

Schwannoma of Lower Lip: An Unexpected Diagnosis - A Rare Case Report

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ABSTRACT

Schwannoma is a benign encapsulated neuronal tumor arising from the perineural Schwann cells. It has predilection for sensory nerves. Only about 1% is located intraorally. When compared to other sites in intraoral region like tongue, palate, floor of mouth and buccal mucosa, schwannoma of lip is very rare. Only a very few cases have been reported in literature. Here, we present a very rare case of schwannoma lower lip in a 18 year young adolescent boy. He came with the history of swelling in the lower lip since 5 years. The swelling was painless and gradually increased in size. The swelling was excised under local anesthesia and sent for histopathological examination. Gross examination revealed a 1x1cm encapsulated grey white homogeneous mass. Histopathological examination revealed characteristic Antoni 'A' and Antoni 'B' patterns. The tumor was positive for immunohistochemistry S-100.

Key words: Schwannoma, Lower lip, Antoni 'A' area, Antoni 'B' area

INTRODUCTION

Schwannoma is a benign encapsulated neuronal tumor arising from the perineural Schwann cells. It grows slowly and pushes the nerve aside. [1] Other names for schwannoma are neurinoma and neurilemmoma. [2] Schwannoma can arise at any age, but they are most common in young and middle aged adults. There is no predilection for gender. [3] Schwannoma is usually asymptomatic; because of displacement and compression of surrounding normal nerve tissue, it leads to pain and paraesthesia. [2]

Schwannoma may arise from cranial, spinal and peripheral nerves. But it has a predilection for sensory nerves, especially eighth cranial nerve. [1] Almost 25 to 45% of schwannomas arise in the head and neck

region. [4] Only about 1% is located intraorally. In the intraoral region, tongue is the common site, followed by palate, floor of mouth, buccal mucosa, lips and jaws. [2,5]

Particularly, schwannoma located in the lower lip is very rare. [6] In the literature also, very few cases of schwannoma of lip have been reported. [7] Here, we present a very rare case of schwannoma of lower lip in a young adolescent boy which was diagnosed by histopathological examination and immunohistochemistry.

CASE REPORT

An 18 year old male patient presented to the general surgery department with history of swelling in the lower lip since 5 years. There was previous history of trauma at that site. The swelling was

painless and gradually increased in size. Clinically, the swelling was solitary and about 2x1cm in size. It was located over the lower lip slightly to the left of midline near vermilion border. The swelling was non tender, firm in consistency and was freely mobile. It was neither fixed to skin or overlying mucosa. There were no cafe au lait spots elsewhere on the body. Haematological parameters, urine examination and all other vital data are normal.

Clinically, a diagnosis of mucous retention cyst was made. The surgery was done under local anaesthesia. The Swelling was excised, fixed in 10% formalin and sent for histopathological examination.

Gross examination showed a encapsulated, grey white, homogenous mass of size 1x1cm. Cut section is also grey white. Histopathological examination showed a capsulated lesion composed of Antoni 'A' and Antoni 'B' areas (Figure1). Antoni 'A' areas are hypercellular composed by spindle shaped cells arranged in fascicles, whorls and nuclear palisading is also seen. At few places, verocay body formation is also noted. The spindle cells have wavy bland nuclei and moderate cytoplasm. Antoni 'B' areas are hypocellular composed of hyalinised fibrocollagenous tissue. Few hyalinised blood vessels are also seen. Immunohistochemistry was positive for S-100.

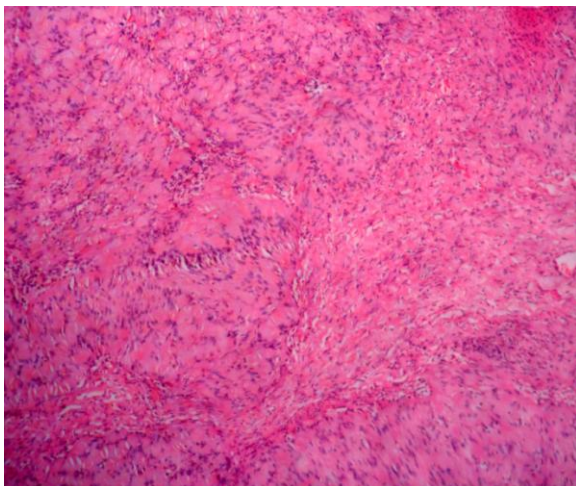


Figure 1- Schwannoma showing Antoni A and Antoni B areas (H & E)(10 X)

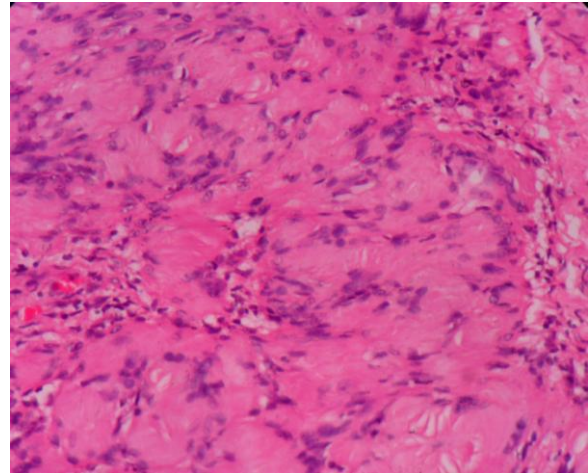


Figure 2 – Antoni A area showing nuclear palisading and verocay bodies (H &E)(40 X)

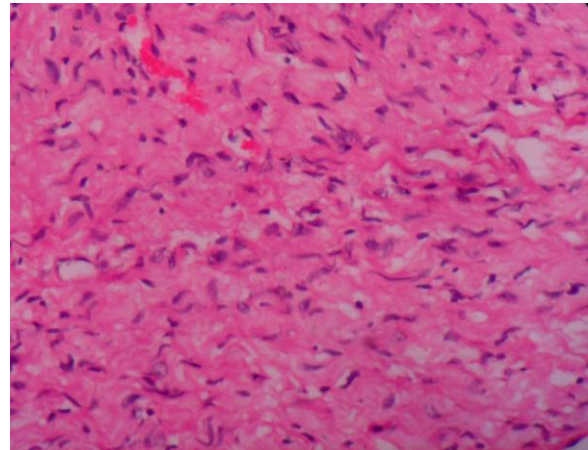


Figure 3 – Antoni B area showing spindle shaped cells in myxomatous stroma(H &E) (40X)

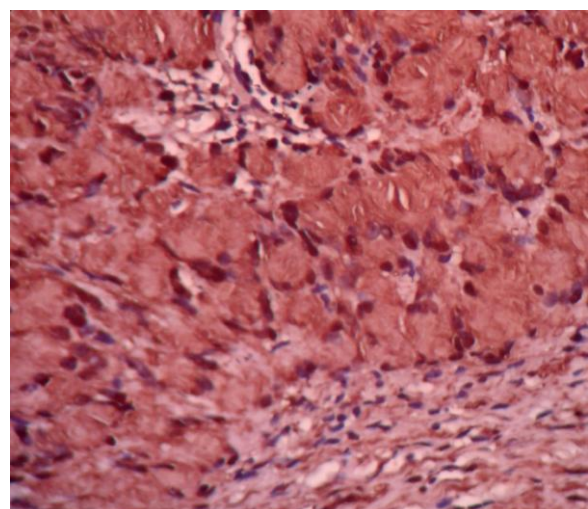


Figure 4 – Schwannoma showing IHC positivity to S-100 (40 X)

DISCUSSION

In 1910, Verocay described a group of neurogenic tumors and named them as neurinomas. In 1935, another term, neurilemmoma was used to name them. In the literature three names are still used, although many names are there. They are neurinoma, neurilemmoma and schwannoma. [1]

Schwannoma of lip is very rare. According to Subha Dhua, [6] only few cases have been reported in the literature. Schwannoma usually presents as gradually enlarging mass. [8] Pain and neurological symptoms are very rare. But if it becomes very big or if it invades submucosal area, it leads to pain and discomfort. Schwannoma can arise as a solitary tumor or it may be associated with von Recklinghausen syndrome. [9]

Grossly, schwannoma is surrounded by a true capsule made up of epineurium. It is variable in size from few millimeters to several centimetres. Cut surface shows smooth appearance. Colour may vary from pink, grey white to yellow. Cystic change, areas of haemorrhage and calcification can be seen. [1]

Microscopically, schwannoma is composed of two types of patterns: Antoni 'A' pattern and Antoni 'B' pattern. Antoni 'A' area is highly cellular, made up of spindle shaped fascicles which form a palisading arrangement around central, eosinophilic, acellular zone known as Verocay bodies (Figure 2). Antoni 'B' area is less cellular, the spindle cells are disorderly arranged in a myxomatous stroma. [1,2] (Figure 3)

Several variants of schwannoma are present which includes Plexiform or multinodular schwannoma, Cellular schwannoma, Ancient schwannoma, Wagner-meissner schwannoma, Nerve sheath myxoma - neurothecoma and Granular cell schwannoma. [1,10,11] But our case is a conventional type of schwannoma which includes both Antoni 'A' and Antoni 'B' patterns.

Differential diagnosis for schwannoma includes Neurofibroma, Myofibroma, Leiomyoma and Palisaded nerve sheath tumor. [7] Sometimes, neurofibroma may present as solitary mass, so differentiation of schwannoma from neurofibroma is important. [12] Based on origin of the cell, schwannoma is derived from the Schwann cell and neurofibroma is derived from the fibroblasts of perineurium. Histologically, schwannoma is composed of Antoni 'A' and Antoni 'B' pattern. While, neurofibroma is composed of mixture of Schwann cells, perineurial cells and endoneurial fibroblasts. [2] The other differential diagnosis we should think while dealing with the lesions of lip are fibroma, inflammatory hyperplasia, lipoma, mucocele, mucous retention cyst and salivary gland tumors. [4,13]

Immunohistochemistry for schwannoma is positive for S-100. It is a neural crest marker which is present in the supporting cells of the nervous system. [1] Our case showed intense and relatively uniform staining for S-100 (Figure 4). Although many differential diagnosis were there for the lip lesions, because of positivity to S-100 IHC marker and the classical characteristic feature of the tumor composed of Antoni 'A' and Antoni 'B' pattern, the diagnosis of schwannoma of lower lip was made for our case.

Schwannoma has good prognosis and the malignant transformation of schwannoma is very rare. [2] The treatment for schwannoma is surgical excision with preservation of neural function. If not possible, end to end anastomosis or interposition nerve grafting has to be done. Recurrence of schwannoma is very rare with complete resection. [1] In our case, the patient had no history of recurrence.

CONCLUSION

We are documenting a very rare case of schwannoma of lower lip in a young adolescent boy while dealing with the lesions of lower lip, even though many differential diagnosis were there, the diagnosis of schwannoma should be kept in

mind, although it is a rare entity. Correlating the histopathological and immunohistochemical findings with the clinical data helped us to make the correct diagnosis of schwannoma lower lip.

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