

## Solitary Buccal Space Neurofibroma: A Case Report

Md. Ashraful Islam<sup>1\*</sup>, Tareq Mohammad<sup>2</sup>, Towsif Bin Mamoon<sup>2</sup>, Md. Sirajul Islam<sup>3</sup>, Md. Aminul Islam<sup>4</sup>, Farid Uddin Milki<sup>5</sup>, ASM Lutfur Rahman<sup>6</sup>, Nazmul Hossain Chowdhury<sup>2</sup> and Kamal Ahmed<sup>7</sup>

<sup>1</sup>MBBS, FCPS, FICS, FRCS (Glasgow), Professor and Head, Otolaryngology-Head & Neck Surgery, Bangladesh Medical College Hospital, Bangladesh

<sup>2</sup>Assistant Registrar, ENT, Bangladesh Medical College Hospital, Bangladesh

<sup>3</sup>Junior Consultant ENT, Bangladesh Medical College Hospital, Bangladesh

<sup>4</sup>Registrar ENT, Bangladesh Medical College Hospital, Bangladesh

<sup>5</sup>Consultant ENT, Bangladesh Medical College Hospital, Bangladesh

<sup>6</sup>Resident Surgeon, Bangladesh Medical College Hospital, Bangladesh

<sup>7</sup>Internee, Faridpur Medical College Hospital, Bangladesh

\*Corresponding author: Islam MA, MBBS, FCPS, FICS, FRCS (Glasgow), Professor and Head, Otolaryngology-Head & Neck Surgery, Bangladesh Medical College Hospital, Bangladesh, Tel: +8801711527954; E-mail: [ashrafis123@yahoo.com](mailto:ashrafis123@yahoo.com)

**Received:** September 14, 2020; **Accepted:** September 22, 2020; **Published:** September 30, 2020

### Abstract

Neurofibroma is a benign nerve sheath tumour that develops in the peripheral nervous system. While 90% of these tumours are solitary, rest of these are multiple and can be found in association with neurofibromatosis type 1 (NF-1). Neurofibromas arise from non-myelinating-type of Schwann cells. The main difference between solitary and disseminated form depends on the nature of involvement such as disseminated form related with systemic and hereditary factors that is absent in solitary cases. This type of neurofibroma is rarely found in the buccal space but commonly present in the other spaces of head neck region. Here we present such a case who presented with solitary swelling in right cheek of 5 years duration. He was operated per-orally under local anaesthesia without sedation with excellent cosmetic outcome.

**Keywords:** Neurofibroma (NF); Buccal space; Per-oral excision

### 1. Introduction

Neurofibromas are benign, slow growing nerve sheath tumour rarely develops in the head-neck region [1]. These can arise either solitary or as a part of syndromic manifestations of neurofibromatosis type-1, which is known as Von Recklinghausen's disease [2]. World Health Organization (WHO) subdivided NF in two broad categories: Dermal and

Plexiform. Dermal neurofibroma arises from single peripheral nerve, whereas Plexiform neurofibroma arises from multiple nerve bundles [3]. Other clinicopathological subtype includes-localized (sporadic), diffuse, plexiform and epitheloid neurofibroma. Oral cavity involvement by a solitary or plexiform neurofibroma is uncommon. Sporadic cases have already been reported to develop in submandibular gland, tongue, lip, Gingiva and periosteum at the mental foramen.

Localized or solitary neurofibroma are rare in infancy, typically develops in late childhood. Solitary NF is most common, usually presents as a focal mass of peripheral nerve origin with a well-defined margin [4,5]. Surgical excision with preservation of the nerve of origin whenever possible is the treatment of choice. Surgery can be done under local or general anaesthesia. Here we present a case of solitary buccal space neurofibroma which was excised per-orally under local anaesthesia.

## 2. Case Report

A 14 year old boy presented to ENT OPD with a swelling in right Cheek of 5 years duration. According to him, his presenting complain started 5 years back when he noticed a small painless bean shaped firm swelling in his right cheek. The swelling was gradually increasing in size and took the current size of a larger plum. He also mentioned about mild local pain without any radiation. He had no other such swelling in anywhere of his body. He had no history of trauma. None of his family members had similar problem.

On local examination, there was a solitary well defined swelling in right cheek of nearly  $5 \times 4$  cm with normal skin colour and without any scar mark (FIG. 1). It was just in front of anterior border of ramous of right half of mandible up to 1 cm lateral to oral commissure. The swelling was non tender, firm in consistency, oval shaped, mobile, smooth surface, well defined margin and present on right buccal space. The intraoral mucosa was completely normal. Parotid duct opening was also normal. All other clinical examination was normal. All cranial nerve examination was normal.



FIG. 1. Cheek swelling (site of lesion).

FNAC reported benign mesenchymal lesion and suggestive of Schwannoma/ fibrous histiocytoma. Colour Doppler study showed no flow within the lesion. Patient was advised for MRI of the oral cavity, but they refused due to financial problems.

With these available investigations, surgical plan was taken. Excision was done under local anaesthesia through per-oral approach with an adequate small sized horizontal incision (FIG. 2).



**FIG. 2. Per-oral Incision.**

The mass was under the facial artery and cheek muscle. Facial artery was accidentally injured and had to ligate here. Nerve of origin could not be identified.

The mass was excised out completely with blunt dissection. It was removed in Toto without any neuronal deficit and measured about  $4 \times 3$  cm. After excision of lesion the incision site was closed in a single layer by mucosal margin closer with 3/0 Vicryl suture with adequate gap in between. No drain was kept. An adequately sized gauze piece was kept in situ to make a pressure. Patient was advised to put external pressure intermittently, to prevent haematoma. Patient was discharged 2 hours after surgery with oral antibiotic and antibiotic mouth wash (FIG. 3).



**FIG. 3. Blunt dissection of the mass (a), gross appearance of excised mass (b).**

The wound was healed within 10 days. On next follow up, 1 month after surgery, there were no neuronal deficit; mucosa was normal, no external swelling (FIG. 4).

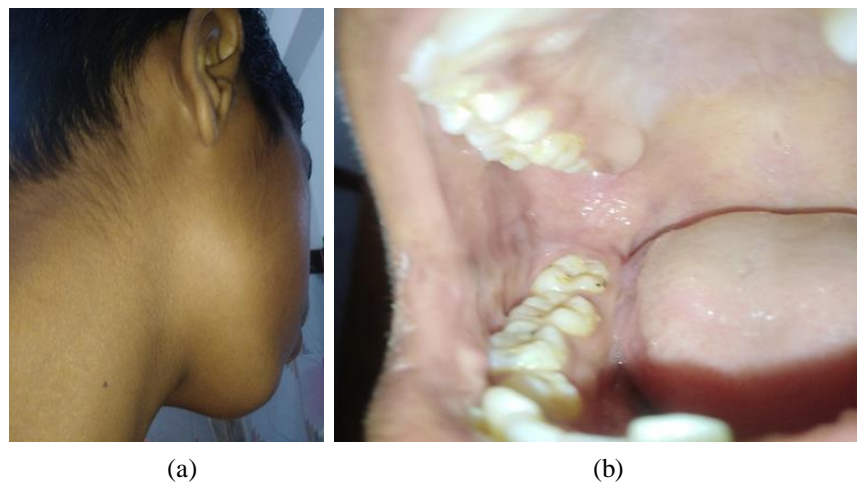


FIG. 4. (a, b). 28<sup>th</sup> POD. Extra-oral and intraoral appearance.

Histopathologically it was a benign tumour composed of spindle shaped cells and fibroblasts in a matrix of collagen fibres, reported as neurofibroma.

### 3. Discussion

Neurofibromas are considered as most common peripheral neoplasm of the skin, but uncommonly reported as buccal neurofibroma. Histologically these are non-capsulated, composed of Schwann cells, fibroblasts, perineural cells within collagenous or myxoid matrix [6]. On histology, these lesions represent all elements of peripheral nerves [7]. NF can be solitary or may have syndromic association with Neurofibromatosis type-1.

However, neurofibromatosis is an autosomal dominant disease, the exact cause of solitary neurofibroma is still unknown. The average age of presentation of solitary NF is variable, ranges from 10 months to 70 years. Though most common in early childhood but can be present at any age [8]. The rate of malignant transformation of solitary NF is very low but in association with NF-1 it is estimated to be of 5%-15% [9].

Patients usually presents with a slow growing asymptomatic swelling of variable size. Usually these are painless mass but patients may occasionally experience local pain if secondarily traumatized due to its locations. Diagnosis can only be confirmed only after excisional or incisional biopsy followed by histopathological examination. Immunohistochemistry also has role in exact diagnosis. MRI is helpful in diagnosis as well as planning of surgery. These lesions are hypointense on T1 sequence and hyperintense on T2 with “target sign” (Hyperintense rim with central area of low signal may be due to dense central area of collagenous stroma), “Fascicular Sign” on T2 sequences with heterogeneous enhancement on T1 contrast [10]. Differential diagnosis includes Schwannoma, Fibrous Histocytoma etc.

Macroscopically these have doughy or firm consistency with a whitish shiny surface. Microscopically NF contains spindle shaped nuclei spreads on a background of delicate connective tissue matrix.

Though asymptomatic, patients come for cosmetic reason and occasionally with a fear of cancer. Surgical excision is the treatment of choice, though rarely there remain chances of recurrence. Multiple recurrences have possibilities of malignant transformation though NF has very low recurrence rate as well very rare malignant transformation rate than neurofibromatosis. These lesion frequently infiltrate the nerve fascicles, that creates difficulty in separating from parent nerve and sacrifice of the nerve often requires [11].

In our case, we have excised the lesion per-orally under local anaesthesia. The limitation in our case is lack of preoperative MRI or a CT, as MRI is very much helpful to identify the nature of the lesion, extension and surgical planning, though it is not mandatory in all cases. In our case patient party refused to do MRI, so surgery was done on the basis of clinical finding.

#### 4. Conclusion

A solitary neurofibroma can be the tip of ice-berg. The patient can be syndromic, there may be other similar swelling anywhere in the body and can be a case of neurofibromatosis type-1. Patient counselling and treatment protocol is different in each case. However, as NF infiltrate between nerve fascicles, these are difficult to separate from parent nerve, so complete excision requires the sacrifice of the nerve. Surgical excision is the treatment of choice in solitary NF and can be done under local anaesthesia in certain cases.

#### REFERENCES

1. Richards D. Neurofibroma of the oral cavity. *Br J Oral Surg.* 1983;21:36-43.
2. Enzinger FM, Weiss SW. *Soft Tissue Tumors.* 3rd ed. St. Louis, Toronto, London: The C.V. Mosby Company, UK; 1995. 610-4 p.
3. Von Deimling A, Foster R, Krone W. Neurofibromatosis type -1. In: Kleihues P, Cavenee WK, editors. *WHO Classification of Tumors. Pathology & Genetics of Tumors of Nervous System.* Lyon: IARC, France; 2000. 216-8 p.
4. Johann AC, Caldeira PC, Souto GR, et al. Extra-osseous solitary hard palate neurofibroma. *Braz J Otorhinolaryngol.* 2008;74(2):317.
5. Karaosmanoglu B, Kocafe ÇY, Söylemezoğlu F, et al. Heightened CXCR4 and CXCL12 expression in NF1-associated neurofibromas. *Childs Nerv Syst.* 2018;34(5):877-82.
6. Richards D. Neurosurgeons of the oral cavity. *Br J Oral Surg.* 1983;21:36-43.
7. Patrick E. Neurofibroma pathology. *DermNet NZ.* 2013.
8. Gomez-Oliveira G, Fernandez-Alba Luengo J, Martin-Sastre R, et al. Plexiform neurofibroma of the cheek mucosa. A case report. *Med oral.* 2004;9(3):263-7.
9. Touil H, Briki S, Karray F, et al. Malignant peripheral nerve sheath tumor of the superficial cervical plexus with carotid extension. *Eur Ann Otorhinolaryngol Head Neck Dis.* 2015;132(2):93-5.
10. Khin T, Fisher C. Malignant peripheral nerve sheath tumor: pathology and genetics. *Ann Diagn Pathol.* 2014;18(2):109-16.
11. Matthew L, Maxime S, et al. Neurofibroma, radiology reference article. <https://radiopaedia.org/articles/neurofibroma?lang=us>