Sialoblastoma: A rare pediatric tumour of the salivary glands
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Citation

Abstract
Sialoblastoma is a rare congenital epithelial tumour of the salivary gland that is diagnosed at birth or shortly thereafter with significant variability in histology and clinical course. We report the case of an eight-year-old boy who had a left parotid tumour for one year. MRI scan demonstrated the tumour to be arising from the superficial lobe of the parotid gland. After making a histological diagnosis by fine-needle aspiration cytology, a superficial parotidectomy was performed with preservation of the facial nerve. There is no recurrence at one-year follow-up.

INTRODUCTION
Tumours of salivary glands are very uncommon in children. Sialoblastoma, which is also called embyroma, represents a neoplastic proliferation of cells of organ rudiments, and it is the most frequent type of congenital epithelial tumour of the salivary glands. Sialoblastoma has been reported to occur predominantly in the parotid gland and very rarely in the submandibular gland. Although it is a slowly spreading tumour, late diagnosis can cause the tumour to spread widely and enlarge the limits of the operation to resect the tumour.

CASE REPORT
An eight-year-old boy was admitted to our surgical unit with the complaint of a mass in the left cheek. The swelling started one year back and had a slow growth in this period. On examination, the mass was firm, 5×4cm, arising from the left parotid gland and was not fixed to the overlying skin or underlying structures (Fig.1).

Figure 1
Fig. 1: Clinical photograph showing classical (L) parotid tumour in a child

The regional lymph nodes were not palpable. Magnetic
resonance imaging (MRI) evaluation showed a well encapsulated tumour arising from the superficial lobe of left parotid gland with normal deep lobe.

**Figure 2**
Fig. 2: MRI scan showing an encapsulated mass arising from the left parotid gland.

Fine-needle aspiration cytology confirmed the diagnosis of sialoblastoma. A formal superficial parotidectomy was carried out with proper identification and preservation of the facial nerve. The postoperative course was uneventful and the boy was discharged five days after surgery.

Histopathology showed a lesion measuring 5×4cm and cut surfaces had a uniform lobular white appearance. Microscopically, the tumour consisted of well-circumscribed nests of basaloid cells separated by a vascular connective tissue stroma containing occasional ducts, consistent with diagnosis of sialoblastoma.

**DISCUSSION**
Salivary gland tumours are rare in childhood and account for 3-5% of all salivary gland tumours occurring in children. Congenital salivary epithelial tumours are characterized by some as medical curiosities, due to their rarity. In 1966, Vawter and Tefft reported two cases in neonates and used the term embryoma to describe them. Since then a variety of names have been used to describe the histologically similar or identical tumours. In 1988, Taylor suggested the term sialoblastoma to describe these lesions because it conveyed the dysontogenetic character and the site of the tumour in a single name. For epithelial neoplasms there is an increasing incidence over the first 20 years. Most are found in children who are between 10-20 years of age, the first decade being a relatively spared time. The majority of the cases of sialoblastoma described in literature belong to antenatal or perinatal age, but this tumour is extremely rare in first decade of life. As a result, the latter neoplasms pose problems in diagnosis and management.

Although rare, pediatric epithelial salivary gland neoplasms have several distinctive features. All have been found in the major salivary glands, mostly in the parotids, the submandibular gland being the more uncommon site. MRI scan is the investigation of choice for sialoblastoma. It manifests as a soft-tissue mass on MRI, which can also detect the involvement of the deep lobe and invasion to the adjacent muscles or bone. The distinction between benign and malignant sialoblastomas may not be as well defined as was thought originally. Review of the limited number of
reported cases reveals that local recurrence is the main concern. Of 24 reported cases, local recurrence has been documented in at least five patients. Some authors propose that sialoblastomas should be regarded as neither benign nor malignant but as one single disease with local infiltrative potential. Based on this concept, surgery alone remains the treatment of choice for sialoblastomas, provided that free margins are obtained. In our case, complete resection was carried out by superficial parotidectomy. Radiation therapy may be considered if the lesions are not completely resectable, but the adverse effects of radiotherapy may be undesirable for growing facial structures. Chemotherapy may be chosen when the tumour is not completely resectable or in cases of recurrent tumour.

From this case report and review of relevant literature, the following facts need special mention:

- Sialoblastoma, a rare tumour of salivary glands, should also be kept in the differential diagnosis of the childhood salivary gland tumours.
- These tumours are locally aggressive, rarely metastasize and wide surgical excision remains the treatment of choice for them, without irradiation or chemotherapy.

References
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