

Abstract

The phakomatosis syndromes are a group of neural crest disorders that bear many features in common.

They include Sturge-Weber Syndrome, Naevus of Ota, Phakomatosis Pigmentovascularis and Klippel-Trenaunay Syndrome. They have numerous ocular manifestations in common, some of which are described in this case. Glaucoma is one of these manifestations and has peculiar characteristics when seen in association with phakomatosis syndromes.