

Isolated Conspicuous Mega Sized Schwannoma of the Soft Palate in a Child – A Rare Case Report

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ABSTRACT

Schwannoma are commonly seen in the head and neck region, of which 4% present in sinonasal area and only 1% are seen in the oral cavity. Of the intraoral presentation, schwannoma are rarely seen arising from the palate. Literature search revealed 17 cases of palatal schwannoma and we are reporting the 18th case of palatal schwannoma. This being a very rare case of Schwannoma arising from the soft palate in a 12-years-old boy as a painless, slow growing, firm mass for 2 years, which was surgically enucleated by trans-oral approach with no recurrence and good speech results.

KEYWORDS: Schwannoma, Head and Neck, Oral Cavity, Palatal Tumor, Hyper-nasality, Neurolemmoma, Excision.

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INTRODUCTION

Schwannoma or neurolemmoma or neurinoma was first reported in the 20th century. Virchow first identified schwannoma in 1908¹. These tumors are uncommon, have a benign nature and arise from Schwann cells. Schwann cells in the nerve sheaths of any nerve trunk, cranial or peripheral, may give origin to a schwannoma. These cells wrap extra cranial nerve fibers and form an insulation (myelin sheath) which improves nerve conductance. These tumors are usually solitary encapsulated and smooth surfaced masses which grow very slowly². These tumors have no gender predilection, however some authors report predominance of tumor in females³. Schwannoma commonly occur in 25 to 55 year of age.

Occurrence in children is rare⁴.

Though usually asymptomatic, symptoms when occur depend on the tumor size and location. Schwannoma are commonly seen in head and neck, out of which 4% are seen in sino-nasal tract and only 1% are intraoral⁵. Palate is a very rare site and so far only 17 cases have been reported in literature⁶⁻⁸.

We report a rare case of a large schwannoma of the soft palate in a 12-year-boy with hyper-nasality which made him report to us. Literature review did not reveal soft palatal schwannoma in a child with this presentation.

CASE REPORT

A 12-year-boy presented to the otolaryngology outpatient with complaint of change in voice for 6 months. Further enquiry revealed a slowly growing, painless swelling on the soft palate for 2 years. Medical history was insignificant. Oral examination revealed a 1.5 cm diameter, firm, irregular mass on the soft palate (Fig-1) along with reduced movement of the soft palate and hyper-nasality. No lymphadenopathy was noted. A provisional clinical diagnosis of pyogenic granuloma was made. Radiological examination including plain x-ray paranasal sinuses did not reveal any remarkable findings. The mass was enucleated along with safety margin, under general anesthesia (Fig-2) with primary closure of the surgical wound.

Following excision biopsy the mass was sent for histopathological examination. Gross examination revealed circumscribed greyish white nodular mass, measuring 2 x 1.5 x 1cm with underlying fibrous tissue (Fig-3). Microscopic examination revealed characteristic features of schwannoma along with fibromuscular tissue and sub mucous glands partly lined by stratified squamous epithelium. There was proliferation of spindle shaped cells with Verocay bodies

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(Antoni-A cellular areas surrounded by nuclear palisades) as well as less cellular Antoni-B areas. There was no evidence of malignancy.

Post-operative course was uneventful. Post-operative follow up revealed reduced nasality due to surgical intervention and good wound healing. Follow up was maintained for 6 months with no recurrence and significant improvement in voice production.



Fig-1: Preoperative Clinical photograph showing an irregular mass measuring 1.5 cm diameter on the soft palate adjacent to the hard palate.



Fig-2: Immediate Post-operative clinical photograph showing surgical wound, following removal of the tumor.



Fig-3: Gross photograph showing enucleated 2 x 1.5 x 1 cm mass which had an irregular firm surface

DISCUSSION

Schwannoma is a benign lesion which develop by proliferation of Schwann cells. Exact etiology is unknown however some factors are incriminated including spontaneous growth, chronic irritation, trauma and exposure to radiation^{9,10}. Of the Head and Neck schwannomas, 4% occur in the sino-nasal tract¹¹ and 1% are intraoral⁵, with the tongue being the commonest site^{1, 3, 5, 12}, followed by palate, bucal mucosa, lip and gingiva. Palate is a very rare site⁶. In a review study, Biswas et al noted only one case of palatal schwannoma out of 31 cases of extracranial schwannomas⁷. Moradzadeh Khiavi et al, in 2014 reported a case of schwannoma of midline hard palate in a 21 year old patient⁸. Schwannoma of the soft palate are even rarer than those of the hard palate. Venkatachala S et al reported a soft palate schwannoma in a 43 year old patient¹³, Hameed et al reported a palatal schwannoma extending from the junction of hard and soft palate to base of uvula in a 18 year old patient⁶. Rahpeyma and colleagues also reported a soft palate schwannoma in a 12 year-old-girl with ulcerated tumor surface⁴, and Chawla O et al reported it in a 9 year old child¹⁴. Though cases of schwannoma of head and neck have been reported in Pakistan.¹⁵⁻¹⁷ However reports of oral schwannoma are rare in literature¹², with no case of soft palate schwannoma reported from this part of the world. Also literature search did not reveal soft palatal schwannoma presenting with hyper-nasality in international literature as well. The present case is very rare and unique, being a child of 12 with soft palate schwannoma who presented with hyper-nasality. This case has similarity to a case reported by Rahpeyma et al, which was of similar age and site of tumor but with no functional symptoms⁴. Presentation of these tumors depend on size and location. Schwannomas usually grow slowly and painlessly making detection of small schwannomas difficult. They usually present when the tumor reaches a size that results in cosmetic or functional issues. Paresthesia's have also been reported¹⁸. In

the present reported case, the tumor has a very unusual presentation since it grew slowly for 2 year without any complaints, although its location could have caused functional issues like chewing and swallowing difficulty, and it ultimately presented due to hyper-nasality.

Differential diagnosis of schwannoma of the palate include minor salivary tumors, fibroma and neurofibroma¹⁹. In case of soft palatal tumor differential also includes pyogenic granuloma and lipoma. The present case gave the clinical impression of a pyogenic granuloma however excision biopsy revealed a schwannoma.

Gross examination usually reveals a yellowish, globular, firm to rubbery mass. Histological varieties include plexiform, cellular, epithelioid and ancient and two distinct patterns are seen which include Antoni Type A (Proliferation of fusiform cells) and Antoni Type B cells (few fusiform cells seen in myxoid stroma along with areas of degeneration, edema, necrosis, hemorrhagic tissue and also cystic formations)²⁰. The present case was composed of both Antoni type A and B pattern with no evidence of degeneration.

Complete Surgical Excision is the mainstay of treatment with a very low risk of recurrence. However, approach to be adopted depends on the lesion's site and size. Even in cases in which nerve of origin is identified, excision of the tumor preserving the nerve of origin is all that is required²¹. However identification of the nerve in large lesions may not be possible²¹. Follow up is essential. We were successful in doing a complete resection of tumor without any recurrence and good speech results.

CONCLUSION

Schwannoma of the soft palate is very rare entity especially in children, and should be kept in mind when dealing with swellings on the soft palate.

CONTRIBUTION OF AUTHORS

Saqulain G: Manuscript writing

Mumtaz N: Literature review

Khan MM: Critical review

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