SCHWANNOMA OF THE HARD PALATE: A RARE EXTRACRANIAL SITE

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ABSTRACT
Schwannoma or neurilemmomas are benign tumours of ectodermal origin derived from Schwann cells lining the nerve sheath. They can arise from any cranial, peripheral, or autonomic nerve having Schwann cells. Approximately 25% to 48% of all schwannomas occur in the head and neck. The most common site to be affected is the eighth cranial nerve. Only 1% of the schwannomas are seen intraorally of which tongue is the most common site. We present a case of schwannoma of the hard palate, a rare site to be affected.

Key Words: Schwannoma, Neurilemmoma, Hard Palate, Extracranial

INTRODUCTION
The various benign neural tumours that can occur in the hard palate include traumatic neuroma, schwannoma, neurofibroma and mucosal neuroma representing MEN type syndrome 3. Traumatic neuroma is a non-neoplastic hyperplastic reaction of axons and Schwann cells. Other benign lesions that can occur in the oral cavity include minor salivary gland tumours or non-neoplastic lesions such as granular cell tumour, mucoceles and polyps (Murthy et al., 2009). Malignancies very rarely occur in the hard palate except in individuals, with a habit of reverse smoking leading to carcinomas.

CASES
A 14 years-old boy presented with a 6 day history of swelling in the roof of mouth. He had slight slurred speech and discomfort while swallowing. The swelling used to bleed occasionally while brushing and on having food. There were no complaints of pain, irritation, sudden increase in size or any other mass in the neck or other parts of the body. His family history was not significant. There was no history of smoking or chewing tobacco or betel nut.

On examination, a 1.5cm diameter globular pink smooth, firm in consistency and well encapsulated mass in the posterior surface of the hard palate just anterior to the junction of hard and soft palate, over the left greater palatine foramen region was present. There was a tiny area of ulceration. It had a soft consistency. There was no evidence of trismus or involvement of nasal floor and septum.

No cervical lymphadenopathy was observed. Neurologically no cranial deficit was elicited. Routine haematological and biochemical investigations were within normal limits. A clinical diagnosis of haemangioma was thought of.

A wide local excision was carried out and subjected to histopathological examination. The sections stained with Haematoxylin and Eosin showed an outer ulcerated epithelium. Beneath there were spindle shaped cells with elongated nuclei arranged in a palisading manner forming Verocay bodies at places (Figure 1). The other areas were hypocellular with few elongated cells and loose oedematous stroma (Figure 2). Few blood vessels were thickened and hyalinised. The microscopic features were consistent with the diagnosis of Schwannoma. Immunohistochemistry for S-100 was positive. The post-operative period was uneventful.
DISCUSSION

Schwannomas or neurilemmomas are benign, slow growing, solitary, well-encapsulated tumours, originating from the Schwann cells of the nerve sheath. They usually present between the second and fourth decade of life but cases during the first year of life are reported. There is no gender predilection. About 25% to 48% of schwannomas originate in the head and neck region. Intraoral schwannomas constitute only 1%. Tongue is the commonest site for intraoral schwannomas. The other intraoral sites to be affected include hard palate, buccal mucosa, lips and the gingiva (Murthy et al., 2009; Roy et al., 2002; Lollar et al., 2010) of these, 8–10% schwannomas are reported to transform to malignancy.

The signs and symptoms of these tumours depend on the location. They usually are asymptomatic. However, if they occur on the base of the tongue or floor of mouth, there may be dyspnoea or dysphagia (Budde et al., 2001; Pfeifle et al., 2001). Otherwise, the presenting feature of an intraoral schwannoma is only a tumour mass. Despite being of neural origin, they usually are painless causing pressure on adjacent nerves rather than nerve of origin and may present with paraesthesia of the region of trigeminal sensory distribution (Murthy et al., 2009; Roy et al., 2002; Isildak et al., 2010). Rarely may they ulcerate.

Figure 1: Hypercellular Antoni A areas, composed of palisading spindle cells [Verocay bodies] H&E X 400
Case Report

Figure 2: Hypocellular Antoni B areas, with loose oedematous stroma. H&E X 400

Histopathologically, the tumour tissue is well encapsulated and consists of Antoni A and Antoni B type cells. Type A tissue shows densely packed, elongated spindle cells, with a regular arrangement forming a palisading pattern. They are associated with delicate reticulin fibres and spindle-shaped nuclei. Between the rows there are fine cytoplasmic fibrils with acellular, eosinophilic masses called Verocay bodies. The palisading nuclei are arranged in rows, surrounding a central acellular eosinophilic zone known as Verocay body.

Antoni B tissue consists of spindle cells haphazardly distributed in a light fibrillar matrix. This type of tissue is less cellular and is formed by irregularly arranged masses of elongated cells and fibres, with areas of cystic degeneration and oedema (Murthy et al., 2009; Dhupar et al., 2011). Immunohistochemistry study of schwannomas show an intense and uniform staining for S-100 protein, which is a marker for Schwann cells. Besides, there may be positivity for Vimentin, Lev 7 antigen and Glial fibrillary acidic proteins. Reports of malignant schwannoma of the oral cavity are extremely rare (Roy et al., 2002; Dhupar et al., 2011).

Treatment mainly is surgical excision. A wide local excision with clear margins gives a good prognosis.

Conclusion
It is imperative that schwannoma should be considered in all cases with intraoral swellings and confirmed histopathologically for better management.
REFERENCES


