Case Report

An Ancient Schwannoma of Hard Palate

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Nayana V G,[1] Akshatha Shetty,[1] Sai manohar S,[1] Roshni Babu[1]

ABSTRACT

Introduction

Schwannomas are the most common benign nerve sheath tumours arising from Schwann cells. Intraoral schwannomas are rare and most common site and in oral cavity hard palate is a very rare site.

Case Report

Here we present a 16 year old boy who presented with a hard palate swelling of 6 month duration. Cytology was negative and completely excision was done. Histopathology proved as ancient schwannoma. He was followed up for 1 year with no signs of recurrence.

Discussion

Any schwannoma with the histologic degenerative changes and cytologic atypia is known as ancient schwannoma. Treatment of this neoplasm is surgical excision with very low rate of recurrence. Malignant transformation has not been described for the ancient variant of schwannoma.

Keywords

Neurilemmoma; Hard Palate; Recurrence; Ancient Schwannoma

Schwannomas are the most common benign nerve sheath tumours arising from Schwann cells. They are typically solitary, well-encapsulated, and slow-growing. They can occur along various nerves, including motor, sensory, sympathetic, and cranial nerves, except for the optic and olfactory nerves which lack schwann cells.[1]

25-40 % of cases are seen in head and neck region.[2] Intraoral schwannomas are rare and most common site in oral cavity is base of tongues, other areas include buccal mucosa ,lip , hard palate and gingiva.[3]

A rare variety of schwannoma is ancient schwannoma which exhibits calcification, cystic degeneration, haemorrhage, myxoid stroma, pleomorphism and nuclear hyperchromatism. Since it bears resemblance with malignancy, it is easily misdiagnosed as malignancy.[4]

Based on previous systematic review on oral ancient schwannoma, it is revealed that average age of occurrence is in second or third decade . There is a female preponderance with male to female ratio of 1:2.[5]

Primary hard palate schwannoma are rare and among that ancient schwannoma of hard palate is quite rare and very few cases have been reported in the literature till date. Here we present an interesting case of ancient schwannoma of hard palate in a 18 year old boy.

1 - Department of ENT, Yenepoya Medical College Mangalore

Corresponding author:

Dr Nayana V G

email: drnayanavg@yenepoya.edu.in

Case report

An 18-year-old male presented with a painless swelling on the right side of his hard palate for 6 months duration. This case is reported after getting an informed consent from patient for publication. The patient developed a small swelling on his palate which was the size of a peanut, 6 months back, it gradually progressed to the current size.

The patient's medical and family history was non-contributory. He did not give history of any recent episodes of high-grade fever, pain or difficulty while swallowing solids/ liquids or any significant weight loss in the recent past.

On examination there was a solitary swelling of size 4x2x2cm on the right side of the hard palate and was covered with a smooth healthy mucosa. On palpation- the surface of the swelling was smooth, and it was firm in consistency, non-tender, with no local rise of temperature.

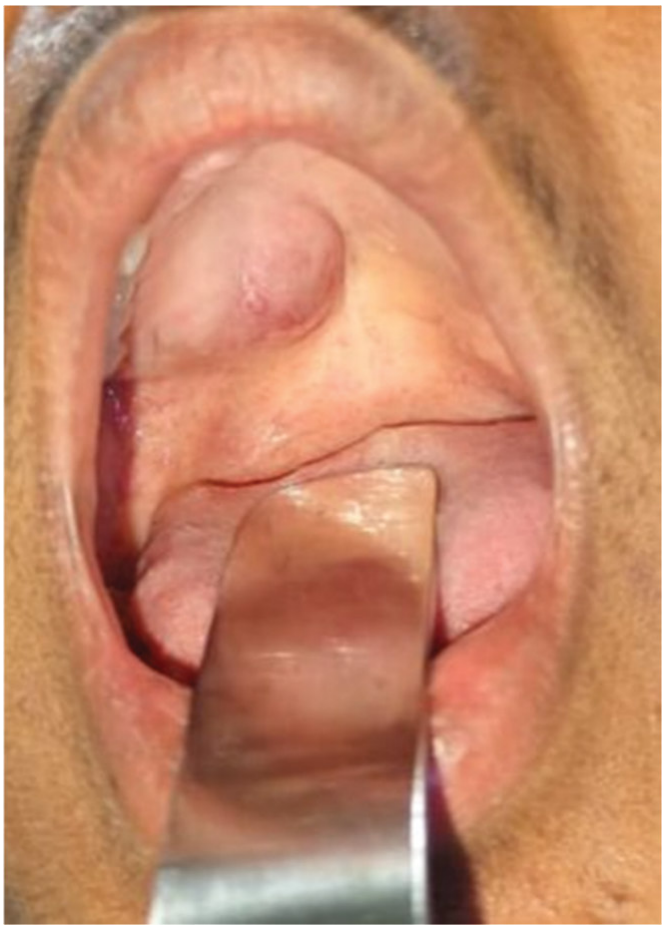

Fig. 1. Pre operative picture showing lesion on the right side of hard palate

Contrast enhanced Commuted Tomography of Nose and Para nasal sinuses showed a well defined subtly enhancing soft tissue attenuating lesion seen arising from the right side of posterior hard palate measuring 2.1x1.5x1.8cm with mild scalloping of the underlying palate and maxilla.

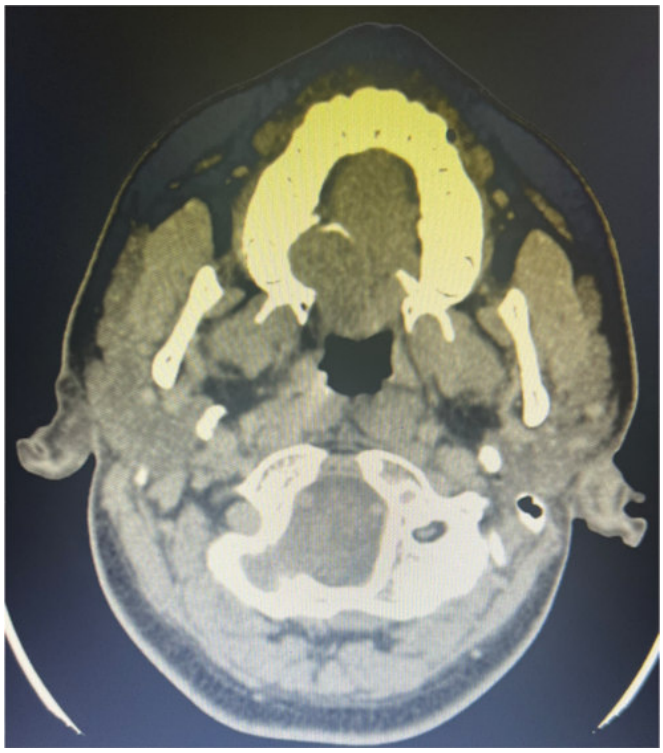

Fig. 2. Showing subtly enhancing lesion on the right side of the palate

Additionally, few subcentemetric level IB lymph nodes were noted with maintained fatty hila. Fine needle-aspiration was performed, it was very painful and no evaluable tumour tissue was obtained.

Based on these data, the most likely diagnosis was considered as palatal tumor originating in minor salivary glands, and complete excision was planned.

The tumor was finally excised via an intraoral approach under general anaesthesia. A linear incision was made anterior to the swelling and posteriorly based mucosal flap was elevated. Tumour was well encapsulated and easily separated from the mucosa. The tumor was separated from the adjacent palatal tissues by careful blunt dissection and complete removal was then accomplished by resection en bloc. The nerve of origin could not be identified.

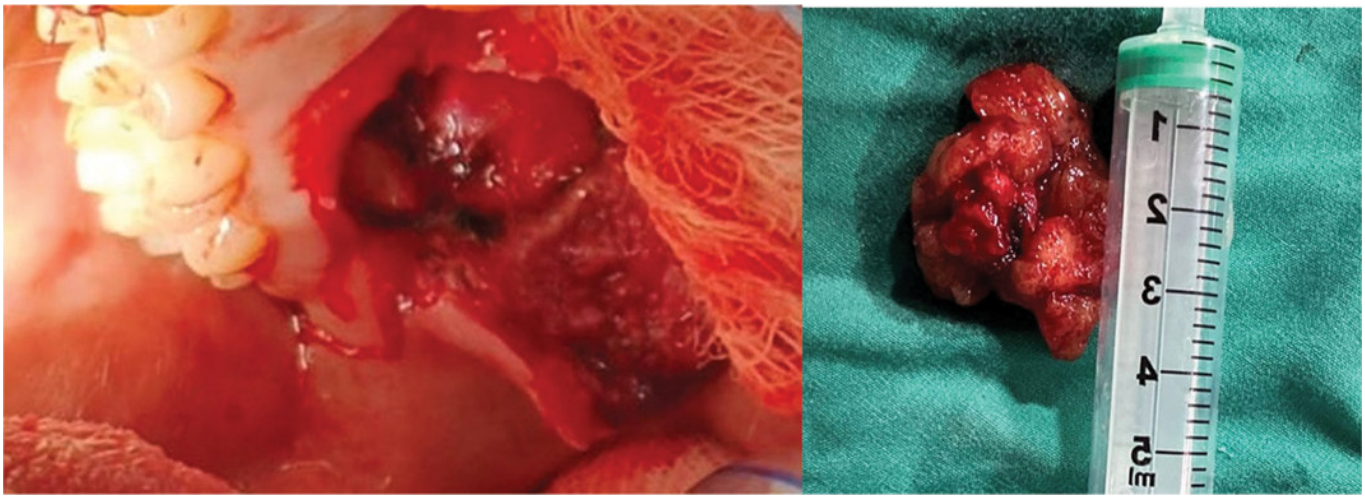

Fig. 3 & 4. Intraoperative picture showing well defined lesion after mucosal elevation and gross specimen

The underlying bone surface was smooth and showed no evidence of compressive resorption. The postoperative period was uneventful.

On gross examination, the resected specimen consisted of a well-circumscribed mass measuring 3.5x2.2x1.2cm. Outer surface appears nodular, cut surface showed a well defined grey white lesion with an area of haemorrhage. Microscopic examination revealed fragments of tissue showing compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas. Cellular areas showed nuclear palisading around fibrillary process. The cells were narrow, elongated and wavy with taped ends interspersed with collagen fibres. Few areas showed cystic degeneration, haemorrhage and thick-walled blood vessels. Histopathology was suggestive of Ancient schwannoma.

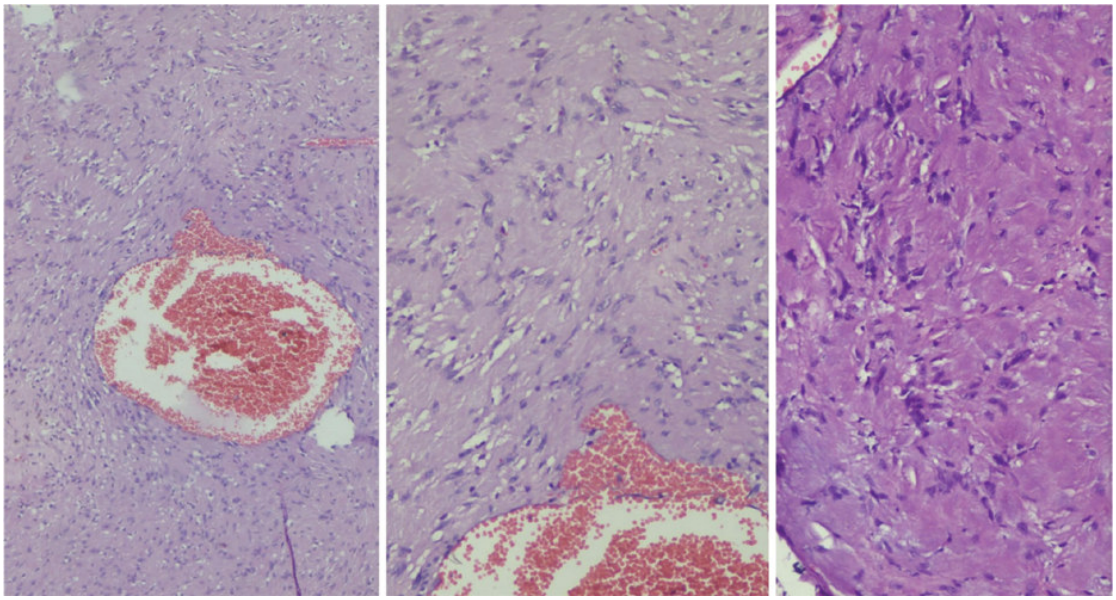

Fig. 5. Histopathology showing Antony A and Antony B cells and degeneration Stain used is eosin and hematoxylin at 10x,20x &20x magnification respectively

Discussion

Schwannoma is also called as neurinoma, neurilemmoma and perineural fibroblastoma.[6] Based on histopathology, five schwannoma variants have been described :common, plexiform, cellular, epithelioid and ancient schwannoma.[7] Ancient schwannoma is one of the rare variant of schwannoma, which has slow growth. The term 'ancient' was proposed to describe a group of neural tumour showing degenerative changes, diffuse hypocellular areas, nuclear hyperchromasia and marked nuclear atypia.[8]

Ancient schwannoma was first described by Eversole and Howell. It was first reported in thorax in 1951 by Ackerman and Taylor.[9] They are rare benign encapsulated tumour of protracted indolent growth , ancient schwannoma denotes extracranial schwannoma, which are solitary and grow to large size.[10]

Intraoral schwannomas are more commonly seen in soft tissues more commonly in tongue. Study conducted by Gallo et al, out of 157 cases reported 45.2% cases involved tongue. Wright and Jackson reported on 146 cases intraoral schwannomas ,among them 52% involved the tongue.[11] Most of the intraoral schwannomas are located in tongue or floor of mouth, hard palate being a rare location.[12]

Atolaiby et al evaluated all neural neoplasm in the oral cavity , which constitutes to 0.2% of all oral specimens. Out of all oral neural specimens, ancient schwannoma accounts for only 0.7% , reflecting the rarity of this pathology.[13]

Most of these patients are asymptomatic and hence they present with long duration and large size. They can experience pain, dysphasia or neurological alteration by the compression of the nerve in case of large sized tumors.

Although according to literature it is long standing and present with large size tumour, our patient presented with only 6 month history of swelling in the hard palate with no other symptoms. Due to short span of presentation with large swelling, differential diagnosis we considered were palatal tumour arising form minor salivary glands and different connective tissue tumours.

Histopathologically schwannoma appears to be made up of two distinct areas: dense areas (Antoni A): These areas contain uniform, elongated cells (spindle cells) with pink cytoplasm (eosinophilic) and oval nuclei. Loose areas (Antoni B): These areas are less populated with cells and have a jelly-like appearance (myxoid). Ancient schwannomas are characterized by degenerative changes, including increased deposition of matrix, perivascular hyalinization, ectatic vessels with thrombus, cystic degeneration, and cellular atypia with paucity of mitosis. These changes are attributed to the long duration of the schwannoma. This altered structure can make diagnosis difficult under a microscope because the usual features of benign nerve sheath tumours may be missing or unclear.[12, 14]

Due to nuclear atypia and hyperchromasia it is often misdiagnosed as malignancy. Dahl in 1977 reported that , out of 11 cases ancient schwannoma, 6 were mis-diagnosed as sarcoma .It has also been misdiagnosed with myxoid neurofibroma and nerve sheath myxoma.[15]

Cytology study by fine needle aspiration of the swelling was done but it did not yield any valid material similar to the first reported case of ancient schwannoma as these lesions have diffuse hypocellularity they are difficult to diagnose by fine needle aspiration cytology.[14, 16]

Contrast enhanced CT scan and MRI are often helpful for diagnosis and also for evaluating the site and extent of the lesion , but exact origin of schwannoma is difficult to know preoperatively. CECT nose and pns was done to our case which revealed subtly enhancing soft tissue arising from posterior part of hard palate and HPE correlation was suggested. MRI was not done in our case. Excisional biopsy was done. According to literature also complete excision of lesion and preservation of the nerve of origin is the expected treatment.[12] Treatment of this neoplasm is surgical excision with very low rate of recurrence.[17, 18]

Malignant transformation has not been described for the ancient variant of schwannoma so far. Although recurrence rate is low, patients have to be kept on regular follow up. This case highlights the importance of recognizing the histologic degenerative changes and cytologic atypia that can be seen in this tumor. By being

familiar with these features, oral pathologists can avoid misdiagnosis and unnecessary treatment.

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