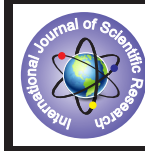


Lingual Paraneurofibroma – A Tongue Based Approach for Resection



Medical Science

KEYWORDS : tongue; neurofibroma; laser

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ABSTRACT

Tumours of the nerve sheath involving areas of the upper aerodigestive tract are not common. The involvement of the tongue is rare with isolated reports in literature. Following a diagnosis of plexiform neurofibroma, it is mandatory to evaluate for other features of neurofibromatosis.

An eight year old boy presented with a protruded tongue and difficulty in articulation due to a swelling involving the right side of the tongue. He was noted to have café-au-lait spots but other features were ruled out. The lesion was removed piecemeal via an incision on the lateral border of tongue using KTP-532 laser. An external scar and a tracheostomy was thus avoided. The histopathology was reported as Paraneurofibroma.

Introduction

Neurofibroma is a benign tumour arising from the peripheral nerve¹. They may be either solid or plexiform type and solitary or multiple. There are seven types of neurofibromatosis (NF), of which NF type 1 is the most common. It comprises of 85-90 % of the cases. NF-1 is characterized by café-au-lait spots, multiple neurofibromas, iris Lisch nodules, CNS tumours, macrocephaly, mental deficiency, seizures, short stature and scoliosis². Plexiform neurofibroma is a term given for a neurofibroma which arises from a large nerve trunk producing multiple irregular fusiform swellings along the nerve creating a tangled worm like mass³. The plexiform variety may occur anywhere in the body but more commonly on the skin. The oral cavity is a rare site where it may involve the buccal region, palate, tongue and lips¹.

Von Recklinghausen disease which included neurofibroma of the tongue was first described in 1849 by Robert Smith³. Plexiform neurofibroma may also be seen as an oral manifestation of the multiple endocrine neoplasia syndrome (MEN) type 3¹.

A transcervical approach has been commonly described in literature for the lingual neurofibroma. A labiomandibulotomy with or without a tracheostomy may be required for adequate exposure and to secure the airway, respectively. Local resection preserving the lingual mucosa has also been described in literature⁴.

Case Report

An eight year old boy presented to the Department of Otolaryngology of a tertiary referral hospital with a complaint of swelling in the right side of tongue noticed by his parents since birth. The swelling has been gradually increasing in size since then. At presentation, the tongue was protruded and the patient could not close the mouth. He had difficulty in articulation. There was no difficulty in swallowing or breathing. On examination, the entire right half of his tongue appeared grossly hypertrophied when compared to the left. The swelling was uniformly spread from the tip up to and just crossing to the base of the tongue on the right side (Figure 1a). The tongue movements were restricted due to the size. On palpation, the swelling was firm but non tender. Café-au-lait spots were noted over the chest.

An ultrasound Doppler was done which showed a heterogeneous echotexture with multiple small non echoic areas, some of them showing colour fill involving the deep part of the posterior portion of tongue with normal musculature in the superficial part. These features were suggestive of a haemangioma.

Under general anaesthesia, using KTP-532 laser an incision was given along the right lateral border of the tongue and submucosal flaps were raised towards the midline and towards the floor of the mouth to expose the lesion. The tumour was removed piecemeal from the underlying muscle using blunt dis-

section and laser (Figure 1b and 1c). The incision was closed with 3-0 Vicryl[®]. The patient was put on intravenous dexamethasone. Extubation was done the following morning over a tube exchanger. There was no breathing difficulty though edema was present. The steroids were continued for three days. The patient was discharged on the fifth postoperative day. On histopathological examination, hyperplastic squamous epithelium overlying a diffuse neurofibroma composed of spindle shaped cells surrounded by collagen, thin walled vascular channels and admixed tortuous nerve profile containing intraneural neurofibroma composed of thickened perineurium encompassing a central distorted nerve bundle surrounded by spindle shaped cells in a myxoid matrix was seen (Figure 2). A diagnosis of Paraneurofibroma (plexiform and diffuse neurofibroma) was made. A work up for NF-1 was made. It was negative except for Café-au-lait spots. A screening for features of MEN 3 was also negative. At the first follow up after three weeks, a residual swelling was noted in the posterior part of the oral tongue and adjoining base of tongue. Using the same approach, but for a more posterior extension of the incision and the residual tumour was resected using KTP-532 laser.

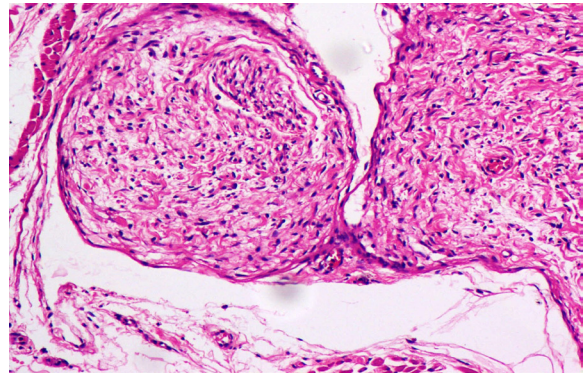
The patient followed up two months later. There was no evidence of residual disease. The patient has been lost to follow up thereafter.

DISCUSSION

Approximately, 9-18 % of neurofibromatosis arising in NF-1 may be located in the head and neck region². The reports in literature involving the oral cavity have been sporadic^{5,6}. The oral manifestation of NF occurs in 4-7 % of patients with neurofibromatosis³. The majority of these cases involve the tongue. The lesion arises from or adjacent to the cranial nerve supplying the tongue i.e the hypoglossal and /or the lingual nerve³. It may be difficult to ascertain the nerve of origin. The clinical presentation may be with pain, sensitivity and paraesthesia. Large lesions involving the tongue can present with difficulty in swallowing, breathing and articulation. The enlarging tongue may protrude out of the mouth keeping it persistently open with drooling of saliva. It may present as a submandibular swelling mimicking a submandibular gland tumour⁷. Differential diagnosis of such a tongue swelling in childhood should include neurofibroma, schwannoma, lymphangioma, cavernous hemangioma, hamartoma, teratoma, lipoma, myofibroma and myofibromatosis, leiomyoma, cystadenoma, pyogenic granuloma, congenital granular cell tumour and cystic lesions such as mucoid cyst, dermoid cysts and cysts of foregut origin⁴. The treatment is surgical in such cases with diagnosis being confirmed only by a histopathological examination. In a swelling involving the tongue, the possibility of neurofibroma should be kept in mind and features of neurofibromatosis looked for. The evaluation for characteristic features of NF-1 i.e café-au-lait pigmentation of skin, multiple neurofibromas, iris Lisch nodules, central nervous sys-

tem tumours, macrocephaly, mental deficiency, seizures, short stature and scoliosis should be done². In addition, evaluation for features of MEN 3 i.e the thyroid, adrenal gland and skin is mandatory¹. The treatment of neurofibroma of the tongue involves resection of the lesion via a transcervical approach with or without a labiomandibulotomy for exposure. A preliminary tracheostomy was considered mandatory in earlier reports in literature^{3,8}. In our case, we had used an incision along the lateral border of the tongue on the side of the lesion using KTP-532 laser. With the use of laser, the bleeding was minimal and it also facilitated dissection of the lesion from the underlying muscle. Flaps were raised medially towards the midline and inferiorly towards the floor of mouth. This gave adequate exposure for removal of the tumour piecemeal. An external scar was avoided. A labiomandibulotomy would cause morbidity in a child and may affect the growth of the mandible causing dentofacial deformity. The patient was extubated via a tube exchanger. Thus, a tracheostomy was avoided.

The nerve from which the tumour arises may be resected if the lesion cannot be dissected. Though it may infiltrate the intrinsic muscle and have a poor cleavage plane, recurrence ~15 % of cases but this is rare in the solitary form of neurofibroma¹. The prognosis of neurofibrosarcoma associated with NF-1 is poor^{2,9}. Patients with plexiform neurofibroma need to be followed up for malignant transformation, recurrence and other symptoms and signs of features of von Recklinghausen's disease⁴.



Legends:

Figure 2: Histopathological examination (20 x) showing plexiform neurofibroma



Legends:

Figure 1:1a:Pre operative swelling over right half of the tongue

1b:Intraoperative picture showing the approach using laser

1c:Approach showing the tongue flaps being raised

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