

A Well-Known Lesion in An Unusual Location: Infantile Myofibroma of the Eyelid: A Case Report and Review of Literature

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Abstract- Myofibroma is a neoplasia of myofibroblasts that can be solitary or multiple and it is found most commonly in the head & neck region including scalp, forehead, parotid region and oral cavity. In the eyelid it is rarely reported. It has a benign course in the solitary form and fatal in its multiple form. A 4 month male infant referred to Farabi hospital –the referral center for eye diseases- with a 2 month history of a mass in his eyelid with gradual enlargement with no other complaints. The only abnormal physical finding was a 2.5 cm mass in the eyelid. This mass was excised and sent to the hospital pathology laboratory. When confronting a spindle cell lesion with a nodular or multinodular growth pattern which appears biphasic due to alteration of light and dark staining areas, the surgical pathologist should think to the possibility of myofibroma. Its pattern of growth and architecture rules out the other differential diagnoses like nodular fasciitis, fibrous histiocytoma, infantile fibromatosis, and peripheral primitive neuroectodermal tumor, mesenchymal chondrosarcoma, malignant hemangiopericytoma, juvenile fibrosarcoma and poorly differentiated synovial sarcoma. In difficult cases immunohistochemical staining is helpful that is Vimentin & Actin positivity & Desmin, CK, EMA & S100 negativity.

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Key words: Myofibroma; myofibromatosis; eyelids; infant

Introduction

Various classifications for eyelid tumors exist .One that is more practical categorized them as epithelial (lick squamous papilloma, BCC, SCC), melanocytic, adnexal, vascular, neurogenic, xanthomatous, cystic, inflammatory, fibroblastic, systemic diseases (like amyloidosis), other lesions (like granular cell tumor) and lacrimal apparatus lesions (1). Myofibroma in this location is rarely reported (2). Myofibroma was initially described in 1951 by Williams and Schrum, who designated the lesions as congenital fibrosarcoma. The lesion measures a few millimeters to several centimeters in diameter. Some are deeply seated. Multifocal pattern of involvement is seen (myofibromatosis). Solitary nodules are found most commonly in the general region of the head and neck, including the scalp, forehead, orbit, parotid region and oral cavity. The trunk is the 2nd most commonly affected site, followed by the lower and upper extremities. There are reports of solitary

intraosseous myofibromas most of which have involved the craniofacial bones. Visceral involvement is rare. In patients with multiple lesions (myofibromatosis), the individual nodules have essentially the same appearance as solitary nodules, they occur in the internal organs ,skeleton and demis & subcutis. Up to 40% have visceral lesions that are invariably present at birth. The nodules may be numerous.

Apart from the soft tissues and the skeleton ,the most common sites of organ involvement are the lung, heart, gastrointestinal tract and pancreas and rarely the CNS. Internal lesions often cause symptoms such as severe respiratory distress, vomiting or diarrhea which often fail to respond to therapy and prove fatal within a few days or weeks after birth. Other cause few symptoms, making it likely that some internal lesions remain unrecognized. The nodules grow principally during the immediate perinatal period, and continued enlargement or formation of new nodules may be observed during infancy or even later in life. Radiographically, the bone

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lesions are circumscribed lytic areas with marginal sclerosis and without penetration of the cortex in most cases. Extra osseous lesions may show weak radiodensity as a result of focal calcification (3).

Case Report

A 4 month male infant referred to Farabi hospital- the referral center for eye diseases-due to a mass in his left upper lid since 2 month ago with a gradual enlargement

with no other complaint .Physical examination revealed a firm solid mass m 2.5cm in diameter. Ocular physical examination and systemic examination was unremarkable.CBC was WBC=5100 Hb =10.2 MCV=74 MCH=23.2 Plt=398000 BS=89 Urea=10 Cr=0.5 the excisional biopsy was done.

Gross examination of the tumoral lesion showed a lobulated solid tan mass with white-gray cut sections m. 2.5×2×1.5cm

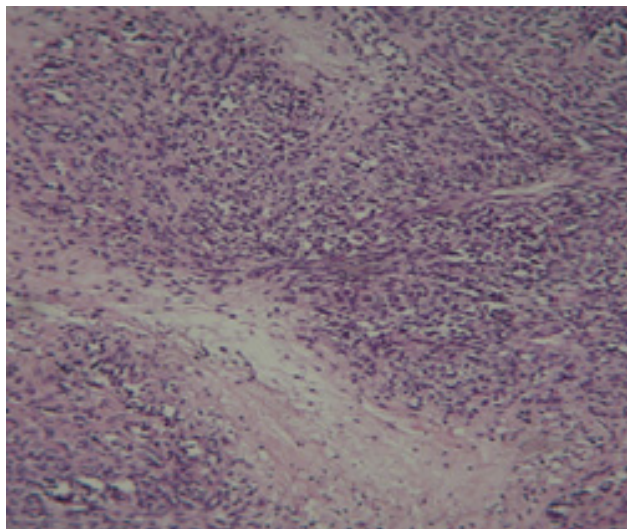


Figure 1. Histology of myofibroma, with biphasic appearance-showing hypercellular ,hypocellular areas. H& E stain, x100

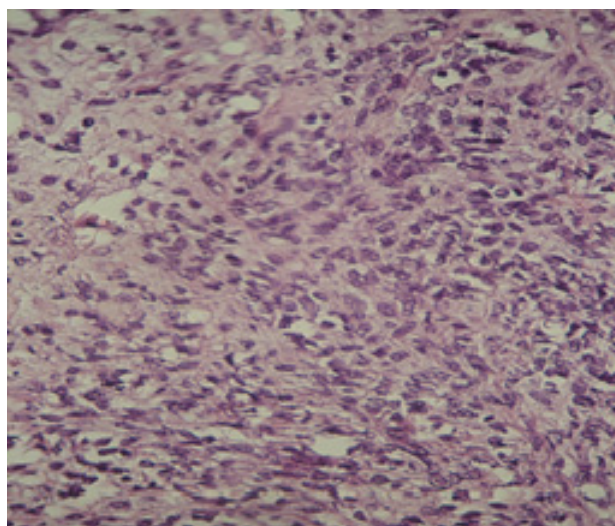


Figure 2. Histology of myofibroma hypercellular area containing round to ovoid cells with hyperchromatic uniform nuclei. H& E stain, x400

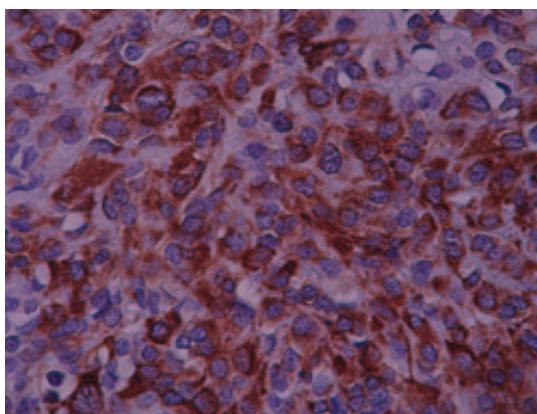


Figure 3. Neoplastic cells of myofibroma reveal positive reaction for SMA , IHC staining , x400

Microscopic examination showed partially encapsulated lobular tumor with biphasic appearance – showing hypercellular , hypocellular areas (Figure 1) - with hemangiopericytoma-like vessels in hypercellular area containing round to ovoid cells with hyperchromatic uniform nuclei (Figure 2). Hypocellular areas show hyalinization and containing spindle cells. Mitotic activity was 3/10 HPF

Immunohistochemical staining showed SMA & (Figure 3) Vimentin positivity (Figure 4) with Desmin/CK/ EMA/ LCA/ Factor VIII & S₁₀₀ negativity (Figure 5) & Ki 67 was 1% positive.

Discussion

Infantile myofibroma of ocular region (including orbit and eyelid) is very rare and there are a few reports of this neoplasm in these areas (4-8).

This case was interesting due to rarity of its report in the eyelid, the congenital pattern of its growth and the necessity that the surgical pathologist thinks about its existence so: when a lesion is seen with a nodular or

multinodular growth pattern which appears biphasic owing to the alteration of light and dark staining areas (light staining areas that are situated haphazardly or sometimes peripherally consist mainly of plump myoid spindle cells with eosinophilic cytoplasm arranged in nodules ,short fascicles or whorls with elongated and tapering or cigar shaped nuclei with atypia and some with extensive hyalinization & dark staining areas usually centrally located and are composed of round or polygonal cells with slightly pleomorphic hyperchromatic nuclei or small spindle cells typically arranged around a distinct hemangio- pericytoma-like vascular pattern. These primitive cells have vesicular nuclei, small amount of acidophilic cytoplasm and indistinct cell margins with slight pleomorphism. Mitotic figure are <8 mitoses/10HPF and some show focal hemorrhage, cystic degeneration or coagulative necrosis and calcification. Peripherally located chronic inflammatory cells including lymphoplasm cells may be seen. Up to 20% show intravascular growth. Early lesions show only dark areas), the possibility of myofibroma/myofibromatosis should be investigated.

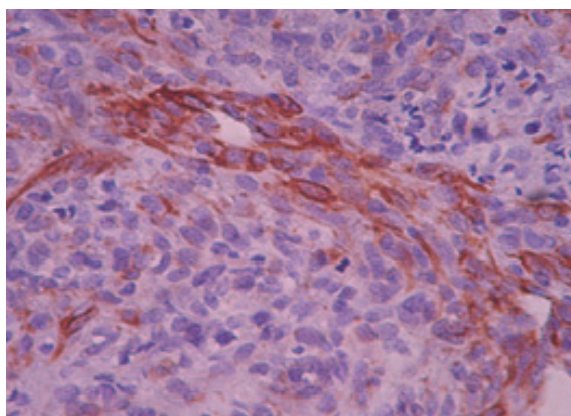


Figure 4. Neoplastic cells of myofibroma reveal positive reaction for vimentin, IHC staining , x400

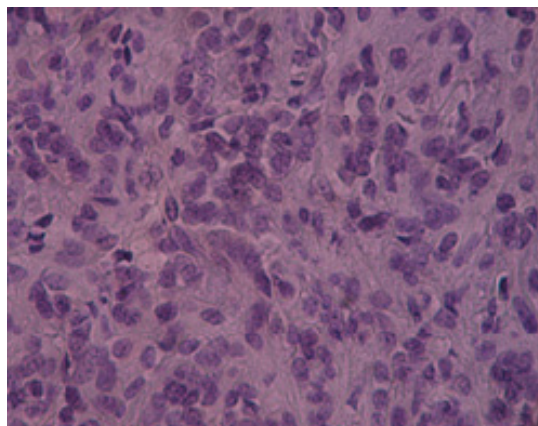


Figure 5. Neoplastic cells of myofibroma reveal negative reaction for Desmin , IHC staining , x400

Immunohistochemical staining shows positivity for Actin weakly and focally but Desmin is usually negative. positivity for Vimentin is seen but CK, EMA, S₁₀₀ are all negative.

Myofibroma and myofibromatosis are expression of benign self-limiting localized or generalized processes that consist to a large degree of cells with the characteristics of myofibroblasts and sometimes of pericytes (9-16).

The differential diagnosis of this lesion depends in part on whether the eosinophilic myofibroblasts or more primitive small cells predominate in a given lesion. Peripheral areas of myofibroma can resemble nodular fasciitis, Fibrous histiocytoma, neurofibroma or infantile fibromatosis. Nodular fasciitis is a rare lesion in newborns and infants and certainly should be considered in the differential diagnosis in adults.

It has a more prominent myxoid matrix and usually contains scattered chronic inflammatory cells and occasional RBCs. The hemangiopericytoma-like pattern characteristic of myofibroma is absent in nodular fasciitis.

The peripheral areas may also resemble neurofibroma but the myofibroblastic cells of myofibroma lack S₁₀₀ protein. Fibrous histiocytoma is composed of a polymorphous proliferation of cells arranged in a more pronounced storiform pattern. Although smooth muscle actin may be found in fibrous histiocytoma, the staining is usually focal and the cells of fibrous histiocytoma usually express factor XIIIa. In infantile fibromatosis the tumor is less well-circumscribed, arise in muscle and show a more uniform spindle cell pattern and it shows neither central necrosis nor a central hemangiopericytoma-like vascular pattern.

Biopsy specimens obtained from the central portion of this lesion may have features that resemble small round cell tumors with hemangiopericytoma-like

vasculature that include peripheral primitive neuroectodermal tumor, mesenchymal chondrosarcoma, malignant hemangiopericytoma and poorly differentiated synovial sarcoma. A battery of immunostains including those for CK, S₁₀₀ & C99 can assist in the differential diagnosis (3).

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