

Rare tumours of the hard palate

Sir,

We read the case report on soft tissue chondroma of the hard palate associated with a cleft with great interest.^[1] As mentioned by the authors correctly, this is a very rare presentation. We would like to share our experience with a tumour of the hard palate.

A 7-year-old boy presented to our outpatient department with a painless slowly growing swelling on the hard palate, the size little larger than a peanut, from last 3 months [Figure 1a]. The child gave a history of trauma to the palate by the back of the pencil at his classroom about 3 months back. A provisional diagnosis of a mucus retention cyst was made and a differential diagnosis of dermoid cyst was kept. Excision of the swelling with its short pedicle was done up to the depth of the periosteum of the hard palate under general anaesthesia. The residual defect was left open and allowed to heal secondarily. The specimen cut into half a revealed a yellowish fibrous core surrounded by a capsule [Figure 1b]. A histopathological study of the specimen stained with haematoxylin and eosin demonstrated characteristic solid sheets of monotonous round-to-polygonal cells with granular cytoplasm and small round nuclei and well circumscribed by stratified squamous epithelium [Figure 1c]. Immunohistochemical evaluation of the tumour cells demonstrated positivity for

vimentin [Figure 1d] but negativity for S100. The diagnosis was congenital granular cell tumour (CGCT). Follow up at 3 months did not reveal any recurrence [Figure 1e].

CGCT or congenital epulis of the newborn (CENB), also commonly referred to as gingival granular cell tumour and Neumann's tumour, was first described by Neumann in 1871.^[2] The tumour is postulated to originate from undifferentiated mesenchymal cell, fibroblasts, myofibroblasts, histiocytes, Schwann cells, or odontogenic epithelial cells. It is considered a benign mesenchymal tumour of unknown origin. While the exact incidence of CGCT is not known, fewer than 250 cases seem to have been reported worldwide till date. This tumour has been reported to occur ten times more frequently in females than males and three times more frequently in the maxilla than mandible.^[3] To the best of our knowledge, not a single incidence of CGCT in a male child has been reported from the Indian subcontinent. CGCT commonly occurs on the gingiva over the maxilla and mandibular alveolar ridge.^[2,3] Excision is curative and recurrence is not seen.

CGCT is different from granular cell tumour (GCT), also known as Abrikossoff tumour, which is an uncommon neoplasm, probably of neural origin derived from Schwann

cells in adults.^[4] CGCT is very similar to GCT histologically, but differs from it epidemiologically as well as in clinical behaviour. According to the published literature, CGCT is seen exclusively on neonatal gingivae, presenting most commonly at birth, having a marked predilection for females, while GCT is rare in the first decade of life, being most frequently diagnosed between the third and sixth decades of life, and affecting a wide variety of visceral and cutaneous sites, also having a predilection for the females. The difference also lies in the immunohistochemical properties of the two. The CGCT/CENB are S100 negative and Vimentin positive while GCT demonstrates immunoreactivity for vimentin, S-100 and neuron-specific enolase which correlates with its Schwann cell origin.^[5,6]

The diagnosis of CGCT should essentially be clinical, but usually poses some difficulty as seen in this case because of the low level of suspicion. The extreme rarity of presentation of CGCT on the hard palate in a 7 year male child in this case prompted us to share the report with our colleagues.

With more frequent reporting of these rare tumours, there arises a necessity to develop a comprehensive health registry with guidelines for research and clinical management. Furthermore, continued advances in molecular biology of such tumours may further elucidate the very distinct clinical behaviours of these tumours and ultimately provide better solutions to their treatment.

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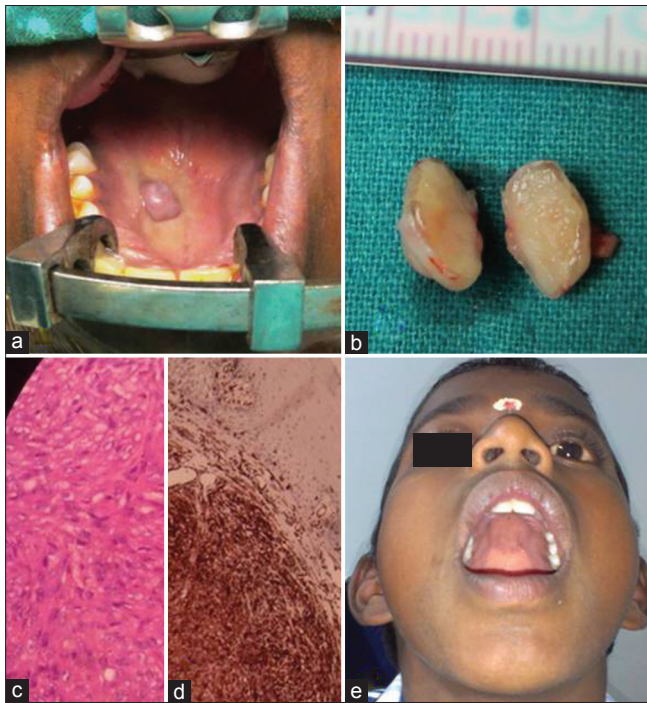


Figure 1: (a) Soft tissue swelling of the hard palate. (b) Cut specimen showing yellowish fibrous core. (c) Haematoxylin and eosin stained specimen showing solid sheets of monotonous round to-polygonal cells with granular cytoplasm and small round nuclei. (d) Immunohistochemical staining of the tumour cells demonstrating positivity for vimentin. (e) Follow up at 3 months did not reveal any recurrence

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| Quick Response Code:  | Website: www.ijps.org |
| | DOI: 10.4103/0970-0358.113740 |