

A Rare Case of Childhood Lipoblastoma presenting as Tongue Mass

<https://doi.org/10.47210/bjohns.2021.v29i1.403>

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ABSTRACT

Introduction

Lipoblastoma is a rare benign tumour arising from embryonic white fat been commonly noted in limbs and trunk, but tongue involvement is rare and has not yet been reported.

Case Report

A child with tongue lipoblastoma is reported, whose imaging reported an encapsulated, well-delineated, fat-containing tumour. Surgical excision was performed with no post-operative morbidities.

Discussion

Lipoblastoma is an uncommon childhood tumour, which rarely affects the tongue. It presents as a progressive painless swelling, rarely causing any symptom. MRI is helpful to assess the precise location and extent of the lesion. Although the ratio of fat to myxocollagenous tissue in the tumour is variable, the diagnosis can be suggested in most cases based on the imaging characteristics. Recommended treatment is complete surgical excision and confirmation of diagnosis by histopathological examination.

Keywords

Lipoblastoma; Child; Tongue

The most commonly encountered paediatric soft tissue tumour in the head and neck region is haemangioma. Lipoblastomas, though rare, should be kept in mind in cases of such presentation. Of these, a clinical diagnosis of lipoblastoma can be made taking into consideration the age of patient and clinical examination first, followed by a probable confirmation with MRI ± CT scan.

Case Report

A 4 year old boy, reported to the otorhinolaryngology outpatient department, with the complaints of painless swelling in the tongue for 45 days. The swelling gradually increased in the size to attain the present size. Lesion did not cause any other symptoms.

On examination of tongue, a smooth, firm, non-tender swelling of 3x2.5x2.5cm, non-compressible swelling noted in the posterior 1/3rd of dorsum of tongue (involving midline and right side of tongue). The function

of hypoglossal and lingual nerve was spared (Fig. 1).

Pre-operative haematological investigations were normal. USG Neck was normal and showed a normal thyroid gland. T2 - weighted images of MRI showed a 2.7x2.3x2.9cm hyper intense, well encapsulated, soft tissue lesion involving midline and right side of tongue. (Fig. 2) The T1 images showed a hyper intense peripheral rim which is compatible with fat and a hypointense centre. (Fig. 3)

Surgical resection was planned. Informed and written consent was obtained. Under general anaesthesia, in-toto surgical resection of encapsulated lesion was done using a midline Glossectomy approach (Fig. 4), without any

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Fig. 1. Pre-op picture showing swelling in posterior aspect of tongue

post-operative morbidity (Fig. 5).

Histopathological examination showed mature adipocytes which were organized in lobules separated by fibrous septae, few immature lipoblasts, myxoid/myxocollagenous stroma, plexiform capillary network and an intact capsule. (Fig 6)



Fig. 3. MRI T1 axial section: Well encapsulated lesion within tongue with rich surrounding capillary network

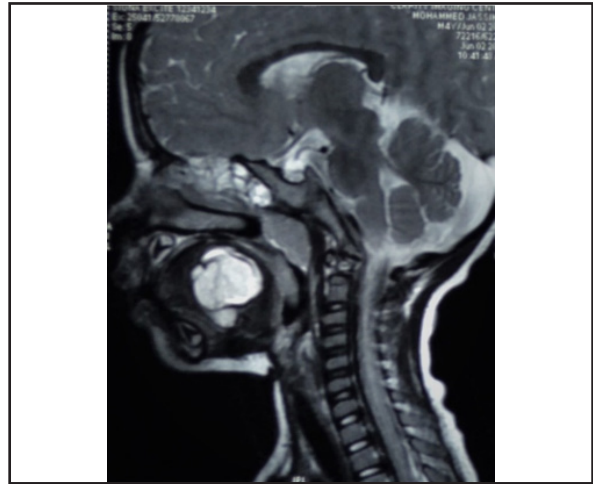


Fig. 2. MRI T2 coronal section: Encapsulated lesion within tongue, with rich vascularity

Discussion

Lipoblastomas and their multicentric/infiltrative forms, lipoblastomatoses, are rare benign soft-tissue tumours of embryonic lipoid cells. Adipose tumours comprise about 6% of soft tissue neoplasms of which 94% are lipomas, 4.7% are lipoblastomas, and 1.3% are liposarcomas.¹ These are known to develop in the first two decades of life.

Lipoblastoma mainly occurs before the age of three



Fig. 4. Intra-op. photograph: Excision of encapsulated mass in-toto

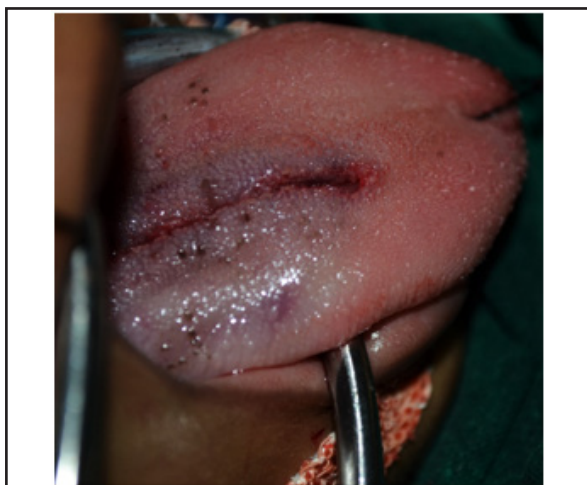


Fig. 5. Appearance post-excision and closure

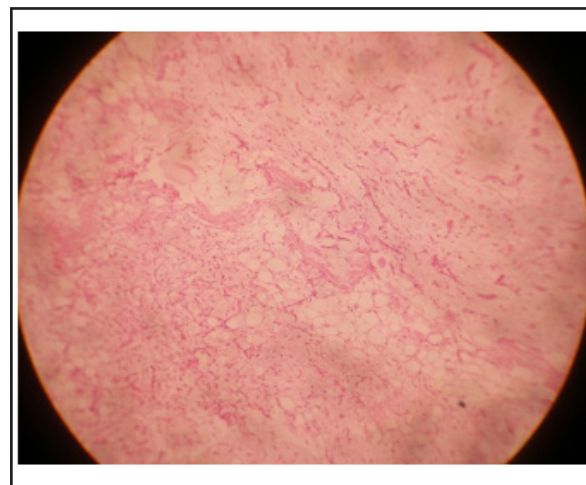


Fig 6. Lipoblastoma: Infiltrating islands of tumour tissue (H&E, 100X)

years and has a male predominance. This tumour is mostly present in the limbs and trunk, with its rare occurrence in the head and neck.² Lipoblastoma is categorized into two types: the circumscribed lipoblastoma (approximately 70% of cases) - a superficial and encapsulated lesion and diffuse lipoblastomatosis (about 30% of cases) - a deeply located, poorly circumscribed lesion with infiltrative growth pattern that may affect surrounding muscle structures.³

The terms lipoblastoma and lipoblastomatosis were first used by Jaffè⁴ and Vellious⁵, respectively. Less than 200 cases of lipoblastoma and lipoblastomatosis at various locations have been reported in the literature.⁶

The most common presenting symptom is a painless, progressively growing mass which is localized superficially. Other symptoms are related to the location and size or mass effect of the lesion. Airway obstruction and respiratory symptoms have been described in patients with pleural, mediastinal, pulmonary, and lower neck lipoblastomas. Gastrointestinal symptoms, such as emesis, diarrhoea, anorexia and abdominal pain occur in patients with mesenteric or retroperitoneal lipoblastomas. Depending on the location, nerve compression and related symptoms can be present.^{7,8}

Head and neck lipoblastoma cases have rarely been reported in literature owing to their low incidence. Calhoun et al. reported the first case of lipoblastoma

in salivary gland which occurred in the parotid gland.⁹ Rasmussen et al. mentioned a case of cervical lipoblastoma causing intermittent airway obstruction.¹⁰ Farrugia et al. reported another case of lipoblastoma in the neck which had presented with rapidly enlarging mass, mimicking cystic hygroma.¹¹ A case of lipoblastoma in parapharyngeal space was reported by DePasquale et al.¹²

On imaging, lipoblastoma appears as a well-defined soft tissue mass, often with lobular appearance and having internal septations. The imaging appearance of lipoblastoma depends on the proportion of fat relative to the amount of myxocollagenous stroma. Fat in lipoblastoma appears as hyperechogenic areas on ultrasonographs, areas of low attenuation on CT images and signal intensity identical to that of subcutaneous adipose tissue. The myxoid components are hypoechoic on ultrasonographs, have low attenuation on CT images (but less hypodense than fat) and on MRI have low signal intensities on T1-weighted images and high signal intensities on T2-weighted images; contrast enhancement of these areas reflects the rich capillary network.^{8,13,14}

Lipoblastoma exhibits a tendency to invade locally. If not excised early, it may enlarge, and infiltrate the various surrounding spaces, present pressure symptoms and may lead to various complications as well.

Fine needle aspiration cytology is the basic investigation for diagnosis. It can be confirmed by excision biopsy and histopathological examination of the specimen. Histologically, the lesions are composed of immature fat cells (lipoblasts) in varying stages of maturity, mesenchymal cells, a plexiform capillary network, myxoid stroma and mature adipocytes organized in lobules by fibrous septa. There is no nuclear atypia.

If the pathologist is unable to differentiate lipoblastoma from myxoid liposarcoma, a cytogenetic analysis should be done. The typical chromosomal abnormality associated with lipoblastomas is breakpoints in the long arm of chromosome 8 (area 8q11-13), whereas myxoid liposarcomas typically show translocation t(12,16) (q13;p11) and atypical lipomatous tumour shows amplification of the MDM2/CDK4 genes on 12q[7-10].¹⁵⁻¹⁹ The age of our patient, the tumour lobulation, its well-circumscribed nature, and the typical histological findings in our case made cytogenetic investigations unnecessary.²⁰

CT scan or MRI is a must to assess the tumour extent and also to plan surgical approach. Reiser et al reported that MRI is the most reliable method, with USG and CT having complementary roles.²¹ It is difficult to distinguish lipoblastomas from liposarcomas by imaging studies because of their similar feature of vascularity. However, lipoblastomas usually occur within the first 3 years of life, and diagnosis after the age of 5 years is unusual. In contrary, liposarcomas are extremely rare prior to 5 years of age.

The natural history of lipoblastoma is to evolve into mature lipomas.^{8,22} The treatment used for lipoblastoma and lipoblastomatosis is wide surgical resection.¹⁷ Recurrence develops in 9 to 25% of cases and is mostly seen in cases of with infiltrative lipoblastomatosis and incomplete resection.⁸ Metastasis has not been reported to date and the prognosis is good.

Conclusion

Though lipoblastoma is an uncommon childhood tumour, it should be taken into consideration as a differential diagnosis of head and neck masses. It usually presents

as a progressive painless swelling, rarely causing any symptom. Imaging is helpful showing the precise location and extent of the lesion. Although the ratio of fat to myxocollagenous tissue in the tumour is variable, the diagnosis can be suggested in most cases based on the imaging characteristics. Recommended treatment is complete surgical excision and confirmation of diagnosis by histopathological examination.

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