Neurothekeoma palpebrae in association with multiple superficial angiomyxomas: Tegumental Angiomyxoma-Neurothekeoma syndrome (TAN syndrome)

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Abstract

We report a case of 10-year-old Indian girl with history of multiple superficial angiomyxoma, presented with three months history of painless right upper lid swelling. There were no visual dysfunctions. Previously, the patient had multiple superficial angiomyxoma (left pinna, left upper cheek, left upper limb, chest, right axilla, hard palate) and epidermal cyst (chin). The histopathological specimens were negative to S-100 protein antibody. Systemic review and family history was unremarkable. Excision biopsy and upper lid reconstruction were performed. Intraoperatively the tumor was multilobulated, firm, well encapsulated and did not invade the underlying tarsal plate. Histopathological features of the upperlid tumor were consistent with nerves sheath myxoma (neurothekeoma). To the best of the authors' knowledge, this is the first reported case of neurothekeoma in association with multiple superficial angiomyxoma.

Introduction

Myxoma is a benign mesenchymal tumor with a hypocellular, hypovascular, bland appearance, composed of fibroblasts embedded in an abundant myxoid matrix.[1]

Allen subdivided myxoid lesions into mainstream myxomas (located in soft tissues, located outside the soft tissue) and non-mainstream myxomas (inadequately substantiated myxomas, myxoid soft tissue tumors not regarded as myxomas, myxoid fatty conditions, other soft tissue lesions that are sometimes markedly myxoid, other soft tissue tumors in which myxoid foci may be seen, nonneoplastic myxoid conditions of soft tissue). Superficial angiomyxoma and neurothekeoma (nerve sheath myxoma) are mainstream myxomas of soft tissues. The other three entities considered as mainstream myxomas of soft tissues are intramuscular myxoma, juxta-articular myxoma and aggressive angiomyxoma.[2] Superficial angiomyxoma arises in the dermis and subcutaneous tissue, and is characterized by prominent thin-walled blood vessels,[3] whereas neurothekeoma originates from the peripheral nerve sheath.[4]We report a case of neurothekeoma palpebrae in a patient with history of multiple superficial angiomyxoma.

Case Report

A 10-year-old Indian girl presented with three months' history of painless right upper lid swelling. The tumor began as a small lump involving the lateral half of the upper lid (Figure 1). It underwent gradual enlargement leading to mechanical ptosis which spared the visual axis (Figure 2). There were no visual dysfunctions. Previously, the patient had multiple superficial angiomyxoma (left pinna, left cheek, left upper limb, chest and right axilla, hard palate) and epidermal cyst (chin) (Figure 3). The histopathological specimens were negative to S-100 protein antibody (Figure 4).

Systemic review and family history was unremarkable. Excision biopsy and upper lid reconstruction were performed. Intraoperatively, the tumor was multi-lobulated, firm, well-encapsulated and did not invade the underlying tarsal plate. Histo-pathological features of the upper lid tumor were consistent with nerve sheath myxoma (neurothekeoma) (Figure 5). At 6 months follow up, there is no recurrence.

Discussion

Neurothekeoma palpebrae are extremely rare; only 10 cases have been reported so far. The average age of presentation is 40-year-old with female preponderance. The tumor is slow growing, painless and often misdiagnosed as

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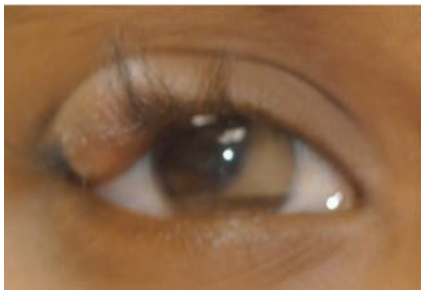

Figure 1. At initial presentation, the tumor was situated at the lateral aspect of the upper lid.

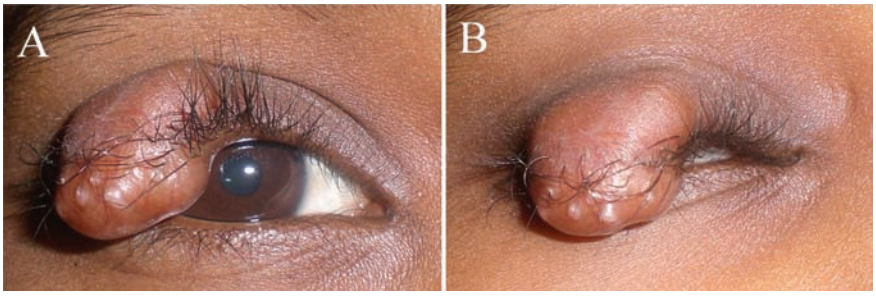

Figure 2. At 4 months, the tumor caused mechanical ptosis sparing the visual axis. The solitary tumor was non-tender, firm, measured 10 mm (horizontal) x 16 mm (vertical). A) The overlying skin was thinned with visible telangiectatic vessels, meibomian glands orifices were enlarged but there was no discharge from the orifices. There was no regional lymphadenopathy. B) Eyelid closure was adequate.

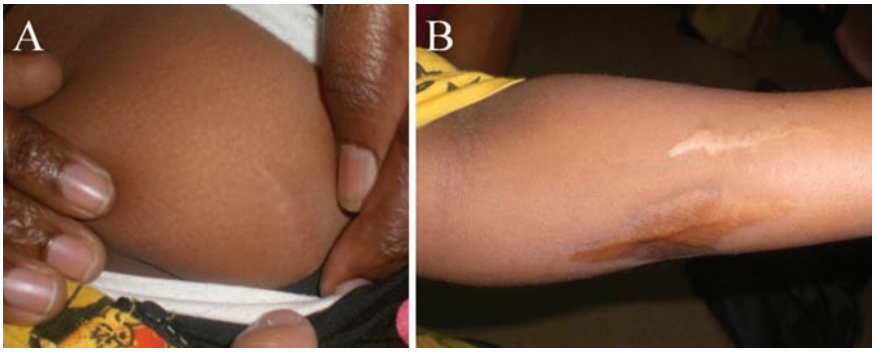

Figure 3. A) Keloid formation over areas of excised superficial angiomyxoma at the chest; and B) left upper limb.

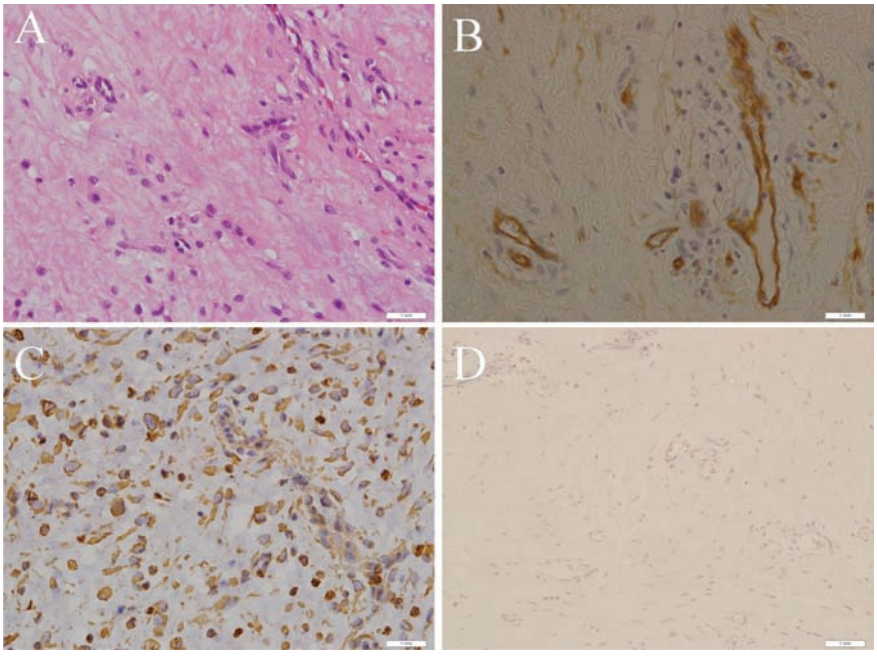

Figure 4.A) Histopathology of superficial angio-myxoma of the hard palate (Specimen number: H 076076). Lobulated tissue composed of proliferation of spindle shaped cells displaying elongated bland nuclei and abundant cytoplasm. The stroma was composed of fibrocollagenous tissue and in areas appeared myxoid with prominent capillary vasculature (original magnification 40x).B) Immunohistochemistry: the vascular channels stained positive to CD 34 (original magnification 40x). C) Immunohistochemistry: sphindle sha-ped cells stained positive to vimentin (original magnification 40x). D) Immunohistochemistry: sphindle sha-ped cells stained negative to S-100 (original magnification 40x). The spindle shaped cells were also negative to SMA, CK, desmin and ALK.

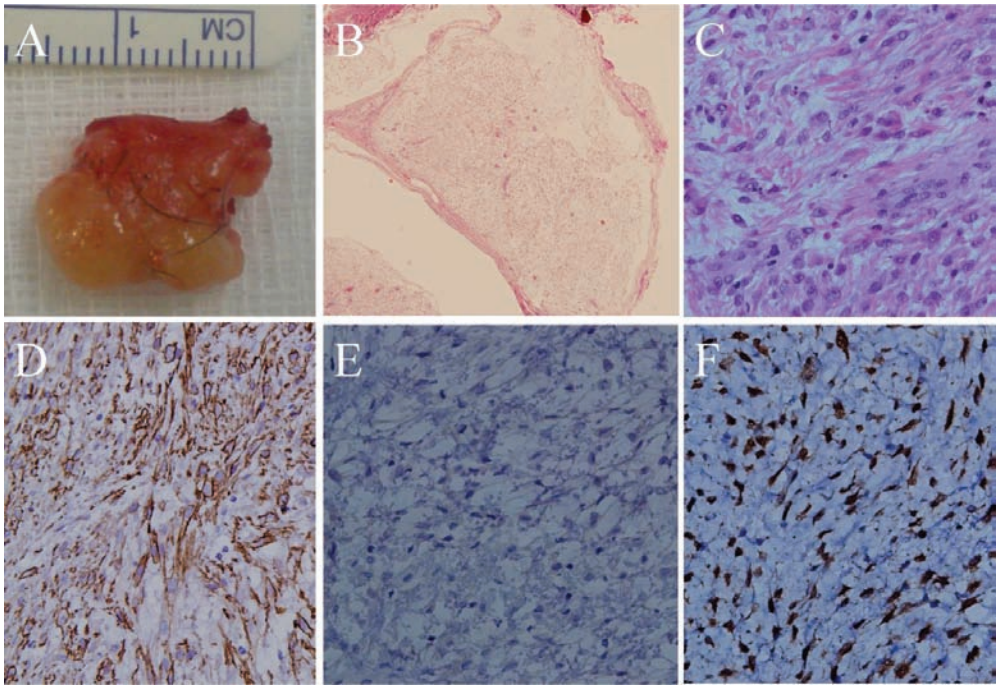

Figure 5. A) Histopathology of upperlid tumour (Specimen number: H 112837). Macroscopy: multi-lobulated, firm tumor removed with intact capsule. B) Histology: well encapsulated tumor (original magnification 1.25x). C) Histology: lobules of spindle to polygonal cells within a myxoid stroma. The cells have eosinophilic cytoplasm and indistinct cytoplasmic membranes. Scattered neutrophils, lymphocytes and plasma cells were present. There were no nuclear atypia, mitosis or necrosis. (original magnification 40x). D) Immunohistochemistry: positivity for CD34 (original magnification 20x) E) Immunohistochemistry: positivity for desmin (original magnification 20x) F) Immunohistochemistry: positivity for S-100 (original magnification 20x).

chalazion or epithelial inclusion cyst.[4]

Malignant transformation has not been reported. Complete resection of myxoma is the best approach to minimize the risk of tumor recurrence. Superficial angiomyxoma and neurothekeoma (nerve sheath myxoma) are mainstream myxomas of soft tissues.[2] Angiomyxomas are derived from fibroblast-like cells, whereas neurothekeomas arise from peripheral nerve sheath cells. Immunohistochemical staining is important to differentiate neurothekeoma from superficial angiomyxoma. Neurothekeomas are positive to S100 antibody, while superficial angiomyxomas are positive to for CD34.[5] Multiple superficial angiomyxomas are associated with Carney syndrome and LAMB (lentigines, atrial myxomas, muco-cutaneous myxomas, and blue naevi).[6] The occurrence of multiple superficial angiomyxoma in our patient is sporadic although some myxoma syndrome are inherited in a autosomal dominant manner. To the best of the authors' knowledge, this is the first reported case of neurothekeoma in association with multiple superficial angiomyxomas. Therefore we take the liberty to name this new syndrome: tegumental angiomyxoma-neurothekeoma (TAN) syndrome.

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