

Lipoblastoma in head and neck – A rare childhood tumour

Sinha BK¹, Thapa N², Banskota DK³

¹Professor, ²Lecturer, ³Associate Professor, Department of ENT and Head & Neck Surgery, Tribhuvan University Teaching Hospital, Maharajgunj, Kathmandu, Nepal

Abstract

Lipoblastoma is an uncommon benign mesenchymal tumour that occurs primarily in children younger than 3 yrs of age. Despite the lesions being benign, great difficulty can be encountered in its management because of its tendency to invade the different fascial planes. A rare case of huge lipoblastoma diagnosed in a 22 months old child, involving various spaces of face and skull base has been reported. This huge tumour was completely and successfully removed through cheek incision without any postoperative complications.

Key words: Lipoblastoma, Benign Mesenchymal Tumour, Head & Neck, C.T., M.R.I., FNAC, Total excision

Lipoblastoma and Lipoblastomatosis are rare benign neoplasms of foetal white fat tissue that occurs almost exclusively in infants and children younger than 3 years of age. A total of less than 100 cases, with only 10 cases involving head and neck have been reported previously in the literature. Among the rarer ones, two cases have been reported affecting the Parotid region and one involving the parapharyngeal space. We are reporting a case, which was involving cheek, parapharyngeal space, pterygopalatine and infratemporal fossae. Such an extensive tumour has not been reported in literature.

Case report

Parents of a 14 months old child noticed a small painless swelling in front and just above the left angle of the mandible which gradually increased within 8 months, to a size involving the entire left side of face and cheek. The mass at the time of presentation was found to be occupying the entire left side of face extending from temple to the lower border of mandible. The swelling was painless to start with but later on, parents noticed that the child was not able to eat or chew any food stuff and used to cry while eating. Few ulcers and raw areas were noticed on the buccal aspect of the left cheek probably due to dental trauma. There was no history of profuse bleeding from the oral cavity, trismus, difficulty in swallowing, change of voice, external trauma or fever.

On examination

There was a diffuse swelling present on left side of her face extending from the lower border of tragus superiorly to lower border of mandible. The swelling

extended anteriorly up to the angle of mouth and posteriorly to the retro mandibular sulcus and ear lobule. The overlying skin was normal. The swelling was soft to firm on palpation and was bimanually palpable measuring about 6x6cm in size. Anteriorly it could be palpated from 1cm posterior to the angle of mouth up to just anterior to the ear lobule posteriorly. Examination of the oral cavity showed diffuse swelling of the buccal aspect of her left cheek extending from retromolar trigone to about 1cm behind the angle of mouth occupying the entire area between the superior and inferior gingivobuccal sulci. There were multiple ulcers adjacent to the teeth.

Investigations

CT scan revealed a mass of fat attenuation value occupying the left parapharyngeal and infratemporal fossa, compressing the posterior wall of the left antrum and also extending to cheek. There was no intracranial extension. (Fig. 1 and 2). Fine Needle Aspiration Cytology (FNAC) from the mass was reported as Lipoblastoma. To confirm the diagnosis, under general anaesthesia, an incision about 1.5 cm long was given just posterior to the angle of mouth and biopsy of the mass was taken and sent for histopathological examination which showed that the tumour contained lobulated mature adipose tissue and myxoid tissue with lipoblasts thus confirming the diagnosis of Lipoblastoma.

Correspondence

Prof. Bimal Kumar Sinha
Dept. of ENT and Head & Neck Surgery
T.U. Teaching Hospital, Maharajgunj, Kathmandu, Nepal
Email: bimal@healthnet.org.np

Fig. 1: C.T. Scan showing the lesion involving parapharyngeal and infratemporal fossae.

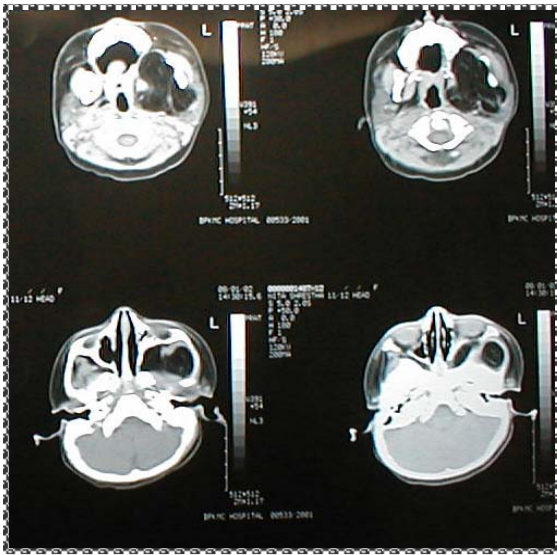
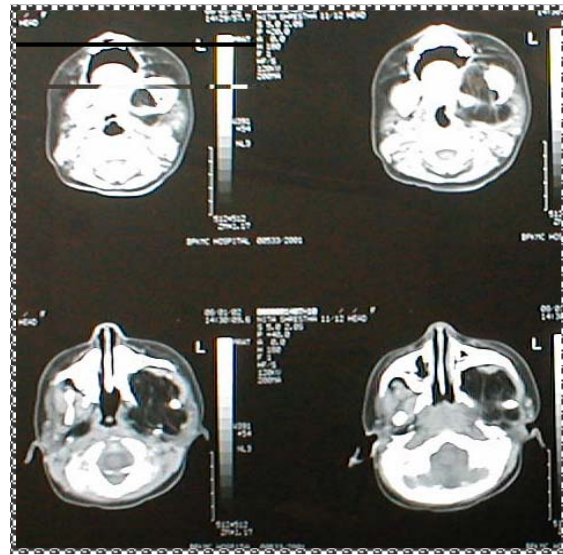


Fig. 2: C.T. Scan showing the compression of the posterior wall of maxilla

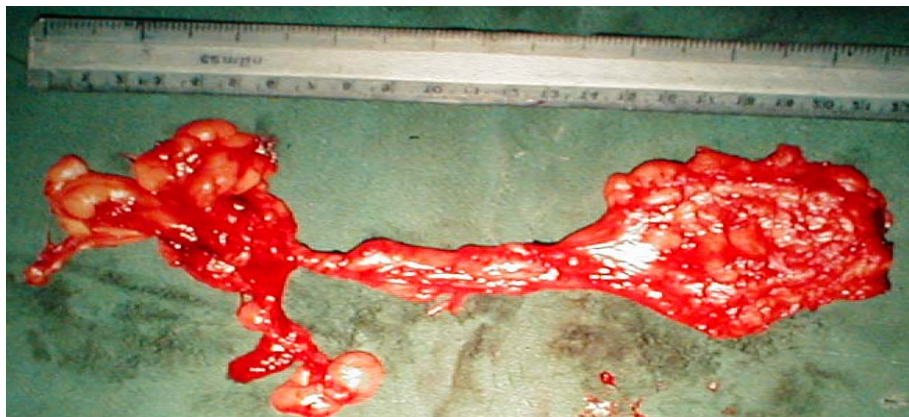


Treatment

After confirming the diagnosis, the child was subjected to surgery under orotracheal general anaesthesia. Boyle-Davis mouth gag was applied and a 5cm vertical incision was given about 1.5cm behind the left angle of mouth. The capsule of the underlying mass was reached. Blunt dissection was done on medial as well as lateral aspect, staying very close to the capsule of the mass. By applying firm traction and extracapsular blunt dissection, the entire mass was removed in toto (Fig: - 3). Perfect haemostasis

was attained and the cavity approximated by applying absorbable sutures. The incision was sutured using 4.0 vicryl sutures. Nasogastric tube was inserted. Pressure bandage was applied for about 24 hrs. The nasogastric tube was removed on the third postoperative day. The postoperative recovery was uneventful. One year follow up has revealed no recurrence clinically, her facial swelling has disappeared and there is no trismus. Repeat C.T. scans could not be performed due to financial constraints.

Fig. 3: Excised mass



Discussion

Lipoblastoma is a rare benign tumour arising from embryonic white fat which occurs in early childhood. It usually occurs in extremities. As mentioned above, very few cases of lipoblastoma in head and neck region have been reported in literature. Dilley et al. reviewed 24 patients that presented to a single institution over a 15-year period. Of these 24 patients, only 4 had the lesion in head and neck region. The tumour may present as focal or diffuse lesion with the former being more common. It has also been mentioned that focal lesions usually do not require further surgery following initial resection.⁴

The case reported here is the first case of lipoblastoma in head and neck reported so far in our context and it is one of the youngest cases of head and neck lipoblastoma reported in literature. Patients with lipoblastoma may present with various symptoms depending upon the site of its involvement. Calhoun et al. reported the first case of lipoblastoma in salivary gland which occurred in the parotid gland.² Rasmussen et al. mentioned a case of cervical lipoblastoma causing intermittent airway obstruction.³ Farrugia et al. reported another case of lipoblastoma in the neck which had presented with rapidly enlarging mass, mimicking cystic hygroma.⁴ A case of lipoblastoma in parapharyngeal space was reported by DePasquale et al.⁵ Lipoblastoma exhibits a tendency to invade locally. If not excised early, it may enlarge, and infiltrate the various surrounding spaces as in this case, present pressure symptoms and may lead to various complications as well. Sun et al. reported the first case of hemiparesis resulting from left sided supraclavicular lipoblastoma compressing the spinal cord.⁶ Singh et al also reported the first case of congenital lipoblastoma in the scalp extending up to the left upper eyelid and eyebrow.⁷ Our patient had a diffuse swelling on the left side of her face, buccal mucosa of cheek and retromolar trigone with multiple traumatic dental ulcers. She had difficulty in eating due to the mass and repeated cheek ulcerations.

Fine needle aspiration cytology is the basic investigation for diagnosis. It can be confirmed by open biopsy and histopathological examination of the specimen. CT scan or MRI is a must to assess the tumour extent and plan surgical approach. Leon et al. described a case of lipoblastoma of the parotid region in a 6-year-old boy which was diagnosed by FNA.⁸ Sakaida et al. described a case of huge lipoblastoma of the neck in which Magnetic resonance imaging (MRI) revealed a 7 x 7 cm neck mass that extended into the parapharyngeal and paratracheal spaces.⁹ In

our patient CT scan was done which revealed a mass of fat attenuation value occupying the left Parapharyngeal and Infratemporal fossa compressing the posterior wall of left antrum and also extending to the cheek. FNAC was done for diagnosis and was confirmed by open biopsy and histopathological examination prior to surgical exploration.

Local excision of the mass is the treatment of choice.¹⁰ However, due to extension of the tumour in different fascial planes, complete excision sometimes may not be possible. In our patient despite the huge tumour involving many spaces, we could remove it in toto by transoral approach without any complications postoperatively. We found that it was important to stay right on top of the capsule of the lesion and dissect bluntly applying gentle traction and remove it as a single piece.

Conclusion

Lipoblastoma is a poorly understood and uncommon soft tissue tumour of infancy and early childhood. Though lipoblastoma is an uncommon childhood tumour, it should be taken into consideration as a differential diagnosis of head and neck masses. Differential diagnosis of Lipoblastoma must be considered in deep parotid, Parapharyngeal, Infratemporal, Temporal or even cheek masses in paediatric population. Sometimes it may present as a huge head and neck mass causing airway obstruction, spinal cord compression or difficulty in swallowing. Though benign, it gives great difficulty in its management, due to its extensions to different fascial planes. However complete excision is still possible despite its involvement of multiple spaces of Head and Neck.

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