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CASE REPORT

SCHWANNOMA OF UPPER EYELID - A RARE DIFFERENTIAL DIAGNOSIS OF EYELID TUMOR

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ABSTRACT

Schwannoma, also known as neurilemmoma, is a benign peripheral nerve sheath tumor arising from Schwann cells. Schwannoma of ocular area are very rare with most common being in the orbit. The eyelid schwannoma are very rare with only limited number of cases published till date. Here we report a case of 26 year female who presented with a slow growing mass on upper eyelid since 10 years. Histopathological examination of the mass showed features of schwannoma. The lesion was positive for S100 on immunohistochemistry confirming the lesion. Eyelid schwannoma although rare, should be considered in the differential diagnosis of eyelid lesions. Excision of the lesion is the recommended treatment.

INTRODUCTION

Schwannoma is a benign tumor that originates from the Schwann cells that form the neural sheath of the sensory nerves. The most common location for schwannoma is head and neck region followed by limbs and trunk (Enzinger and Weiss, 1995). In ophthalmic region schwannomas are most commonly seen in orbit: other locations include choroid, caruncle, sclera and conjunctiva (Le Marc'hadour *et al.*, 1996). The occurrence of schwannoma in eyelid is extremely rare. Here we report a case of an eyelid schwannoma in a 26 year old female and discuss the various differential diagnoses in these cases.

CASE REPORT

A healthy 26 year female presented with a slow growing painless left upper eyelid mass of 10 years duration. There was associated complaint of heaviness in the upper eyelid and progressive ptosis. There were no associated complaints of itching, redness, photophobia, watering, flashes, and floaters. On examination a firm non tender nodule of size 1x 0.5x 0.50cm was present on left upper eyelid at 2'0 clock position. The mass was not adherent to skin or the underlying tissue. Rest of the ocular examination was normal. The lesion was removed under local anesthesia by a full thickness resection of the lid around the tumor. On macroscopic examination the nodule was well circumscribed, non encapsulated, measuring 1.5 cm in the largest dimension. Microscopic examination tumor showed hypercellular (Antoni A areas) and hypocellular

(Antoni B areas) [Figure 1 a, b]. The Antoni A areas revealed fascicles of spindle shaped cells with elongated bland nuclei, with surrounding collagenized fibers. No mitosis was seen. On immunohistochemistry the tumor cells were strongly positive for S-100 thus confirming a diagnosis of schwannoma (Figure 2).

DISCUSSION

Schwannoma is made up of proliferating Schwann cells of peripheral nerve sheaths. It can occur wherever Schwann cells are present. Most commonly schwannoma occurs as single benign neoplasm. If multiple schwannoma are present it is usually indicative of neurofibromatosis (Le Marc'hadour *et al.*, 1996). Clinically the tumor is a solid, painless and slowly progressive lesion. In our case the patient presented with painless slowly growing mass of 10 years duration involving the left upper eyelid. It was a solitary lesion with no systemic complaints. The main differentials in such cases are chalazion, inclusion cyst and dermoid cyst. Excisional biopsy with histopathological examination is required for the diagnosis. Microscopically, schwannoma is a capsulated lesion and classically shows a mixture of two patterns, the Antoni A (dense cellular pattern) and the Antoni B (edematous disorganized pattern). However, the most important feature for diagnosis is the strong reactivity to S100 protein in immunochemistry and rare mitotic figures (Touzri *et al.*, 2009; De Jong *et al.*, 2010; Shields and Guibor, 1984; Patil *et al.*, 2010; Jakobiec *et al.*, 2016; Baijal *et al.*, 1994; Shields *et al.*, 1994; Siddiqui *et al.*, 2005; López-Tizón *et al.*, 2007; Chung *et al.*, 2007; Lee *et al.*, 2009; Yuichi *et al.*, 2009;

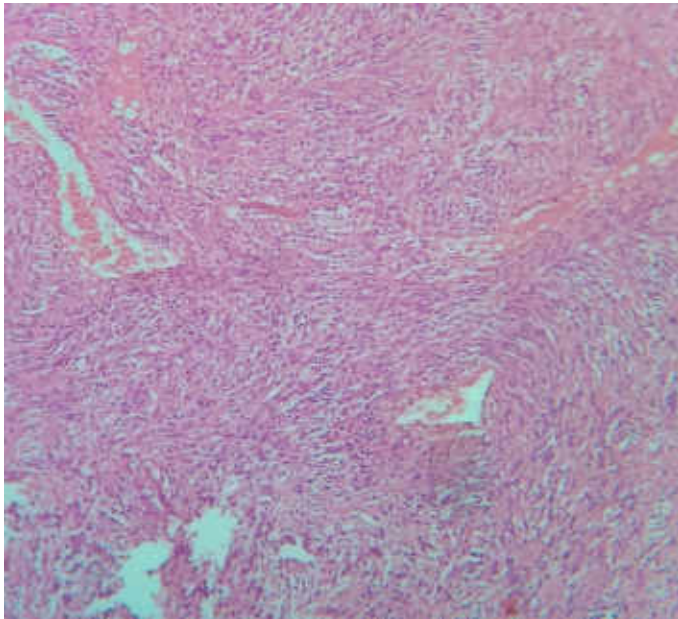


Figure 1a. Histopathological examination showed a tumor composed of hypercellular areas (Antoni A) with focal pallsading

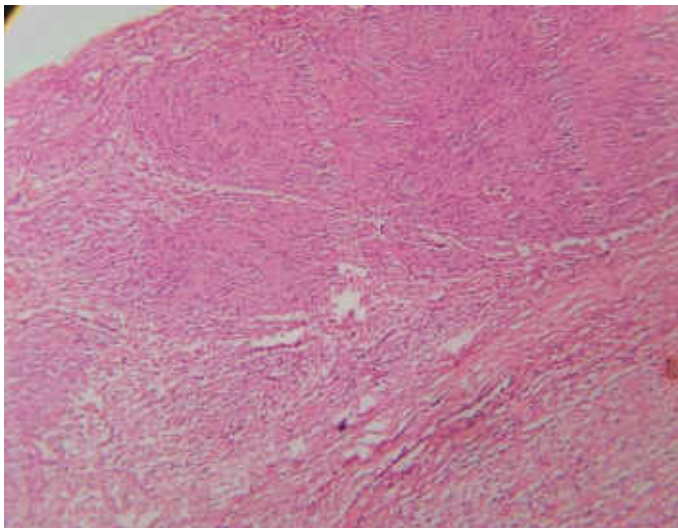


Figure 1b. Tumor also showed focal hypocellular areas (Antoni B)

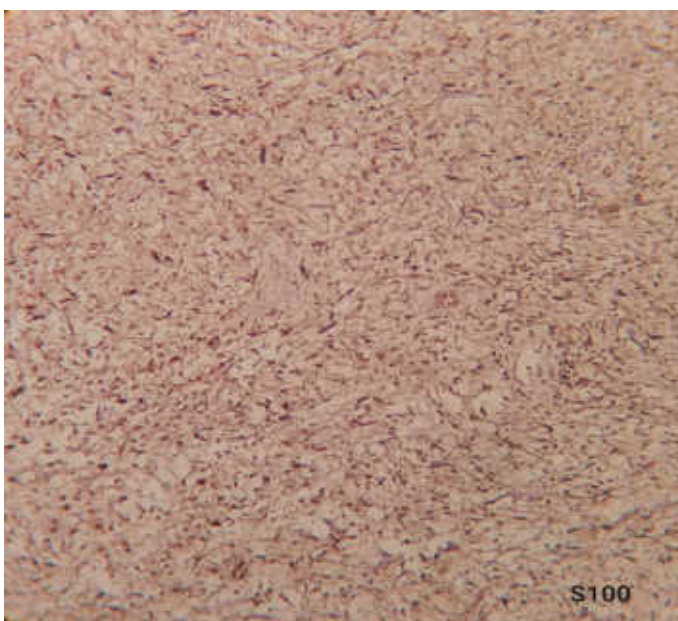


Figure 2. On immunohistochemistry the tumor cell were positive for S 100

Dervis_og̃ ulları *et al.*, 2016; Shields *et al.*, 1994). Poor prognosis has been described in some cases if the cells are fusiform, contain melanin granules, or if epithelioid cells are present (Jakobiec *et al.*, 2016). No features of malignancy were seen in our patient. Nevertheless, malignant transformation has not been reported in eyelid schwannoma (Jakobiec *et al.*, 2016). Schwannomas in the eyelid are very rare with only few cases reported till date (Touzri *et al.*, 2009; De Jong *et al.*, 2010; Shields and Guibor, 1984; Patil *et al.*, 2010; Jakobiec *et al.*, 2016; Baijal *et al.*, 1994; Shields *et al.*, 1994; Siddiqui *et al.*, 2005; López-Tizón *et al.*, 2007; Chung *et al.*, 2007; Lee *et al.*, 2009; Yuichi *et al.*, 2009; Dervis_og̃ ulları *et al.*, 2016; Shields *et al.*, 1994). Schwannoma of the eyelid margin in adults was first reported in 2007 by Lopez-Tizon *et al.* (2007). The second report was in 2012, by Cheng *et al.* (2012). It represents only 0.1–0.7% of the eyelid neoplasm (Touzri *et al.*, 2009; López-Tizón *et al.*, 2007). Eyelid Schwannomas are presumed to originate from supraorbital, supratrochlear and infraorbital nerves. Management of schwannoma of the eyelid is complete excision with clear margins to establish the histopathological diagnosis and to prevent recurrence. Incomplete excision is associated with recurrence and more aggressive behavior. There have been reports of malignant changes in a previously incomplete removal of benign schwannoma (Jakobiec *et al.*, 2016). In conclusion, isolated eyelid schwannoma is extremely rare. It is highly recommended to be considered in the differential diagnosis of eyelid tumor. Accurate histopathological diagnosis and early complete excision of the tumor should be the objective in the management of eyelid schwannomas.

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