Case Report

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Surgery Section

Schwannoma of Upper Lip: Report of a Rare Case in a Rare Age Group

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ABSTRACT

Schwannoma is a benign, encapsulated perineural tumour originating from the schwann cells of the neural sheath of peripheral motor and sensory nerves. It may develop at any age but is extremely rare in paediatric age group. The tumour is frequently located on the head and neck region, the tongue being the most common site followed by the palate, floor of mouth, buccal mucosa, lips and jaws. Schwannomas rarely occur in the lip area and it is exceedingly rare in the upper lip. The lesion is usually solitary but can be multiple when associated with neurofibromatosis. The diagnosis is usually confirmed after biopsy and anti-S100 protein immuno-histochemical staining is usually used to identify the tumour. In the present study the patient was a 14-year-old young girl with the schwannoma on the upper lip which is probably the third such case in a paediatric age group being reported and was excised without any recurrence at 2 year after excision.

Keywords: Nerve sheath tumour, Paediatric age group, Upper lip location

CASE REPORT

A 14-year-old young female patient presented to us with a swelling in the upper lip [Table/Fig-1] of about 10 years duration which has started without any predisposing cause. The swelling gradually increased in size and was painless. As the patient was from a very remote part of Meghalaya, she did not consult any doctor earlier. Clinically the swelling was seen in the upper lip, firm in consistency without being reducible and was globular in shape. The swelling was about 6.5 cm in diameter. The surface was variegated on palpation and the swelling was not mobile, but the margins were well-defined. No tenderness could be elicited from the swelling and it was non-transilluminant. Clinically a diagnosis of lipoma was made. Fine Needle Aspiration Cytology (FNAC) showed aggregates of spindle cells with schwann cell processes and eosinophilic cytoplasm. So a diagnosis of schwannoma was made.

The tumour was excised under local anaesthesia. The patient recovered without any complications [Table/Fig-2]. Histopathological examination of the excised tumour came as schwannoma. There were specific hypercellular areas known as Antoni A areas, with frequent nuclear palisading arrangements (Verucay bodies), and less dense reticular areas called Antoni B or reticular-type areas. [Table/Fig-3] showing postoperative image after 5 days of surgery. The patient did not have any recurrence till 2 years of follow-up.

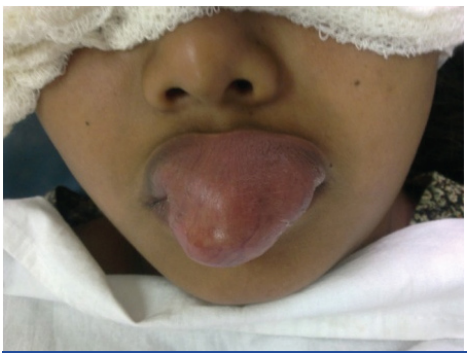
DISCUSSION

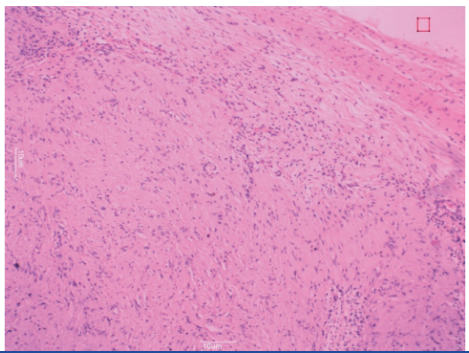
Schwannoma is an uncommon benign tumour arising from schwann cells. It can originate from peripheral, cranial, spinal or autonomic nerve sheaths. The exact aetiology of schwannoma is not known. They are found to arise from a nerve trunk and usually located in the deep layers of soft tissues [1-3]. Schwannomas usually affect young and middle-aged adults in their fourth decade of life [1,4] and rare in the paediatric age group. There is no predilection for gender and may be found equally in both the genders [2,5].

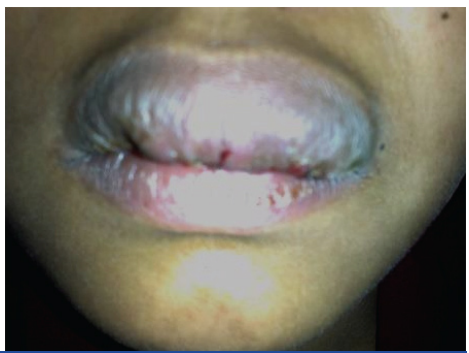
About 25 to 45% of all schwannomas are located in the head and neck region [1]. It is extremely rare to find schwannoma in the upper lip and more so in paediatric patients. The first paediatric case of schwannoma in upper lip was reported by Hashiba et al., in a 12-year old girl [6].

In the review of 26,792 histopathological records from the year 1946 to 2012; Curra et al., found 1195 biopsies of lip lesions [7]. Out of the 1195 biopsies of lip lesions, only four (0.33%) cases of schwannoma on the upper lip were found. The mean age their patients were 45 years and none of their reported cases were found in the paediatric age group [7].

A schwannoma is usually encapsulated and shows proliferation of spindle-shaped cells which are arranged in two patterns: hypercellular







[Table/Fig-1]: Showing schwannoma of upper lip. **[Table/Fig-2]:** Showing histopathological features of Schwannoma (H and E, 20x), hypercellular areas known as Antoni A areas, with nuclear palisading arrangements (Verucay bodies). **[Table/Fig-3]:** Showing postoperative healing after 5 days.

areas (Antoni type A) and hypocellular areas (Antoni type B). The spindle shaped cells may show a palisaded appearance usually arranged around eosinophilic areas, thus forming the so-called Verocay bodies observed in the hypercellular Antoni type A pattern. The differential diagnosis of other spindle shaped cells lesions usually include neurofibroma, palisaded nerve sheath tumour, myofibroma, leiomyoma, etc., and hence the diagnosis may be confirmed by intense immunohistochemical labeling for S-100 protein [1-3].

The various differential diagnoses which should be borne in mind while dealing with lesions in the upper lip include fibroma, inflammatory hyperplasia, lipoma, mucocele, mucous retention cyst and salivary gland tumours [1,2].

Treatment of schwannomas is surgical excision and there is no need to include a margin of safety. Recurrence is rare with complete enucleation of the tumour and the prognosis is usually favourable [1].

The case that is being reported here is the third such case of upper lip schwannoma in a paediatric age group; the second such case was reported by Tuba Bayindir et al., in a 15 old boy in the year 2013 [8]. Abdulkadir Özgür et al., have also reported a schwannoma in the upper lip in the year 2015, but the age of their patient was 19 years which may not qualify in the paediatric age group [9].

CONCLUSION

Schwannoma should be considered as a differential diagnosis while dealing with swellings in upper lip in adults and may rarely present in paediatric patients. Even though rare, a systemic work-up for an accurate diagnosis and simple surgical excision usually results in good clinical outcomes in this subset of patients.

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