

## Schwannoma of the Left Superior Eyelid in a 10-Year-Old Child

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### Abstract

Schwannoma is a rare benign neurogenic tumor whose origins come from Schwann cells located at the myelin sheath of peripheral nerves. It is frequently associated with the orbit, but ocular tissues in general and eyelids in particular are very rarely affected. There are very few reports that can be found in the Literature describing eyelid schwannomas and we have found only two articles describing it affecting children. To our knowledge, this is the first case report about eyelid schwannoma in Brazil - and it involves a child.

**Keywords:** *Neurilemmoma/Pathology; Neurilemmoma/Surgery; Eyelid Neoplasms/Pathology; Eyelid Neoplasms/Surgery; Child; Case Reports*

### Introduction

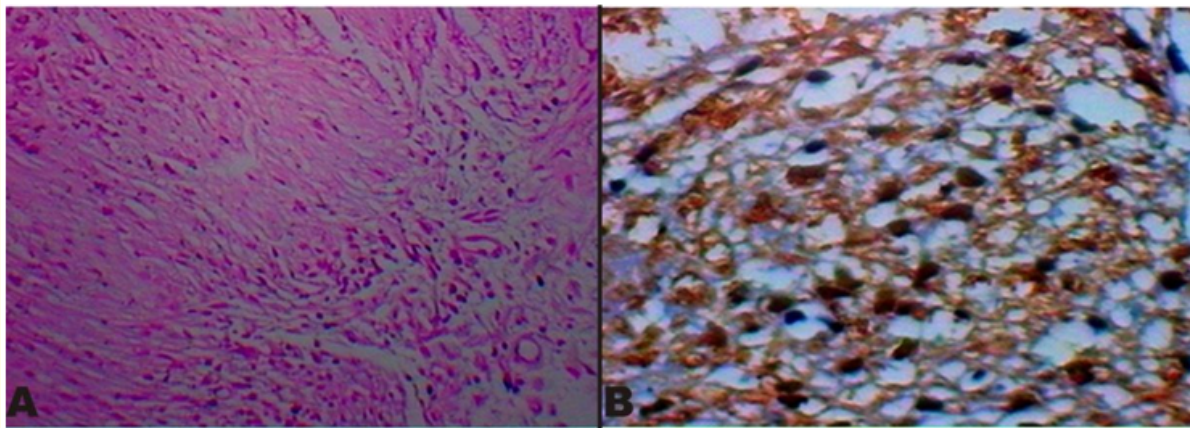
Schwannoma or Neurilemmoma is a rare benign neurogenic tumor, originating from Schwann cells in the myelin sheath of peripheral nerves [1-11]. They are encapsulated tumors of slow growth, whose location in the ocular tissues is not common, the orbit being the most frequently affected site (1 to 2% of all orbit tumors) and the involvement of the eyelids, very rare [1-11]. It frequently develops in adulthood, isolated or associated with neurofibromatosis [2,5] and it is known that the presence of multiple schwannomas are an indicative of this entity.

There are very few reports described in the literature on palpebral schwannoma [1-11] and this is, to our knowledge, the first case reported in Brazil.

### Case Report

Female patient, 10 years old, presented with a tumor in the lateral corner of her left upper eyelid, was diagnosed with chalazion and underwent surgical excision. During the surgical procedure, the macroscopic aspects of the tumor were not compatible with clinical di-

agnosis of chalazion, but suggestive of a tumor of hardened and well-defined subcutaneous tissue. The material was excised and sent for histopathological analysis, which macroscopically showed an irregular, white, elastic tissue fragment, measuring 1.1 x 0.7 x 0.2 cm, with a compact, white cut surface. Microscopy showed histological sections consisting of proliferation of spindle cells with oval nuclei and elongated and oval cytoplasm forming bundles, most of them loose, corresponding to Antoni's pattern A for schwannoma, and in small zones, arrangements corresponding to Antoni's pattern B (Figure 1A). It was defined that it was a case of schwannoma (neurilemoma) with a histopathological predominance of Antoni's pattern A. Was also performed immunohistochemical examination of the lesion which demonstrated immunopositivity with the S-100 protein and immunonegativity with others tested (melan-A, smooth muscle actin, GEAP - glial fibrillar acid protein and Ki-67 antigen), with evidence of low cell proliferation (Figure 1B). This results in immunohistochemical exams confirm the diagnosis of schwannoma.



**Figure 1:** A: Histological section shows proliferation of spindle cells with oval nuclei and elongated and oval cytoplasm forming bundles, most of them loose, corresponding to Antoni's A pattern for schwannoma, and in small areas, arrangements corresponding to Antoni's B pattern; B: Positive immunostaining for protein S-100.

The patient evolved well in the postoperative period, with complete healing of the wound. After six months, the patient did not show any sign of recurrence of the lesion. It still remains on track.

### Discussion and Conclusion

Schwannomas or neurilemmomas are ectodermal originated benign tumors of the cranial and peripheral nerves, derived from Schwann cells of the myelin sheath [1-10]. They are encapsulated, with solid and cystic areas, firm consistency and yellow-brown color, of slow growth and asymptomatic [5].

They can be associated with neurofibromatosis, but, when isolated, they are not related to this entity [2,5].

It presents two histological patterns. Antoni's type A, found in the patient in the case in question, presents with spindle cells, arranged in compact form, usually on palisades. Antoni's type B is characterized by cells dispersed in a myxoid matrix [3,5,6].

Schwannomas located in the eyelid region are extremely rare and usually occur in adults [2-7,9,10]. Only two other cases have been reported in children [1,8]. The patient referred here was 10 years old at the time of diagnosis. Among the cases reported in the literature, there does not seem to be a preference for sex in the incidence of schwannoma (6 male patients and the present case is the eighth reported in a female patient) (Table 1).

Author	Year	Country	Sex	Age
Mishra, Sharan [11]	1960	India	Male	18 years
Baijal, <i>et al.</i> [10]	1980	India	Male	19 years
Shields, Guibor [9]	1984	USA	Female	63 years
Butt, Ironside [7]	1994	England	Female	55 years
Shields, <i>et al.</i> [8]	1994	USA	Male	8 years
Siddiqui, <i>et al.</i> [6]	2005	England	Male	53 years
Chung, <i>et al.</i> [4]	2007	Korea	Female	66 years
López-Tizón, <i>et al.</i> [5]	2007	Spain	Female	41 years
			Female	70 years
Kumar, <i>et al.</i> [3]	2008	India	Male	19 years
Touzri, <i>et al.</i> [2]	2009	Tunisia	Male	47 years
			Female	20 years
Onaran, <i>et al.</i> [1]	2009	Turkey	Female	13 years

**Table 1:** Reported cases of palpebral Schwannoma.

The development of schwannomas in the ocular tissues is more frequent in the orbit, constituting 1 to 2% of orbital tumors [1,5,6], being extremely rare in the eyelid [6,9]. Other sites described are conjunctiva, uveal tract and sclera [1,2,5,9]. The nerves commonly responsible for orbital schwannomas are oculomotor, ciliary and supraorbital. Branches of the supraorbital, supratrochlear and infraorbital nerves are the presumed origins of eyelid schwannomas [1].

Clinically, they are manifested as a solid, slow-growing, painless mass and, due to their rarity, they are constantly confused with chalazion [2,4,9] or inclusion cyst [2,5]. Differential diagnosis must be made with chalazion, sweat gland tumor, sebaceous gland carcinoma, hair follicle tumor and other neurogenic and soft tissue tumors [9].

Rarely, schwannoma can become malignant [2]. Computed tomography is an important exam in the location of the tumor during the surgical approach, since, to avoid recurrence or greater risk of malignant transformation, complete excision of the lesion is necessary, which constitutes the curative treatment [4,7-11].

The diagnosis of palpebral schwannoma can be suggested by a clinical and radiological study, in conjunction with macroscopic data found during surgery, but, as it is a rare entity, it is hardly considered in the differential diagnosis of eyelid tumors, being frequently diagnosed only histopathologically [1-11], as well as in the report described here. It is important to consider this hypothesis, especially in cases of recurrent chalazion. The search for similar lesions in other areas and screening for neurofibromatosis is also valid, as this tumor may be associated with this entity [2,5]. In our case, no other lesions were found, discarding neurofibromatosis.

Schwannomas in the ocular region are quite rare, although they correspond to approximately 1 to 2% of orbital tumors [1,5,6]. Its occurrence in the eyelid is extremely rare [6,9]. Although uncommon, schwannoma must be taken into account in the differential diagnosis of any eyelid tumor.

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