

CASE REPORT

A Large Extra-Osseous Solitary Neurofibroma of the Hard Palate

Amer Sabih Hydri¹, Syed Shaukat Hussain², Iqbal Hussain Udaipurwala³, Fatima Siddiqui⁴

ABSTRACT:

Solitary and isolated extra osseous neurofibroma of the hard palate is a rare clinical entity and literature search showed that only three cases have been published so far. The diagnosis is often arrived on immuno-histochemistry after surgical excision of the tumour. We are reporting a case report of a large and solitary extra-osseous neurofibroma of the hard palate, which was not associated with generalized neurofibromatosis in a 30 years old lady. She presented with a painless and gradually progressive swelling on the hard palate which was interfering with the swallowing and speech. On examination there was a large pedunculated mass arising from the hard palate near its alveolar margin. Surgical excision of the swelling was done and histopathology showed the diagnosis of neurofibroma. There is no recurrence after a follow-up of 18 months till so far.

Keywords: Neurofibroma, Hard palate, Oral tumours, Von Recklinghausen's disease.

INTRODUCTION:

Neurofibroma is defined as a well-demarcated intraneural or diffusely infiltrative extraneural tumour arising from Schwann cells and perineurial fibroblasts. Neurofibroma of the oral cavity often presents as a sub-mucosal, non-tender, discrete mass. The head and neck region is commonly involved by neurofibroma because of the rich innervation of this area. The lip, tongue, buccal mucosa, and vestibular area are the common extra-osseous sites. Hard and soft palates are rarely affected by these lesions.

Neurofibroma may occur as solitary lesion or as part of a generalized syndrome of neurofibromatosis (von Recklinghausen's disease) or very rarely as multiple neurofibroma without any association with neurofibromatosis syndrome. Generalized neurofibromatosis is divided into two clinical forms NF1 (peripheral) and NF2 (central). Both of these types are clinically and

genetically different. The predominant type is NF1 which occurs in almost 90% of the cases. It is imperative to ensure that there is no confusion between the diagnosis of isolated neurofibroma and those associated with NF1 since both have divergent clinical behavior, treatment, and prognosis.

An extensive review of the internet and medical literature revealed only three published reports of extraosseous solitary neurofibroma of the hard palate not associated with type 1 neurofibromatosis. We are reporting this case of a large and solitary extra-osseous neurofibroma of the hard palate.

CASE REPORT:

A 30 year-old-female reported to the ENT OPD with the history of a lump on her palate. She had noticed this lump about a year back and it had grown steadily to its present size (Figure 1a). The lump was painless but had now started interfering with her speech and deglutition, especially solid food. On examination, the patient had full, healthy dentition with a solitary, smooth, firm, non-tender, elongated mass of about 4.5 x 2.5 x 2.5cms on the hard palate and extending on the soft palate. It appeared sessile at first but detailed examination revealed a small pedicle, which was arising from the left mid hard palate near the alveolar border. This mass was extending across the midline to the right side as well (Figure 1a). Apart from this growth, the intra-oral examination was unremarkable. Both the nasal cavities were normal on anterior rhinoscopy and x-ray PNS (Water's view and Sub mento-vertical view) were also normal. There was no extension of the mass in the nose or sinuses.

Excisional biopsy was planned for obtaining a histological diagnosis. After counseling the patient, informed consent was obtained and the patient was operated under general anesthesia. After inserting a mouth gag, the pedicle was identified as being attached to the greater palatine nerve (Figure 1b). The pedicle was meticulously removed by electrocautery, while preserving the nerve and the unblemished growth removed completely (Figure 2a). There was no bleeding after excision of the mass (Figure 2b).

Post-operatively there was immediate improvement in the patient's speech and deglutition and she was discharged the following day. Histopathology report

✉ Dr. Amer Sabih Hydri

Assistant Professor
Department of ENT
PNS SHIFA Hospital
Bahria University Medical & Dental College
Karachi

Email: draamerhydri@gmail.com

✉ Dr. Syed Shaukat Hussain

Assistant Professor
Department of ENT
PNS SHIFA Hospital
Bahria University Medical & Dental College
Karachi

✉ Dr. Iqbal Hussain Udaipurwala

Professor & Head
Department of ENT
Bahria University Medical & Dental College
Karachi

✉ Dr. Fatima Siddiqui

Registrar
Department of ENT
Bahria University Medical & Dental College
Karachi

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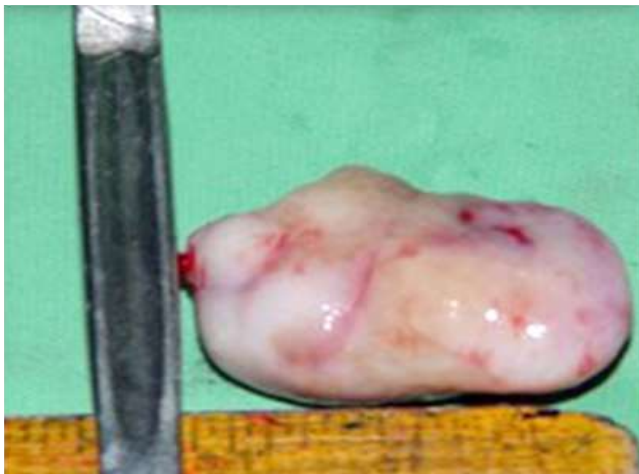
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revealed the mass to be a neurofibroma. On her follow-up visit the patient was re-evaluated for type I neurofibromatosis in mind but no additional evidence

Figure: 1a
Swelling seen on the palate



Figure: 2a
Complete mass after excision



could be elicited. At the 18 months follow-up the patient is asymptomatic and free from any recurrence.

Figure: 1b
After inserting mouth gag, pedicle of the swelling was identified



Figure: 2b
Surgical site after excision of the mass



DISCUSSION:

Palatal tumours of the neural origin are very uncommon. Clinically, oral neurofibroma usually appears as pedunculated or sessile mass, with a very slow growth rate. Malignant transformation of solitary neurofibroma takes place very rarely. Recurrence is also rare although some authors suggest higher rate of recurrence in head and neck region in case of solitary neurofibroma.

Of the three cases reported earlier,^{6,7,8} all were small and sessile growths. Our case is the first where the solitary neurofibroma was pedunculated and was also very large in size (4.5 x 2.5 x 2.5cms). Due to the slow growth of neurofibroma patients are usually asymptomatic, but depending on the location (e.g., tongue, palate), it may be traumatized and give rise to symptoms.

Regarding the soft tissue manifestation of neurofibromatosis, palate is affected in 8% of the patients, while the solitary extra osseous lesions are rare. Despite the

large size of the palatal mass our patient remained asymptomatic for almost a year and only reported after it altered her speech. Although Sheejith⁹ and Bongiorno¹⁰ their separate studies claim no predilection to gender or race yet Cherrick¹¹ reported a preference for females. Our patient was incidentally a female patient. Treatment for solitary neurofibroma is surgical excision, while preserving the nerve from which it originates and recurrence is rare as is evident from the follow up of all reported cases including our case. Confirmation of diagnosis is by histopathology and immuno-histochemistry.¹² Neurofibroma is immuno-positive for S-100 protein in 85 to 100% of the cases.

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