A case of symmetrical lipomatosis of the tongue presenting as macroglossia

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Lipomas are the most common benign soft tissue mesenchymal tumours composed of mature adipose tissue. They are uncommon in the oral and maxillofacial regions, with 15-20% of cases involving the head and neck region and less than 5% of all benign oral lesions. Multiple symmetric lipomatosis is rare and characterized by diffuse growth and nonencapsulated lipomas. It is usually found in the posterior neck and upper trunk and they are relatively infrequent on the oral and maxillofacial regions like Madelung disease. In the report, we describe a rare case of symmetrical lipomatosis of tongue with OSAS and Dysarthria. This lesion was resected under general anesthesia. Intraoperative findings revealed only adipose tissues with replacement of lingual muscles and no capsulation. The lesion was finally diagnosed as symmetric lipomatosis of the tongue based on clinical radiological and histologic examination. SLT (Symmetrical lipomatosis of the tongue) is an extremely rare case that appears like a macroglossia. Partial glossectomy is the treatment of choice because of the improvement of symptoms and the low rate of recurrence.

KEY WORDS: Macroglossia, Oral lipoma, Tongue lipomatosis

Introduction

Lipomas are the most common benign soft tissue mesenchymal tumours composed of mature adipose tissue. They are uncommon in the oral and maxillofacial regions, with 15-20% of cases involving the head and neck region and less than 5% of all benign oral lesions. Lipoma of oral cavity may occur in any site but the cheek is the commonest site of occurrence followed by tongue, mouth floor, buccal sulcus and vestibule, palate, lip and gingiva. Multiple involvements are rare. A few cases of bilateral symmetrical lipomatosis (BLS) have been reported in literature and this disease usually affects the neck and upper trunk with only rare involvement of the tongue. Symmetrical lipomatosis of the tongue (SLT) is a symmetrical and diffuse swelling of the bilateral tongue border and some investigation have suggested that SLT may be a subtype of BLS. We report a rare case of SLT infiltrating grown of nonencapsulated mature adipose tissue, presenting as macroglossia with obstructive sleep apnea syndrome (OSAS) and dysarthria.

Case Report

A 68-year-old Italian male was referred to our department with gradually progressive painless slowly growing and worsening swelling on both lateral borders of the tongue that had been present for over 5 years.
The patient had tongue dysmotility and dysarthria, upper airway obstruction with OSAS, dysphagia and psychosocial impairment for the last 8 months; he denied other neurosensory diseases, including dysgeusia, motor or autonomic neuropathy. The patient was not obese and did not abuse alcohol; there was no family history of illness. Clinical examination showed symmetric bilateral tongue masses: the size of the tongue was 15 cm x 9 cm x 8 cm with a soft and elastic consistency (Fig. 1-2). The overlying mucosa was normal in color and texture with no ulceration or inflammation. On palpation the lesions were rubbery and no fluctuant. There were no cervical masses and no other swellings anywhere in the body. Magnetic resonance imaging (MRI) was performed for differential diagnosis and preoperative evaluation of the lesion. T1-weighted images revealed a widespread high signal intensity of the tongue, while fat saturation T1-weighted images revealed low signal intensity in the same region (Fig. 3). Polysomnography revealed AHI index >15 >30 and oxygen saturation was > 80%.

Other investigation included complete blood tests examination revealed that the full blood count, electrolytes, liver function test and blood glucose were all normal, only the cholesterol was elevated.

An incision biopsy showed lobules of mature adipose tissue in the tongue submucosa suggested lipomatosis. The lesion was removed under general anesthesia and we performed a glossectomy reduction by variant of Dingman glossectomy a peripheral excision category and suffer from the drawback of reducing the bulk of the tongue at the periphery while leaving the centre and base bulky. The operative technique used in this case is based on the submucosal vascular plexus, which allows excision of an intramuscular wedge to reduce the bulk of the tongue, along with the longitudinal wedge excision to reduce the width, and the transverse wedge to reduce the length. This helps in giving a uniform reduction of the macroglossia in all three dimensions. While reducing the bulk of the tongue it spares the papillae at the dorsum which are responsible for salty taste, the papillae at the sides which are responsible for sour taste, and the papillae at the base for bitter taste.

**Pathological finding**

Surgical specimen was constituted of two excision respectively of cm 10x4x6 and cm 7x3x2 (Fig. 4). The cut surface showed multiple yellowish areas, with soft consistency.
Histological examination showed a normal squamous epithelium overlying a mature adipose tissue lesion, that replaces the normal smooth muscle tissue. No pleomorphism or atypia was observed. Albeit lipomatous cells showed a vague variety of size, no evidence of nuclear atypia or mitotic figures was detected. No multivacuolated lipoblasts were seen (Fig. 5).

An immunohistochemical analysis was performed against S-100 (Novocastra clone S1/61/69 BOND ready to use) Vimentin (Dako Anti-Vimentin clone V9, IS630, ready to use), Actin (Novocastra clone HHF35 BOND ready to use), and Desmin (Novocastra clone DE-R-11, BOND ready to use), to define the cells population. Negative controls were achieved by substitution of the primary antibodies with normal rabbit serum. Blood vessels, normal tissue macrophages and muscle cells were used as positive controls. The fat tissue was positive for S-100 and Vimentin, while the residual muscular component interpersed into the adipose tissue, was positive for Desmin, Actin and Vimentin.

The morphological and immunohistochemical analysis were consistent with a diagnosis of multiple lipomas of the tongue / lipomatosis of the tongue.

The patient’s postoperative course was uneventful and without functional deficits. The mobility of the tongue is not affected as the tongue base and frenulum are not touched (Fig. 6).

In order to assess the patient’s dysarthria, OSAS and dysphagia, we evaluated his speech function on two weeks after surgery using a listening test in which he was asked to speak Italian words. The listening test indicated that the patient’s dysarthria had resolved. Regarding dysphagia and OSAS, his tongue was now accommodated comfortably within the oral cavity with significant improvement of deglutition.

Post-operative Polysomnography revealed, also, normal saturation and no more apnea events. He was well with no local recurrence and regrowth after 36-month follow-up period.

Discussion

Lipomas is a benign tumour composed of adipose tissue, and is the most common form of soft tissue tumour. Presentation as multiple tumours is infrequent, some are associated with inherited disorders, for instance as in Gardner’s or Bournville Syndrome, multiple lipomatosis or macroglossia.\(^4^,^5\)

Enzinger and Weiss classified benign lipomatous lesions into 5 categories: lipoma, variants of lipoma, heterotopic lipoma, infiltrating or diffuse neoplastic or non-neoplastic proliferation of mature fat, hibernoma; the fourth group includes also BSL, diffuse lipomatosis and pelvic lipomatosis.\(^6\)

Lipoma of the tongue is rare and often presents as a single, superficial, pedunculated or sessil lesion, which accounts
Lipomatosis of the tongue (SLT), first reported by Desmond in 1944, involves only the tongue and was reported predominantly in Asian people in absence of obesity and sometimes also without a history of alcoholism. Although SLT is been previously described as a variant of BSL, this condition is likely separate dese 8,9. Our case of SLT is particular because the patient was not an Asian man and had not a history of alcoholism and obesity. Only the clinical and histopathologic features are common with other reported cases of SLT in literature. Microscopically, it is composed of mature adipocytes; however, in 20% of cases, it demonstrates histologic variants that includes spindle cell lipoma, pleomorphic lipoma, angiolipoma, fibrolipoma, myxoid lipoma, and atypical lipoma.

Preoperative magnetic resonance imaging is useful for the management plan. The strikingly high intensity signal on both T1- and T2-weighted images are suggestive of lipoma and it further delineates the extent of tumor involvement. Lipomatosis of the tongue had an ill defined margin because of infiltration of muscles 10. A lipoma differs from normal body fat in that its lipid is not available for metabolism and it is surrounded by a capsule. In accordance with others Authors (Chikui) in our case we observed high signal intensity masses detected bilaterally in the tongue and curved lines of low signal intensity running bilaterally from the lingual septum (the remnants of the internal lingual muscles) 11,12.

The treatment for the SLT is surgical resection when associated with functional disorder (obstructive sleep apnea, dysarthria, etc.), with the aim to restore function. Lipomatosis penetrates deep into the lingual tissue therefore complete resection of the lesion is almost impossible so partial glossectomy is the surgical treatment of choice 13. The local recurrence of intramuscular lipoma has been reported to range from 19 to 62,5% 5 due to the difficulty of radical surgical excision. No recurrence case in the tongue have been reported in literature, regardless post-operative follow-up is raccomended.

Surgical excision provides more tissue for examination than fine needle aspiration and sometimes the only way to make certain diagnosis and to exclude other pathology for differential diagnosis of macroglossia: Congenital (Down’s syndrome, Cretinism, Beckwith-Wiedemann syndrome or exomphalos, Macroglossia or gigantism syndrome); Inflammatory (Lymphoedema); Trauma (Lingual haematoma); Neoplasm (Benign like neurofibroma, haemangioma, lymphangioma or malignant like squamous cell carcinoma); Metabolic disorders (Glycogen storage disorders, Amyloidosis); Endocrine (Acromegaly, Myxoedema); Miscellaneous (Angioneurotic edema, Madelung Disease) 7-10.

Conclusion

SLT is an extremely rare case that appears like a macroglossia. Partial glossectomy is the treatment of choice because of the improvement of symptoms and the low rate of recurrence described in the literature. Our case was clinically and histopathologically typical and the treatment performed was effective in restoring normal tongue function and speech.

Riassunto

I lipomi sono i più comuni tumori mesenchimali dei tessuti molli, composti da tessuto adiposo maturo. Sono rari nell’ambito orale e nella regione maxillo-facciale, con il 15-20% dei casi riguardanti il capo e la regione del collo, e rappresentano meno del 5% di tutte le lesioni benigni del cavo orale. La lipomatosi multipla e simmetrica è rara, ed è caratterizzata da lipomi non capsulati ed a crescita diffusa. Si localizzano abitualmente posteriormente nel collo e nel tronco prossimale, e sono relativamente rari a livello orale e maxillo-facciale come nel caso della malattia di Madelung.

In questa presentazione si descrive un raro caso di lipomatosi simmetrica della lingua con OSAS (Obstruction Sleep Apnea Syndrome) e disartria. La lesione è stata asportata sotto anestesia generale. Il reperto intraoperatorio ha dimostrato soltanto tessuti adiposi senza capsula e sostituzione dei muscoli linguali. La diagnosi finale è stata di lipomatosi della lingua in base ai reperti clinici, radiologici ed istologici. La LSL (Lipomatosi simmetrica della lingua) rappresenta un caso estremo che si presenta clinicamente come macroglossia. Il trattamento di scelta è una glossettomia parziale che determina un miglioramento della sintomatologia ed una bassa incidenza di recidiva.

References

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