Surgical Management of Recurrent Neurofibroma of Infratemporal Region: A Case Report with 20-Year Follow-up

Abstract
Head-and-neck region is one of the common locations for neurogenic tumors such as neurofibroma. Neurofibromas are usually found in individuals with neurofibromatosis, which is an autosomal dominant disease. Although mostly associated with neurofibromatosis, solitary forms have also been reported. Solitary neurofibromas are rare tumors and widely described in the literature as benign. Mostly, these solitary tumors tend to occur in the gastrointestinal system. Neurofibromas of the head and neck are not uncommon, but rarely been reported to occur in the infratemporal region. We report a very rare case of recurrent solitary neurofibroma originating from the infratemporal region. Complete excision of the primary tumor was done before 20 years. Tumor arising in this anatomical location requires a conservative surgical approach for cosmetic reason.

Keywords: Infratemporal region, neurofibroma, recurrent neurofibroma

Introduction
Neurofibroma is a benign peripheral nerve sheath tumor that can be occasionally found in the head-and-neck region as multiple lesions associated with neurofibromatosis type 1 or as a solitary tumor. They usually occur between the ages of 30 and 50 years. Neurofibromas arising from the infratemporal region are quite rare, especially in children. An unusual case of recurrent infratemporal neurofibroma, diagnosed on histopathology with immunohistochemical staining, is being described. Primary tumor in the infratemporal region was surgically excised at the age of 10. Recurrence of the tumor in the same site after 20 years has been diagnosed and surgically managed.

Case Report
A 30-year-old male patient reported to our hospital with the chief complaint of swelling in the preauricular region of the right side for the past 6 months. He had a surgical history of excision of tumor in the same region before 20 years in our hospital. The tumor was excised intraorally and confirmed to be neurofibroma [Figure 1]. No positive family history was elicited. There was no similar swelling elsewhere in the body. On physical examination, the patient presented with facial asymmetry, and on extraoral examination, there was a mild diffuse swelling in the right temporal region. The swelling was soft in inconsistency and with no secondary surface changes [Figure 2a and b].

Computed tomography (CT) imaging revealed large soft-tissue density mass measuring approximately 4.21 cm × 4.55 cm within the right infratemporal fossa (ITF). It involves the medial and lateral pterygoid musculature of the right side with mild-to-moderate erosion of the pterygoid plate and posterolateral wall of the right maxilla and inferior aspect of greater wing of sphenoid bone. It extends outward through the ITF with intense postcontrast enhancement. Contrast-enhanced magnetic resonance imaging (MRI) of the head and neck was obtained which revealed a well-circumscribed mass involving the right infratemporal region [Figure 2c and d].

Histopathology revealed partially encapsulated neoplasm with skeletal muscle surrounding it. The lesion was composed at the periphery by dense fibrocollagenous connective tissue with scattered and elongated thin-walled blood vessels. Toward the center, the tumor abruptly becomes hypercellular and myxoid composed of elongated stellate...
and spindle-shaped cells with eosinophilic fibrillary cytoplasm with dark but fairly uniform nuclei. The lesion shows a prominent network composed of arcades of thin-walled vessels with the condensation of cells around the blood vessels. Immunohistochemical examination showed that the tumor cells were strongly positive for vimentin staining and parts of the cells were positive for S-100 and CD57, which was consistent with a neurogenic origin.

Surgical removal of the mass was planned under general anesthesia.

**Surgical procedure**

Under general anesthesia, standard surgical preparation was done. Preauricular transzygomatic approach to the ITF was done to access the tumor from the side. A semicoronal incision was made in front of the right ear (previous surgical scar) and the underlying tissues were dissected. The temporalis muscle was exposed, cut at its origin, and reflected to expose the skull bone below. The zygomatic arch was osteotomized to access and excise the neurofibroma nestled in the ITF. The tumor was accessed superiorly below the zygomatic arch and released laterally, medially, and anteriorly. This was followed by a transoral approach as retrieval and removal of the lesion were difficult through the site of the temporalis muscle incision. For the transoral approach, a right-sided gingivolabial sulcus approach was taken to retrieve the excised lesion. While the tumor was cut through the preauricular approach and pushed down, it was removed through the oral cavity through the gingivolabial sulcus incision. The fractured zygomatic arch was fixed with bone plate, and the temporalis muscle was repositioned. The wound was closed intraorally with 4–0 vicryl and extraoral preauricular incision with staples. Postoperative results were satisfactory with better healing of wound and esthetics [Figures 3 and 4].

**Discussion**

Neurofibroma is a benign peripheral nerve sheath tumor occurring rarely in the head-and-neck region; there is no sex predilection for neurofibromas and they commonly occur in the third or fourth decade of life. Solitary neurofibroma arises along a nerve trunk, occurring as a spontaneous lesion. The tumors are frequently centrally located and are usually not tender.

Neurofibromas should be differentiated from neoplasms derived from fibroblasts (fibromatosis and fibrosarcoma); fibrous histiocytoma; and from osseous and fibro-osseous lesions (fibrous dysplasia, ossifying fibroma, and osteosarcoma). Some neurofibromas grow by expansion and thus can erode the adjacent bone by pressure. Some can infiltrate locally, even into the bone, and can lead directly to bone resorption. Thus, bone erosion does not necessarily indicate the presence of a malignant lesion. In our case, there was erosion of the pterygoid plate, posterolateral wall of the right maxilla, and the inferior aspect of greater wing of sphenoid bone.

Tumors of the ITF are rare, constituting <1% of tumors of the head-and-neck region. Tumors involving the fossa usually extend from the surrounding areas such as sinuses, throat, parotid gland, or ear. Rarely, tumors may arise from the tissues of the ITF itself; these are called primary tumors. Tumors arising in the ITF can remain asymptomatic until they are quite large, producing symptoms late in the clinical course.

Primary neurofibroma arising from the ITF is rare; only about 12 cases have been, thus far, reported, but the recurrent neurofibroma in the same region is reported to be very few. The ITF is a quadrangular space bounded anteriorly by the maxillary sinus, medially by the

![Figure 1](image1.png)

**Figure 1:** (a-d) Excision of neurofibroma through intraoral approach before 20 years. Tumor is accessed through preauricular incision and released medially and laterally. Note the excised specimen

![Figure 2](image2.png)

**Figure 2:** (a and b) Preoperative image. Note the previous surgical scar in the right temporal region. (c) Computed tomography postcontrast image showing well-differentiated mass in the right infratemporal region. (d) Head magnetic resonance imaging showing infratemporal neurofibroma. Coronal section postcontrast T1-weighted image demonstrating a right-sided well-defined enhancing oblique
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Indian Journal of Dental Research | Volume 28 | Issue 6 | November-December 2017

pterygoid plate, laterally by the temporalis muscle and ramus of the mandible, and superiorly by the greater sphenoid wing in the floor of the middle cranial fossa. This cavity contains several structures including blood vessels and nerves, which are responsible for supplying the face and head. Due to the complex regional anatomy of the ITF, the surgical approach to this region requires careful consideration.

MRI with gadolinium is the best imaging modality of choice for evaluating the soft tissue within the infratemporal region. As can be seen in this case, initial CT scan showed erosion of the skull base and lateral pterygoid, but did not allow a clear definition of the neurofibroma [Figure 2c]. MRI with gadolinium, however, clearly revealed a well-defined enhancing mass involving the medial and lateral pterygoid [Figure 2d].

Surgical approach to tumors of the ITF is always challenging because of its deep location. They can be approached either anteriorly (from front through the face or through inside the mouth), laterally (from the side of the face near the ear), or inferiorly (from below the lower jaw at the side). To ensure thorough excision of the tumor and to overcome relatively limited access of any one single...
approach, a two-way combination approach was employed in our case which offers maximum benefit.

Recurrence and re-growth is common with plexiform neurofibroma but not with solitary neurofibroma.[9] In our case, there was recurrence after about 20 years of excision of primary tumor. The tumor was successfully removed, and the patient is now without residual or recurrent tumor 2 years’ postsurgery.

The potential for malignant transformation of neurofibroma is higher than for schwannoma.[10,11] Due to this consideration and its nonencapsulated characteristics, some authors[10] suggest that neurofibroma should receive a more radical excision.

**Conclusion**

The ITF is a complex and irregularly shaped space located deep into the zygomatic arch and the ramus of the mandible near the base of the skull. Due to its concealed regional anatomy, the ITF is inaccessible for clinical examination. In general, any lesion that occurs in the fossa usually goes unnoticed for some time until symptoms appear. Moreover, the symptoms are minimal, thus leading to delayed diagnosis. We, therefore, described a rare case of recurrent neurofibroma arising from the ITF in an adult after 20 years. A preauricular and intraoral approach was used to remove the tumor with excellent postoperative esthetics and normal facial nerve function. As these neurofibromas are of nonencapsulated nature and associated with higher risk of malignant transformation, it should receive a more radical surgical excision to ensure complete removal of the tumor.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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