ORBITAL TUMOURS: A RADIOLOGICAL AND HISTOLOGIC CORRELATION

DISSERTATION SUBMITTED IN PART FULFILLMENT FOR THE DEGREE OF MASTERS OF MEDICINE IN DIAGNOSTIC RADIOLOGY OF THE UNIVERSITY OF NAIROBI.

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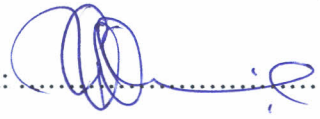
DECLARATION

This dissertation is my original work and has not been presented for a degree in any other university.

Signed: 

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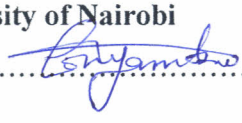
   Signed..... 

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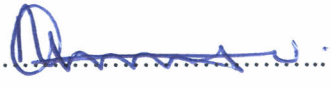
   Signed..... 

DEDICATION

This work is dedicated to Ann, Sharleen and Ambigel the three people who have sacrificed a lot with me in my pursuit of higher learning.

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TABLE OF CONTENTS

ABBREVIATIONS

AIDS	Acquired Immunodeficiency Syndrome.
CNS	Central Nervous System.
C T	Computed Tomography Scan.
IOID	Idiopathic Orbital Inflammatory Disease
DDR	Department of Diagnostic Imaging and Radiation medicine.
K.N.H	Kenyatta National Hospital.
MALT	Mucosa Associated Lymphoid Tissue.
MRI	Magnetic Resonance Imaging.
PP	Page.
SPSS	Statistical Packages of Social Sciences.
UON	University of Nairobi.
U/S	Ultrasonography

ABSTRACT

Introduction

Orbital tumours constitute a heterogeneous array of benign and malignant lesions. These tumours lead to morbidity and mortality in affected patients. Diagnosis of orbital tumours depends on appropriate imaging and histology results.

Objectives

The main objective of this study was to determine the level of correlation between computed tomography diagnosis and histologic diagnosis of commonly occurring orbital tumours in KNH.

Methods

This was a one year retrospective cross sectional hospital based study done from June 2007 to June 2008 at Kenyatta National Hospital (KNH). It included all patients seen in KNH during the period of study with the relevant computed tomography and histology results. The imaging modality evaluated was Computed Tomography (CT). A correlation was then made between computed tomography and histologic diagnosis and the results presented with the aid of tables, pie charts and graphs.

Results

A total of 35 cases were recorded over a one year period and were later analyzed. The most prevalent tumour overall was Retinoblastoma 20%, followed by squamous cell carcinoma 11% and Rhabdomyosarcoma 9%. The overall mean age was 20.6 years. In at least 68.6% of cases the radiological and histologic diagnosis correlated. Overall there was no statically significant gender predominance.

Conclusion.

There is a high level of correlation between computed tomography and histopathology diagnosis of orbital tumours especially taking into account that computed tomography was mainly done on patients whose diagnosis could not be made clinically and on Ultrasonography (U/S). The high level of correlation is essential for optimal patient management.

INTRODUCTION

Orbital tumours constitute a heterogeneous array of benign and malignant lesions. The tumours arise from a variety of tissues within the relative confines of the orbit as well as metastases to the orbit and tumours due to contiguous spread.

Orbital tumours constitute a significant proportion of orbital diseases. In a study done in Congo whose aim was to determine the occurrence of different orbital diseases, it was found that orbital tumours made up 45 % of orbital diseases. The other significant orbital diseases included inflammatory conditions and injuries. This study concluded that early diagnosis is essential for appropriate management. (1)

Given the value attached to human eyesight and the variety of structures within the relative confines of the orbit a systematic approach is necessary to achieve an accurate diagnosis which is the basis of appropriate management. This includes clinical history, examination and imaging. Before the advent of computed tomography, U/S was the only modality for imaging orbital tumours. It has the advantage of not exposing the patient to ionizing radiation however it is unable to show orbital wall involvement as well as intracranial extension.

COMPUTED TOMOGRAPHY

Imaging of the orbit has progressed very rapidly since the introduction of cross sectional imaging which include computed tomography, ultrasonography and magnetic resonance imaging. (2)

Since the advent of CT the imaging of the orbit has improved significantly. This is due to its cross-sectional nature and also ability to reformat. This technique is easily available and because of the speed of CT, motion artifacts are eliminated which is the main drawback of MRI in investigation of orbital tumours. It is also capable of depicting clearly small osseous and soft tissues lesions which are not visible on plain X-rays. Better visualization of orbital tumours can be enhanced by the use of intravenous iodinated contrast medium. It can also be done on out-patient basis and the fact that it is non-invasive is an added advantage.

In this study the CT were done by radiographer and the images were printed on hard copies However its main drawback is its radiation dosage to the patient and especially to the lens. The total radiation dose to the patient in standard CT examination of the orbit varies between 12 to 21 rads. Hypersensitivity reactions to iodinated contrast medium may also occur. However this is becoming less of a problem with introduction of non-ionic contrast medium.

LITERATURE REVIEW

In 2003 Samira et al at KNH did a cross- sectional hospital based study. The aim of the study was to determine the correlation of clinical, diagnostic imaging and histologic findings in patients presenting with orbital tumours. She also determined the etiology of proptosis and the accuracy of CT diagnosis in these patients. A total of 39 patients were studied. A detailed examination of the eyes and whole body was followed by U/S and CT. It was found that the accuracy of CT diagnosis was 71.4% for rabdomyosarcoma, 66.7%for Burkitt's lymphoma, 50% in benign lacrimal gland tumour and non specific in squamous cell tumour. It was concluded that CT diagnosis could be more accurate if more information was given to the radiologist. (2)

In 1989 Malakwen et al did a cross sectional hospital based study at KNH .The aim of the study was to determine the pattern and characteristic of orbital tumours by use of CT and U/S. He also determined the advantages and disadvantages of these U/S and CT. He studied a total of 38 patients. In all the patients CT and U/S were done .He concluded that these forms of orbital imaging provide a good degree of accuracy but they were not available outside Nairobi. He also found that U/S was affordable to the patients while CT was expensive and only a few patients could afford. Malakwen et al recommended that there was need for radiologist specialised in orbital pathology. (3)
Unlike when this study was done, CT and U/S are now more widely available and so can be used to evaluate orbital tumours

In 1998 Luo et al and associates did a retrospective study on orbital invasion in nasopharyngeal carcinoma and evaluation with computed tomography .The aim of the study was to determine the role of CT in documenting orbital invasion and determining the pathway of tumour spread. A total of 562 patients with histologic proven nasopharyngeal tumours were examined using CT. They concluded that CT provides essential information in documenting orbital invasion and determining the pathway of tumour spread. The study also showed that coronal sections best showed these findings on CT

In 2004 Xiao LH and associates did a retrospective study on diagnosis by U/S and CT in orbital vascular tumours and malformations in china .The aim of this study was to analyze the value of CT in the diagnosis of orbital vascular tumours and malformations .One hundred and eight cases of orbital vascular tumours and malformations were studied. In one hundred and eight cases examined by CT the tumour could be diagnosed by CT in all cases (100%).The nature of the tumour could be determined by CT in 92% of cases pre-operatively. They concluded that CT can estimate the nature, extent and location of orbital vascular tumours and malformations and can be useful for selection of the route of orbital operation. (5)

ANATOMY OF THE ORBIT

The orbits are formed by a bony skeleton. The orbit has four walls, the roof, the medial wall, lateral wall and the floor .These walls form a cavity which is pyramidal in shape with the apex situated posteriorly and the base anteriorly.

The roof of the orbit is formed by the orbital plate of the frontal bone anteriorly. It is completed posteriorly by the lesser wing of sphenoid. It separates the orbit from the anterior cranial fossa. Posteriorly there is a communication between the middle cranial fossa and the orbit at the junction of the roof and the medial wall.

The medial wall of the orbit is thin: it is formed by the frontal process of the maxilla, the lacrimal bone, the orbital plate of ethmoid and the body of the sphenoid bone. It is related to the lacrimal groove, ethmoidal air sinuses and sphenoidal air sinuses.

The lateral wall of the orbit is the thickest and is formed by the frontal process of the zygomatic bone anteriorly and posteriorly by greater wing of the sphenoid bone. The lateral wall separates the orbit from the temporal fossa and the middle cranial fossa respectively.
The floor of the orbit is formed mainly by the orbital surface of the maxilla, by the lower part of the orbital surface of zygomatic bone and by the palatine bone posteriorly. It is relatively thin and forms the roof of the maxillary sinuses. (6)

Contents of the orbit.

The extraocular muscles are the levator palpebral superioris, superior rectus, medial rectus, inferior rectus, lateral rectus, superior oblique and inferior oblique muscles. The four rectus muscles arise from the common annulus of Zinn and insert on the upper, medial, inferior and lateral surface of the sclera of the eyeball respectively. The muscles are joined together by connective tissue thus forming the intraconal and extraconal spaces. (6).

The periorbita is continuous posteriorly with the dura matter and the sheath of the optic nerve. It covers the bony orbit and is continuous with the periosteum covering the orbital margin. It is loosely attached to the bony orbit and can be easily stripped .It is firmly attached at the fibrous pulley of the tendon of superior oblique muscle, at the lacrimal groove and at the suture lines. At the inferior and superior margins of the orbit it sends off projections, which form the orbital septum. This divides the orbit into a preseptal space and postseptal space. (6)

The nerves of the orbit include the optic nerve (II), Oculomotor nerve (III), Trochlea nerve (IV), Abducent (VI), and the sympathetic nerves. The trochlea nerve supplies the superior oblique muscle; the abducent nerve supplies the lateral rectus muscle while the rest of the extraocular muscles are supplied by the oculomotor nerve. (6)

The globe is spherical and consists of three layers. The outer layer called the sclera, the middle layer that consists of the choroid, iris and the cilliary body collectively known as the uveal tract and the inner layer the retina which is the photosensitive layer.

The globe is divided into an anterior chamber and a posterior chamber by the lens. The anterior chamber contains the aqueous humour and the posterior chamber the vitreous humour and the retina.

The blood supply of the orbit is from the intracranial part of the internal carotid artery via the ophthalmic artery. It is given off as the first branch of the internal carotid artery medial to the anterior clinoid process .The artery enters the orbit through the optic canal together with the optic nerve both of which lie in a common dural sheath .The artery gives off branches to the orbit, the main ones being the central artery of the retina, the lacrimal branch and other branches from the main trunk. The venous drainage is via the superior and inferior ophthalmic veins. The lymphatic drainage is into the preauricular and submandibular lymph nodes. (6)

PATHOLOGY

ORBITAL NEOPLASMS

Neoplasm of the orbit can be primary, secondary from adjacent structures or metastases. Orbital neoplasms can be classified as benign or malignant. This classification is may be deficient as it does not take into account the fact that some of the orbital neoplasms can be both malignant and benign.

In this study an anatomical classification will be used. The classification will be based on the tissues of origin of the various orbital tumours. The involved surgical space and other radiological characteristics like contrast enhancement are also used.

CLASSIFICATION OF ORBITAL NEOPLASMS

1. Lacrimal gland tumours.
2. Tumours of vascular origin for example cavernous and capillary haemangioma.
3. Nerve tumours.
4. Muscular tumours of the orbit.
5. Intraocular tumours for example retinoblastoma
6. Metastasis
7. Others

LACRIMAL GLAND TUMOURS

The lacrimal gland is a minor salivary gland hence demonstrates pathology similar to other salivary glands in the region of head and neck. Fifty percent of salivary gland tumours are epithelial in origin and half of these are benign and the other half are malignant. Patients with lacrimal gland tumours present with superolateral orbital mass and inferomedial proptoses. (7)

The most common lacrimal gland tumour is the benign mixed tumour also known as pleomorphic adenoma. Adenocystic carcinoma is the most common lacrimal gland malignancy. Other lacrimal gland tumours include the dermoid and epidermoid tumours and epithelial cysts. (7)

Benign mixed tumour

It is also known as pleomorphic adenoma and is the most common benign tumour of the lacrimal gland .It is usually located in the superolateral extraconal space and majority originate in the orbital lobe of the lacrimal gland. It presents in the middle age by which time it may have been present for 1-2 years. It has a male preponderance. The tumour is well-encapsulated and grows slowly by expansion and may show calcification.

On CT the tumour shows a homogenous density soft tissue mass. It may show remodeling of the lacrimal fossa and punctate calcification. On enhanced CT it shows mild contrast enhancement.

On MRI it is hypointense on T1WI and hyperintense on T2WI with heterogeneous areas and shows moderate to marked enhancement.

Treatment is usually by surgical removal and recurrence is rare. (7)

Adenoid cystic carcinoma

This is the most common carcinoma of the lacrimal gland. This tumour can arise from the benign pleomorphic adenoma or may a rise de-novo in the lacrimal gland. It has an extraconal origin in the orbital lobe of the lacrimal gland, is a soft tissue mass rapidly growing and classically invasive. It is not encapsulated .Presentation with pain and parasthesia is characteristic. It occurs in young adults to old age with a peak in the fourth decade. Male to female ratio is equal in adults.

CT demonstrates a poorly defined isodense homogeneous soft tissue mass with marked focal enhancement on intravenous contrast administration. Erosion of the lateral wall of the orbit is common. It may be mimicked by pseudotumour or Wegeners granulomatosis thus need for histology.

Treatment is by radical surgical resection of early tumour with wide margins and may include exenteration but recurrence is common .It has a poor prognosis and high mortality. (7)

Tumours of Vascular Origin

These comprises of capillary haemangiomas, cavernous haemangiomas, lymphangiomas, and haemangiopericytoma. They represent about 15% of retrobulbar tumours.

Capillary haemangiomas

This is a benign tumour of infancy and arises from the endothelial cells. It is most commonly located in the superior medial extraconal space. It occurs at birth in 30% and most in the first six months of life. Clinically it presents as a unilateral soft tissue mass with bluish discoloration of the skin and enlarges on crying or valsalva. Capillary haemangioma is not capsulated as opposed to cavernous haemangioma .It may show secondary remodeling and enlargement of the orbit without bone destruction.

The CT findings are of a slightly hyperdense lobulated mass of homogenous density which may also be infiltrative. It shows marked contrast enhancement. On MRI the tumour is hyperintense on T1WI and T2WI images and shows diffuse intense enhancement. It may regress spontaneously after one year. Treatment is by expectant observation unless complications occur in which case corticosteroid treatment, intratumoural laser therapy or surgical ligation may be done. (8)

Cavernous Haemangioma

It is a venous malformation of the orbit, characterized by large cavernous spaces. Most of the tumours occur in the intraconal space; however the tumour can occur anywhere in the orbit. It is the most common benign orbital tumour in adults. Age at presentation is 10 to 60years with a peak at 40years. Clinical presentation is by slowly progressive painless proptosis. Tumours may also present with pressure symptoms especially when located at the

orbital apex. CT shows a well-defined homogenous isodense mass with marked enhancement after intravenous contrast administration. (8)

Haemangiopericytoma

This is a vascular tumour occurring more commonly in adults but can occur in children. It shows similar clinical presentation and appearance as the cavernous haemangioma. Both tumours are encapsulated, and on CT show homogenous density and marked enhancement on intravenous contrasts administration .Haemangiopericytoma can be both benign and malignant, it thus may show bone destruction and muscle infiltration and distant metastasis. Treatment is by surgical excision but it may show recurrence. (8)

Lymphangioma

Lymphangioma is a harmatoma and arises from the primitive vascular tree. They are composed of solid and cystic material including fibrous tissue,dilated lymphatic channels and lymphoid tissue, they present in childhood with pressure symptoms.

Lymphangioma occur in extraconal space but can cross boundaries. CT shows a poorly defined, lobulated mass, of mixed attenuation with variable enhancement on intravenous contrast medium. (9)

Tumours of the optic nerve

Neural tumours arise from the optic nerve itself or its dural sheath or from other cranial nerves within the orbit. These tumours include the optic nerve glioma and the optic nerve sheath meningioma. Optic nerve glioma can arise from the intraorbital part of the optic nerve inside the orbit and thus just like the optic nerve sheath meningioma is a recognized tumour of the orbit. The presentation of the two tumours also resembles other orbital tumours. (9, 10)

Optic Nerve Glioma

Optic nerve glioma is an uncommon tumour and occurs more commonly in children aged 5-10years.It is more common in females. This tumour may arise in any part of the visual pathway from the nerve to the optic tract. The tumours commonly arise from the pilocytic astrocyte thus they are commonly juvenile pilocytic astrocytomas.

Optic nerve gliomas are associated with Neurofibromatosis one (NFI) in 30-40% of cases while 10-15% of patients with NF1 have optic nerve gliomas. NFI is an autosomal

condition with the gene, a tumour suppressor located on the long arm of chromosome 17 q11 especially with bilateral optic nerve gliomas. High-grade tumours rarely involve the optic nerve. (9, 10)

CT shows fusiform or uniform thickening of the entire optic nerve with mild contrast enhancement. MRI is more sensitive than CT in evaluation of intracanalicular and intracranial extension. It is isointense to muscle on TIWI and hypointense on T2WI.It may also show nerve kinked behind the globe both on CT and MRI.The main differential diagnosis is optic nerve sheath meningioma. (10, 11)

Optic nerve sheath meningioma

Meningioma in the orbit may be either primary or secondary. Primary meningioma originate from the intraorbital optic nerve sheath. Secondary meningioma originate along the sphenoid wing and subsequently invade the orbit.
Secondary meningioma is much more common than primary intraorbital meningioma and account for 90% of all meningioma in the orbit. Meningioma is slow growing tumour. (9, 10)

Primary meningioma is found most often in middle-aged females but can occur in children where it is more aggressive. It is occasionally associated with neurofibromatosis type two (NF2) in juvenile patients. The tumour originates from the optic nerve sheath and may circumferentially encase the nerve.

Clinical presentation is with progressive loss of visual acuity, papilloedema and pallor of the optic nerve.

CT is the imaging modality of choice and shows a hyperdense mass surrounding the nerve. The mass may contain calcification and enhances markedly with intravenous contrast. It may also show tram-track appearance that is characteristic but not pathognomonic. (9, 10)

Sphenoid wing meningioma is also known as hyperostotic meningioma en plaque. It presents with slowly progressive unilateral painless exophthalmos, numbness in the distribution of the trigeminal nerve and sometimes headaches and seizures. CT shows a hyperdense mass

pushing the eyeball from the lateral side. It may also show calcification and hyperostosis of the bone .It enhances markedly with intravenous contrast medium. (10)

MUSCULAR TUMOURS

Rhabdomyosarcoma

This is the most common primary malignancy in the pediatric age group. It can occur from birth to adulthood; with the average presenting age being 6yrs. Rhabdomyosarcoma is mostly intraconal but can cross boundaries. It presents with rapidly progressive exophthalmos and proptosis.

Bone destruction and extension into the Paranasal sinuses are common with rhabdomyosarcomas and spread outside the orbit showing a poor prognosis especially with intracranial extension.

CT shows a mass isodense to the extraoccular muscle which it may involve; there could also be bone destruction and extraorbital involvement.
Treatment is by surgical debulking, chemotherapy and radiotherapy to which the tumour shows good response. (11)

ORBITAL LYMPHOMAS

Orbital lymphomas are uncommon and when they occur they are more commonly non-Hodgkin in type. They occur without systemic disease, which develops subsequently in a third of patients especially when bilateral .There has been an increase in the orbital lymphomas due to increased occurrence in Aids patients.

Lymphoma may occur at any age but is commonly seen in 30 to 40years age group. It is more common in females. It shows slow growth and variable presentations.

CT shows an infiltrating hypodense mass with moderate enhancement on intravenous contrast medium administration. (12)

INTRAOCULAR TUMOURS

Retinoblastoma

Retinoblastoma is the most common intraocular tumour in childhood with a mean age of presentation at 18 months with more than 90% of cases diagnosed in patients below 3 years. The non hereditary form accounts for 94% and familial form 6%.Retinoblastoma can be bilateral, trilateral or quadrilateral with a suprasellar cistern focus.

Clinical presentation is determined by the extent of the tumour at the time of diagnosis with the most common presenting symptoms being leukocoria, strabismus, ocular inflammation and proptoses. In a rare case lesion could be found on routine examination. (13)

Retinoblastoma is highly malignant and aggressive premature neuroectodermal tumour with frequent intraocular, transcleral or haematogenous spread.

Ultrasound in good hands is up to 95% diagnostic. (13)

CT shows a solid smoothly marginated lobulated retrolental hyperdense mass with calcification. Tumour shows marked enhancement on intravenous contrast injection. (13, 14)

MELANOMA

Melanoma is the commonest primary tumour of the globe in adults. It arises from the choroid layer of the globe. In our setup melanoma is rare.(15) It is highly invasive and recurrence is common. Metastasis is common to the brain, liver, chest and bone.

CT shows an ill-defined hyperdense thickening of the globe. It enhances markedly with intravenous contrast medium. (15)

METASTASES

Metastases to the orbit are more common than primary malignancies in adults. Patients usually have a known primary malignancy elsewhere, the commonest site being the breast, kidney and lung .In children wilms tumour is a common source of metastasis.

CT shows ill-defined mass of variable density to the vitreous cavity and variable contrast enhancement. (16)

RESEARCH QUESTION

What is the pattern of presentation of commonly occurring orbital tumours in KNH and what is the level of correlation between computed tomography diagnosis and histology diagnosis?

Justification

The research review done shows that there is no consistent data in the sensitivity of computed tomography diagnosis of orbital tumours in Kenya. Computed tomography is an important imaging modality in the diagnosis of orbital tumours as well as the intracranial extension of orbital tumours. The only study that attempted to document the sensitivity of CT in diagnosis of orbital tumours in Kenya was done by Samira .The patients sampled did not have computed tomography as the only mode of imaging. Some of the patients had both U/S and CT while others it was either U/S or CT alone. The results that were obtained from this study are therefore not representative and consistent with other studies done elsewhere. This study aims to address the above shortcoming of Samira's study and document the sensitivity of CT in the diagnosis of orbital tumours in Kenya.

OBJECTIVES

Broad objectives

The main objective of this study was to determine the level of correlation between computed tomography diagnosis and histology diagnosis of commonly occurring orbital tumours in KNH.

Specific objectives

1). Determine the CT findings of commonly diagnosed orbital tumours at KNH

2). Determine the histology results of these tumours, and then correlate the CT diagnosis and the histological diagnosis.

3).Asses the involved surgical compartment of commonly occurring orbital tumours.

MATERIALS AND METHODOLOGY

Study Area

This study was conducted at Kenyatta National Hospital. Kenyatta National Hospital is both a referral hospital as well as a teaching hospital in Kenya. It has modern diagnostic equipment including U/S, CT and MRI.

Study Population

The study was designed to include all patients who had been seen at KNH Ophthalmology, Neurosurgery and Radiology Departments with a CT diagnosis of orbital tumour, and histology results. Patient details were entered into data collection forms by the researcher. The details were entered into computer software and analyzed.

Study Design

This was a one year retrospective cross-sectional hospital based study done from June 2007 to June 2008 at KNH. Kenyatta National Hospital being both a referral hospital as well as a teaching hospital in Kenya thus the only patients seen were referrals from other hospitals. The tumours were classified as intraconal, extraconal, intraconal and extraconal. They were also classified as well defined or diffuse.

The limitations of this classification include the fact that tumours are not confined to one surgical space. It was not always possible to define the surgical space that the tumour originated .Tumours arising from the extraocular muscles that define the surgical spaces posed a challenge placing them in the surgical spaces.

The CT was done by radiographers under the guidance of a radiologist. The necessary precontrast and postcontrast axial views are taken and then reviewed by the radiologist. The radiologist can then recommend for reconstructions as necessary. The CT was reviewed by the researcher and a qualified radiologist on the counsel.

The biopsies were done by the opthamologist under anaesthetic and reported by a pathologist within a period of three days.

Sample size determination

This was determined using Fisher's formula.

$$n = \frac{z^2 p}{d^2}$$

Where n = Sample size

z = Standard normal deviate value corresponding to 95% confidence level (=1.96)

p = estimated prevalence of orbital tumours. The value used was 45%

d = Degree of precision set at 5%

Using the above formula the calculated sample size was 35. This number of patients was studied.

Sampling method

All patients with orbital tumour, CT and histology result for the period specified and meeting the inclusion criteria were sampled.

Inclusion criteria

Patients of all age groups who had CT diagnosis of orbital tumour and histology results were considered.

Exclusion criteria.

All patients with no CT or histology results for whatever reason were not included in this study. Some patients records where incomplete, inadequate or lost hence they had to be excluded from the study. Some of the patients had a clinical diagnosis of orbital tumours but had no CT or histology results. These patients were also excluded from the study.

Material and procedures

The relevant records were obtained from the records departments. The patients particulars age and sex were obtained from the patients file and entered into data collection sheet.

The histology results were obtained from the patients file and noted on the data collection form appendix 2. Where necessary further clarifications were obtained from the histology departments and records departments of KNH.A correlation was then made of the CT diagnosis and histologic diagnosis.

Data analysis

A separate data collection sheet (appendix 1 and appendix 2) were used for each patient. This data was then entered into a computer and analyzed using Statistical Package for Social Sciences (SPSS). Data cleaning was done by running frequencies and all missing data was corrected by confirmation using the questionnaires. Statistical analysis was carried out.

Study limitations

The study limitations included the following:

1). A short duration of study in this case one year thus not possible to sample some of the rare orbital tumours.

2.) A small sample size due to the relatively short duration of study.

3). This was a retrospective study, a prospective study would have been better.

ETHICAL CONSIDERATIONS

Before commencement of this study, the proposal was submitted to the ethical committee of KNH and written consent for the study was obtained. This was a retrospective study and did not involve direct patient contact. The patients did not incur any danger or expense. In addition, patients name and details were not used and therefore confidentiality was maintained thus there was no need for a signed consent. The results of this study will be delivered to the KNH Ethical committee.

RESULTS

Complete records of 35 patients with a clinical diagnosis of orbital tumours were reviewed. The CT findings were recorded from the CT counsel. The histology results were obtained from the patient files or from the histology department.

Demographic characteristics

A total of 35 patients were reviewed 15 patients were males and 20 were females.

Age distribution

Overall the patients age ranged from 1.67years – 80 years .The mean average age 20.6 years.

Frequency of orbital tumours

Eight commonly diagnosed orbital tumours in KNH were identified and their occurrence was as follows: Retinoblastoma accounted for 20.0%, squamous cell carcinoma 11.4%, Rhabdomyosarcoma 8.6%, Lymphoma 5.7%, dermoid cysts 8.6%, meningioma 5.7%, Idiopathic Orbital Inflammatory Disease (IOID) 5.7%, capillary haemangioma 2.9% and others 31%.

Table showing age frequency of orbital tumours

AGE IN YEARS	FREQUENCY (n)	%
0-5	12	34.3
6-10	6	17.1
11-15	3	8.6
16-20	2	5.7
21-25	1	2.9
26-30	0	0.0
31-35	1	2.9
36-40	2	5.7
41-45	1	2.9
46-50	4	11.4
51-55	1	2.9
56-60	0	0.0
61-65	1	2.9
66-70	0	0.0
71 and above	1	2.9

Table showing frequency of orbital tumours as seen on computed tomography

CT Diagnosis		
	Frequency(n)	%
Advanced Retinoblastoma	6	17.1
Squamous cell carcinoma	3	8.6
Rhabdomyosarcoma	3	8.6
Meningioma	2	5.7
Lymphoma	4	11.4
IOID	1	2.9
Dermoid cysts	3	8.6
Capillary haemangioma	0	0.0
Others	13	37.1

Location of commonly diagnosed orbital tumours at KNH

Surgical spaces	Intraconal space	Limited to Extraconal space	Intraconal and Extraconal space	Intracranial Extension	Total
Advanced Retinoblastoma	0	0	7	0	7
Squamous cell carcinoma	0	4	0	0	4
Lymphoma	0	0	2	0	2
Meningioma	0	0	2	1	2
Dermoid cyst	0	2	0	0	2
Rhabdomyosarcoma	0	0	3	0	3

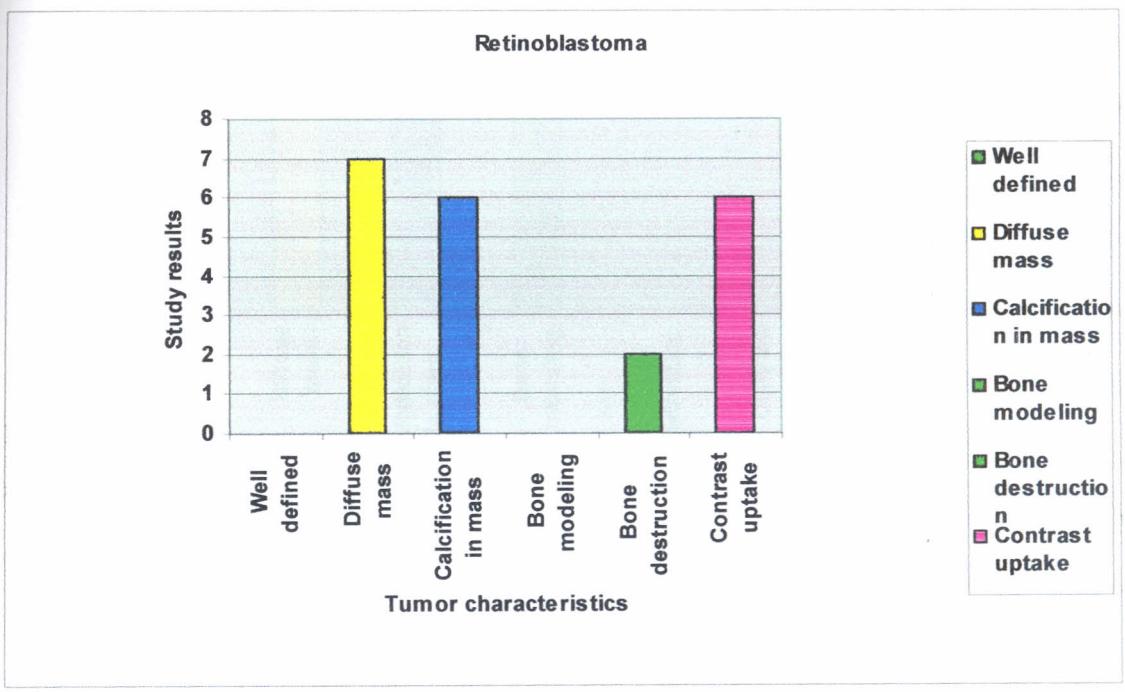
Table showing CT appearance

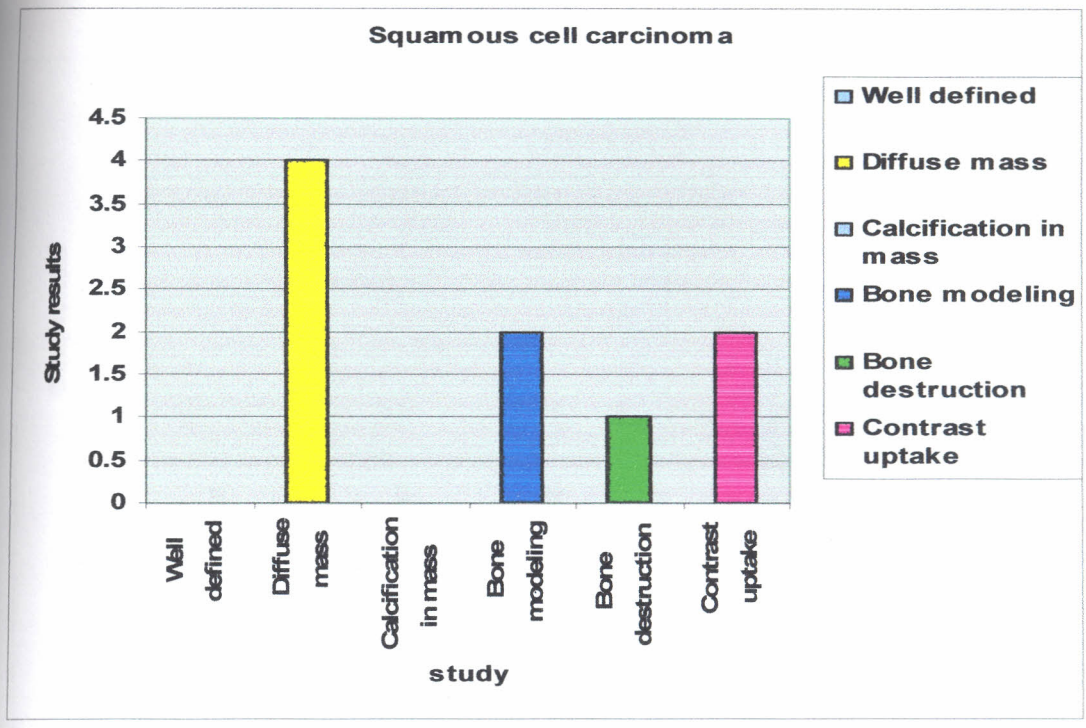
CT density	Hypodense	Isodense	Hyperdense	Heterogeneous	Total
Advanced Retinoblastoma	0	0	6	1	7
Squamous cell carcinoma	0	4	0	0	4
Lymphoma	0	2	0	2	2
Meningioma	0	0	1	1	2
Dermoid cyst	2	0	0	0	2
Rhabdomyosarcoma	0	2	0	1	3

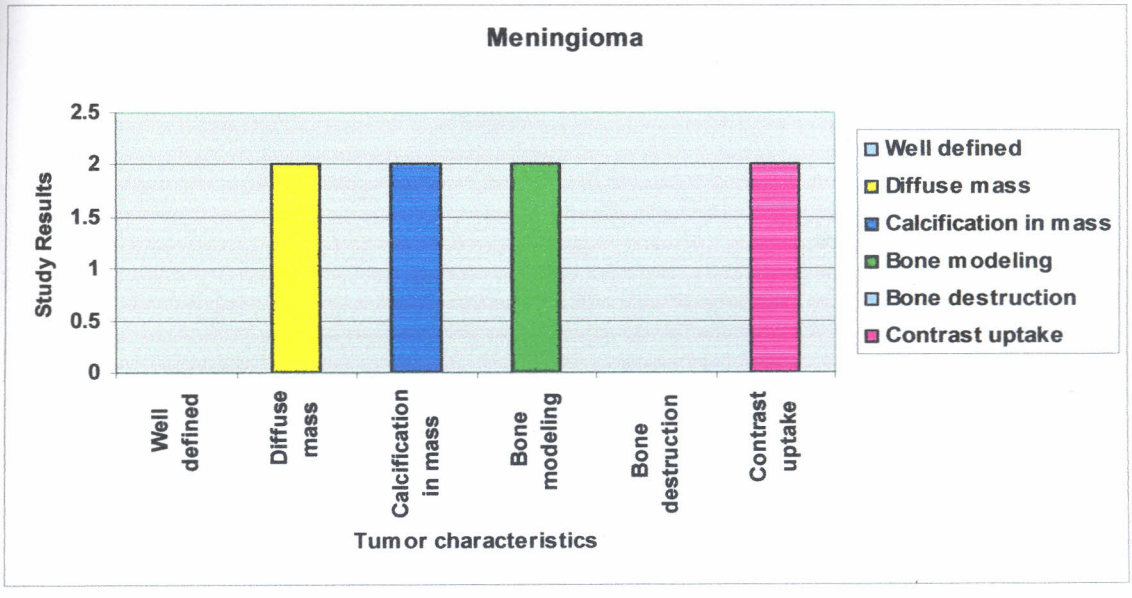
Table showing CT characteristics of commonly diagnosed orbital tumours

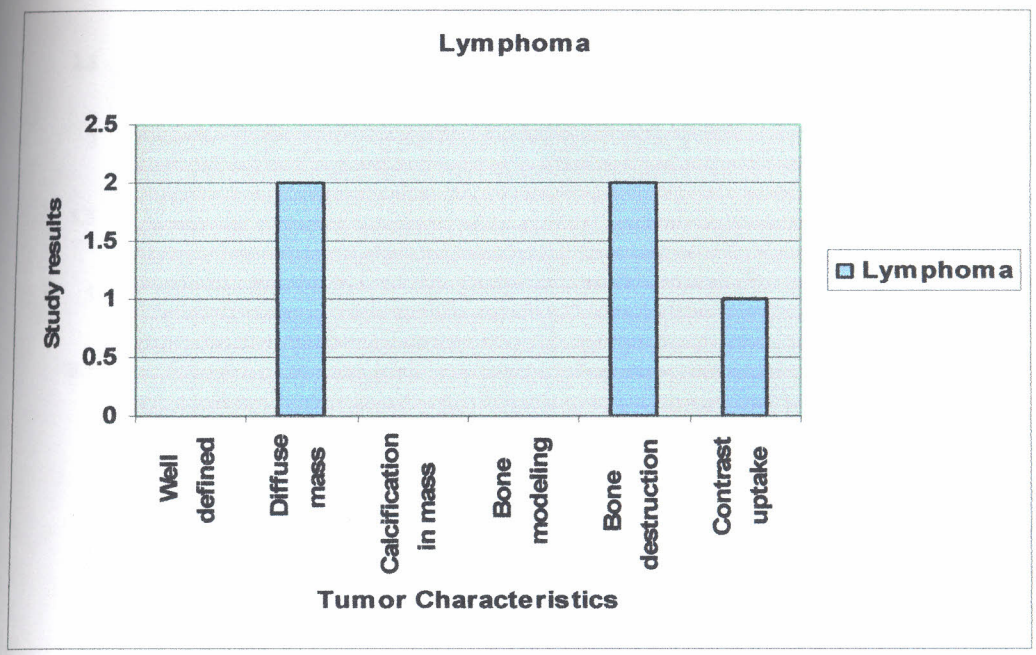
Tumour studied	Well defined	Diffuse mass	Calcification in mass	Bone modelling	Bone destruction	Contrast enhancement
Retinoblastoma (Advanced)	0	7	6	0	2	6
Squamous cell carcinoma	0	4	0	2	1	2
Meningioma	0	2	2	2	0	2
Lymphoma	0	2	0	0	2	1
Rhabdomyosarcoma	0	3	0	0	0	1
Dermoid cyst	2	0	0	2	0	0

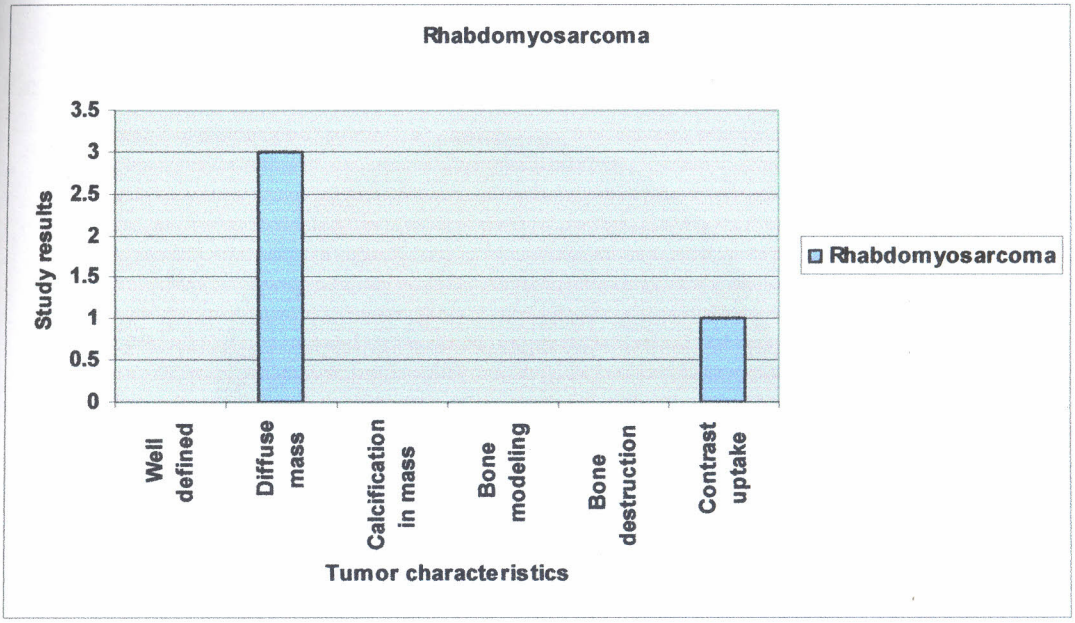
Bar graphs showing CT characteristics of commonly diagnosed orbital tumours

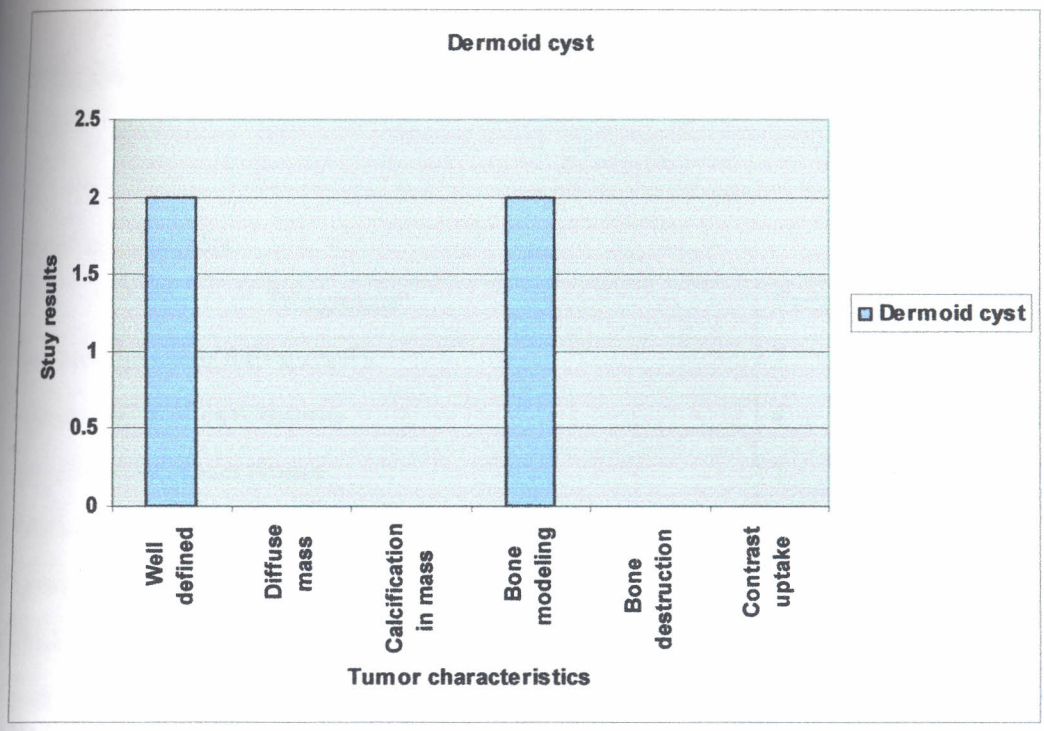



Squamous cell carcinoma
Study results
4.5
4
3.5
3
2.5
2
1.5
1
0.5
0
Well defined
Diffuse mass
Calcification in mass
Bone modeling
Bone destruction
Contrast uptake
study


Meningioma
Study Results
2.5
2
1.5
1
0.5
0
Well defined
Diffuse mass
Calcification in mass
Bone modeling
Bone destruction
Contrast uptake
Tumor characteristics


Lymphoma
Study results
2.5
2
1.5
1
0.5
0
Well defined
Diffuse mass
Calcification in mass
Bone modeling
Bone destruction
Contrast uptake
Tumor Characteristics
Lymphoma


Rhabdomyosarcoma
Study results
3.5
3
2.5
2
1.5
1
0.5
0
Well defined
Diffuse mass
Calcification in mass
Bone modeling
Bone destruction
Contrast uptake
Tumor characteristics
Rhabdomyosarcoma



Pie Chart showing distribution of the tumours as seen on computed tomography

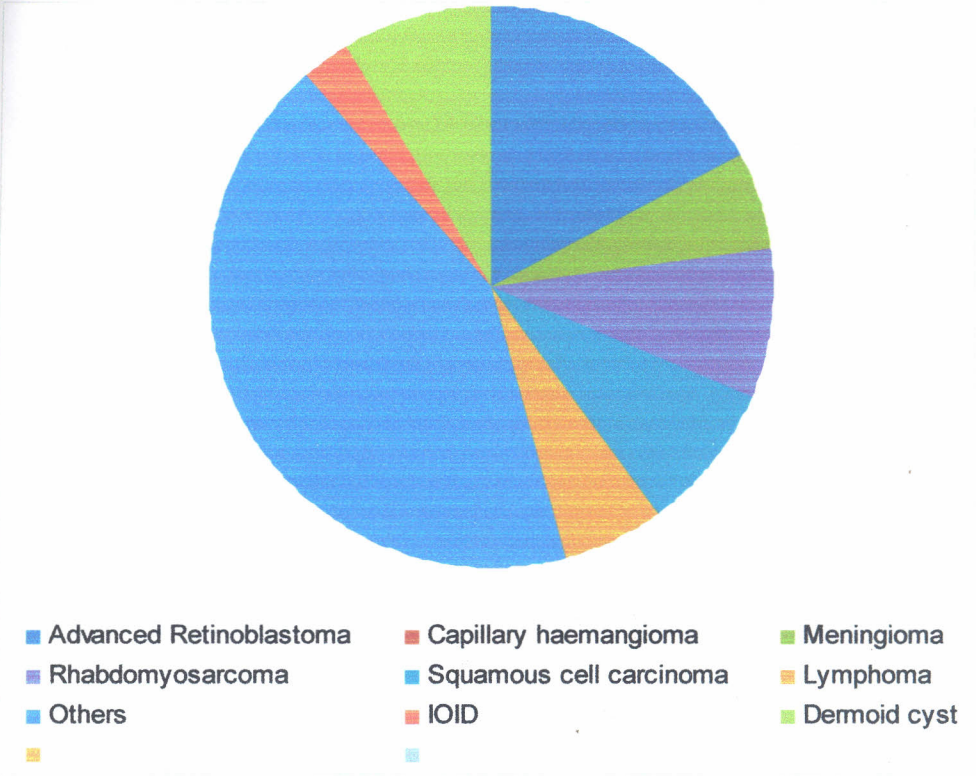
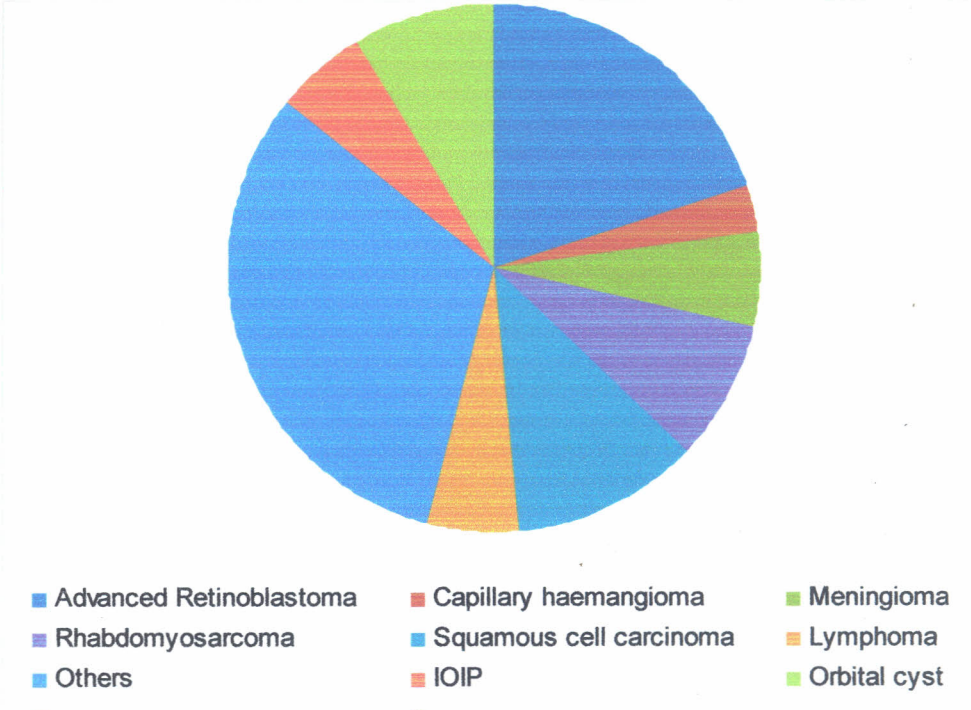


Table showing Histology Results of commonly diagnosed orbital tumours

Histology Results		
Diagnosis	**Frequency(n)**	**%**
Retinoblastoma(Advanced)	**7**	**20.0**
Squamous cell carcinoma	**4**	**11.4**
Capillary haemangioma	**1**	**2.9**
Rhabdomyosarcoma	**3**	**8.6**
Lymphoma	**2**	**5.7**
I.O.I.D	**2**	**5.7**
meningioma	**2**	**5.7**
Dermoid cysts	**2**	**8.7**
Others	**11**	**31.0**

PIE CHART SHOWING PERCENTAGE DISTRIBUTION OF ORBITAL TUMOURS ON HISTOLOGY.



COMPARISON OF CT DIAGNOSIS AND HISTOPATHOLOGY DIAGNOSIS

Diagnosis	CT-scan	Histopathology
Retinoblastoma (advanced)	6	7
Squamous cell carcinoma	3	4
Rhabdomyosarcoma	3	3
Lymphoma	2	2
meningioma	2	2
IOID	1	2
Dermoid cysts	3	3
Capillary haemangioma	0	1
Others	13	11

BARGRAPH SHOWING COMPARISON OF CT DIAGNOSIS AND HISTOPATHOLOGY DIAGNOSIS

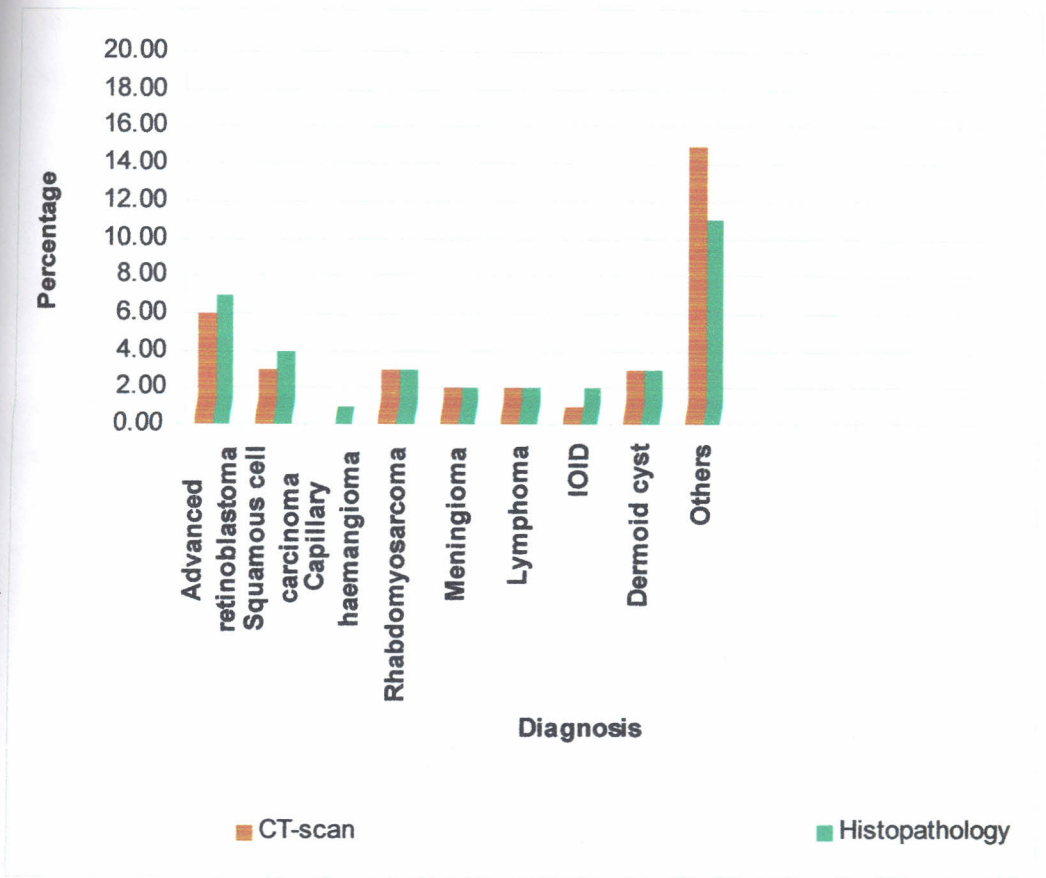


Table showing Correlation

Correlation	
Spearman's rho Correlations	0.686
P - Value	< 0.001

Correlation between Computed tomography and histology diagnosis

The sensitivity of the CT diagnosis as compared with the histology diagnosis was 68.6% using the spearman's rho correlation. This means that the positive predictive value of CT in making a diagnosis in orbital tumours similar to the histological diagnosis is 68.6%. This was statistically significant with a P- Value of <0.001.

Retinoblastoma

The diagnosis of retinoblastoma was first made clinically by an opthamologist and CT was later done. The percentage correlation for this tumour was 85.7%.A total of six patients with retinoblastoma was diagnosed on CT. Histology results showed seven patients had retinoblastoma. One patient with retinoblastoma was not diagnosed on CT. This patient was diagnosed as having rhabdomyosarcoma. The age of the patients ranged from 1.67 to 13 years. The average age was 5years.There were five females and two males, this was not statistically significant. All the patients presented with proptosis and impaired vision in the affected eye .This was because of late presentation in six of the patients and enucleation in one of the patients. Computed tomography showed a diffuse mass with infiltration of adjacent structures, the soft tissue mass was hyperdense with calcification within the mass in 6 out of the 7 cases .The tumours showed bone destruction in 2 out of 7 cases .Contrast enhancement was noted in 6 out of the 7 cases. There was no bone remodelling noted.

Squamous Cell Carcinoma

The percentage correlation for this tumour was 75.0%.This was the second commonly diagnosed orbital tumour at KNH on computed tomography over the specified period of study. The total number diagnosed on CT and confirmed on histology was three. Histology showed four patents had squamous cell carcinoma, thus CT failed to diagnose one patient. This was diagnosed as a sinonasal tumour which had extended to the orbit possibly from the ethmoid sinuses. The patients age ranged from 33years to 80 years, the mean age was 50.5years.The sex distribution was 3 males and 1 female. Computed tomography showed an extraconal diffuse mass which had a tendency to invade adjacent structures. The masses were isodense and had no calcification. Bone remodelling was noted in 2 while bone destruction was noted in one. Two of the tumours showed contrast enhancement .There was no intracranial extension.

Dermoid cyst

The percentage correlation for this tumour was 66.7%.Three patients were diagnosed to have dermoid cyst by computed tomography. Two were confirmed by histology results. Thus CT over diagnosed dermoid cyst. Histology showed the above one patient to have a mucocele. The two patients presented with proptosis and impairment of vision. The patients age were 2years 10 months and 3 ½ years.

The two patients had well defined, extraconal, hypodense masses. The tumours showed no calcification, no contrast enhancement and no infiltration to adjacent tissues. There was no bone destruction or intracranial extension; however there was bone remodelling in the two tumours.

Sphenoid wing meningioma

The percentage correlation for this tumour was 100%.Two patients with sphenoid wing meningioma were diagnosed using computed tomography and were later confirmed by histology results. The patients were aged 17 years and 38 years. The two patients were females and presented with proptosis and impaired vision.

Computed Tomography showed both masses to be in both intraconal and extraconal compartments and to have optic nerve involvement. The two cases showed presence of calcification in the mass, hyperostosis and contrast enhancement. There was no bone destruction; however there was intracranial extension in one of the patients.

Lymphoma

The percentage correlation for this tumour was 50%.Four patients were diagnosed to have orbital lymphoma by computed tomography. Two patients were later confirmed by histology results as having lymphoma. The other two patients had malignant fibrous histiocytoma. The two patients confirmed by histology to have lymphoma were females and were aged eleven years. The patients presented with proptosis and impaired vision. Computed tomography showed diffuse tumours with involvement of both intraconal and extraconal compartments. There was involvement of adjacent structures which included optic nerve and extraocular muscles. The tumours were heterogeneous in appearance with no calcification .There was contrast enhancement in one case while in the other contrast was not given. The two cases showed bone destruction but no intracranial extension.

Rhabdomyosarcoma

The percentage correlation for this tumour was 100%.Three cases of rhabdomyosarcoma were diagnosed on computed tomography and were later confirmed on histology .The patients were females, aged 2, $2^1/_2$ and 7years respectively. The patients presented with proptosis and impaired vision. On computed tomography the tumours were located in both intraconal and extraconal compartments. The extraocular muscles were involved in all the patients and tumours were isodense to extraocular muscles. Contrast enhancement was noted in one of the tumours .There was no bone destruction, remodelling or intracranial extension.

DISCUSSION

In this retrospective study of orbital tumours a total of 35 cases were reviewed using computed tomography and histology results at Kenyatta National Hospital. The patients had CT and histology results. Any patient who did not have CT or histology results was not included in the study.

It was found that of the 35 patients reviewed 15 were males and 20 were females. The sex ratio in this study was 1:1.3.The gender difference was not statistically significant (P>0.05).

In another cross- sectional hospital based study done by Samira et al in KNH a total of 39 patients were studied, she found that 22 were females and 17 were males.(1) This was in keeping with this study.

In another cross sectional hospital based study done in KNH by Malakwen et al to determine the pattern and characteristic of orbital tumours by use of CT and U/S as well as the advantages and disadvantages of these diagnostic criteria, a total of 38 patients were studied, 25 were males and 13 were females. (3)

In another study done in Congo there were 99 (60%) male and 65 (40%) female for a sex ratio of 1.5: 1. (22) These two studies findings are in variance with our study.

The cause of the variance in the two studies can be explained by the small sample size used in our study thus the need for another study with a larger sample size that can be extrapolated to the general population.

The minimum age of the orbital tumours in this study was 1.67 years and maximum age was 80years.The mean age for our study was 20.6years.

In a similar study, Samira et al found the age range to be between 0.33years to 50.0years with a mean 12.27 years. (1)

In a similar study, Malakwen studying a total of 38 patients found that the age ranged from 2years to 70years. (3) These studies are in keeping with age range as our study.

Retinoblastoma was found to be the most common tumour diagnosed accounting for 20.0%,squamous cell carcinoma 11.4%, Rhabdomyosarcoma 8.6%, Lymphoma 5.7%, Dermoid cysts 8.6%, Meningioma 5.7%, Idiopathic Orbital Inflammatory Disease (IOID) 5.7%,and capillary haemangioma 2.9% and others 31%. In another study in Congo retinoblastoma was the most common histologic form, representing 31.7% of all malignant tumours of the eye and adnexa. Metastatic tumours accounted for 9.7% of all cancers.

cancers. Burkitt's lymphoma was seen in 3% of cases. Lymphoma and adenocarcinoma both accounted for 1.8% of all cases of cancers while adenoid cystic carcinoma, liposarcoma and rhabdomyosarcoma accounted for 1.2% of all cases. (22) In this study retinoblastoma was the most common histologic form. This was in keeping with our study. The histologic forms described in this study were the same forms encountered in our study though some were classified as others in our study.

Most patients with retinoblastoma are diagnosed clinically and confirmed with U/S. Computed tomography was mainly done for patients suspected of having intracranial extension or those with a difficult diagnosis.

Six out of the seven patients (86%) had calcification within the mass and also showed contrast enhancement within the mass. This is in keeping with other studies done which showed calcification in 92% and contrast enhancement in 86%.(14)

The seven patients with retinoblastoma had a diffuse mass on CT and bone destruction was noted in two patients .This is not in keeping with the literature as retinoblastoma is an intraocular tumour in early disease and not a diffuse tumour as noted in our study but this can be explained by the fact that our study dwelt mainly with advanced disease. (14)

It was also found that imaging in squamous cell carcinoma was not for diagnostic purpose but for staging and to plan surgery and for suspected intracranial extension. The four patients with squamous cell carcinoma presented with an extraconal diffuse mass and the mass was isodense in four cases on CT. Two cases showed both bone modelling and contrast enhancement while one case showed bone destruction. This is in keeping with the literature as other studies show this tumour to be extraconal in location and can show bone modelling and bone destruction. (14)

Two cases of sphenoid meningioma were reviewed .The tumours were both intraconal and extraconal compartment .One tumour was hyperdense and the other was heterogeneous. The two tumours showed contrast enhancement, calcification and bone modelling. This is in keeping with the literature as meningioma is known to be a hyperdense tumour, with calcification within the tumour and shows moderate contrast enhancement. Optic nerve meningioma was not diagnosed in this study.

For the other tumours imaging was done for diagnostic purposes, to plan surgery and also to follow-up patients post-treatment progress. The findings posed the same challenges as the ones discussed above.

There was good correlation between CT and histology results. The histology result was taken as the gold standard. The sperman's rho correlation was 68.6% with a P value of <0.001.This means that the percentage of accuracy in diagnosing orbital tumour is approximately 68.6% when using computed tomography.

In another cross- sectional hospital based study done by Samira et al in KNH. It was found that the accuracy of CT diagnosis was 71.4%. (1) This is in keeping with our study which showed a correlation of 68.6 %.(23)

In another study done in Tongji Medical University the CT correlation with histological diagnosis was 67.8%. This is in keeping with our study which showed a correlation of 68.6%. (23)

CONCLUSION

1. The age for patients with orbital tumours ranges from1.67years to 80 years.
2. The male to female ratio for orbital tumours is approximately1:1.4
3. The most common tumour was retinoblastoma.
4. Computed tomography is a very useful imaging modality in the diagnosis of orbital tumours.
5. The accuracy of computed tomography in diagnosis of orbital tumours is 68.6%.

RECOMMENDATIONS

1. A detailed history, clinical findings and a provisional diagnosis is necessary to the radiologist reporting orbital computed tomography.
2. A regular clinical meeting should be arranged for ophthalmologists, radiologists and histopathologists where clinical, computed tomography and histopathology diagnoses can be compared.
3. A select group of radiologists should be trained with special interest in imaging of orbital pathology and this would improve the accuracy with which orbital tumours are diagnosed.
4. CT should be made more available and affordable in the district hospitals throughout the country.
5. Another study should be done with a larger sample size that can be extrapolated to the general population.

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APPENDICES

APPENDIX 1

DATA COLLECTION FORM ONE

General Patients Information:

PATIENTS HOSPITAL NUMBER

PATIENTS STUDY NUMBER

PATIENT'S CT-SCAN NUMBER

.

PATIENT'S LAB NUMBER:

AGE:

SEX:

MALE [ ]

FEMALE [ ]

APPENDIX 2

DATA COLLECTION FORM TWO

PATIENTS CLINICAL SUMMARY

[A] CLINICAL DIAGNOSIS

OD: PROPTOSIS [ ]

OS: PROPTOSIS [ ]

[B] IMPAIRED VISION

COMPUTED TOMOGRAPHY FINDINGS

SITE OF THE LESION

1.	Intraconal	[ ]		
2.	Extraconal	[ ]		
3.	Intraconal and Extraconal involvement	[ ]		
4.	Optic Nerve involvement	[ ]		
5.	Muscle involvement	[ ]		
6.	Well defined	[ ]		
7.	Diffuse or infiltration	[ ]		
8.	Density of the lesion ;-(a) Hyperdense	[ ]	(b) Isodense	[ ]
	(c)hypodense	[ ]	(d) heterogeneous	[ ]
9.	Presence of calcification	[ ]		
10.	Contrast enhancement	[ ]		
11.	Bone remodelling	[ ]		
13.	Bone destruction	[ ]		
14.	Intraocular	[ ]		
15.	Intracranial extension	[ ]		

RADIOLOGICAL DIAGNOSIS.....

HISTOPATHOLO RESULTS .....