Clear cell sarcoma of tendons and aponeuroses: histogenesis and mode of treatment.

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Abstract:

Clear cell sarcoma of tendons and aponeuroses is a rare, slow-growing malignant tumor arising from the tendons and aponeuroses of the lower extremity, more often in the foot. Although it appears to be a benign tumor, the clear cell sarcoma can be a malignant sarcoma capable of local recurrences or distant metastases. Microscopically, there is a rather distinctive picture characterized by discrete nests and fascicles composed of spindle cells, and a small round-to-ovoid nucleus containing a central prominent basophilic nucleolus. Since there is some controversy over the best way to treat this disease, and since its histogenesis is still unknown, the authors elected to report the case of a 28-year-old white male for three reasons: first, to illustrate that the tumor is of neural crest derivation; second, to show that an ultrastructural evaluation of the tumor is imperative in some cases; and lastly, to suggest that amputation is the treatment of choice in localized disease.