Case Report of a Granular Cell Tumour in the Nasal Septum of a Child

J S G Hwang, H K Ang, C Y Aw

ABSTRACT

A six-year-old child with a nasal septal polyp presents with pain and nasal discharge. The diagnosis of a granular cell tumour was made histologically on excision of the polyp. This interesting case of an uncommon lesion presenting in a relatively rare paediatric age group is, as far as we know, the first report of a granular cell tumour involving the nasal septum.

Keywords: Granular cell tumour, nasal septum, child

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INTRODUCTION

While the differential diagnosis of a nasal mass in a child is wide ranging, granular cell tumours (GCTs) have not been described. GCTs are uncommon lesions occurring predominantly in the fourth to sixth decades of life[1]. Although they are most frequent in the head and neck, these have been localised largely to the tongue and the larynx. We document in this report a previously undescribed origin of a granular cell tumour of the nasal septum in a 6-year-old child.

CASE REPORT

A six-year-old Chinese girl presented at the Otolaryngology service with pain and discharge from the left nostril for a month, not relieved with topical creams. On clinical examination, a sessile nodule with a smooth surface was seen at the left anterior nasal septum. Endoscopy of the rest of the nasal cavity and the nasopharynx was normal. No other nodules were found in the head and neck region. Clinical history and examination was otherwise unremarkable. The polyp was subsequently excised under general anaesthesia. Macroscopic examination revealed a 6 x 5 x 3 mm mucosa-covered polyp. Histologically, this polyp was an unencapsulated submucosal tumour composed of cellular sheets and nests divided by slender fibrous connective tissue (Fig. 1). The polygonal to elongated cells exhibited ample pale eosinophilic granular cytoplasm with indistinct cytoplasmic borders, nuclei

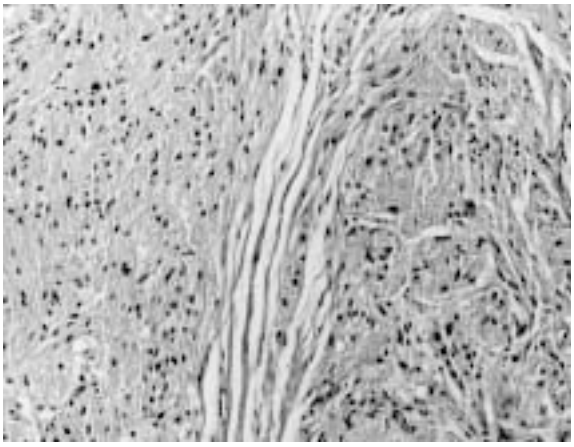

Fig.1 Sheets and nests of granular cells with small nuclei. (H&E stain, original magnification X100)

that were small and hyperchromatic in areas and more vesicular in others. Occasional eosinophilic cytoplasmic globules were seen with surrounding halo. Neither mitotic activity nor necrosis was evident. The tumour cells revealed positive immunohistochemical staining with S-100 protein and CD 68 and negative staining with muscle markers: desmin, myoglobin, myo-D1, HHF35. The deep surface of the tumour was circumscribed with nests of tumour cells forming a lobulated growth pattern. Closer to the surface, the tumour was more ill-defined with cells appearing more elongated. The tumour extended towards the overlying squamous and transitional type epithelium but no epithelial hyperplasia was demonstrated. Gomori methenamine silver stain for fungal organisms and Ziehl Nielsen stain for acid fast bacilli yielded negative results. The patient was well with no evidence of tumour recurrence six months after the excision.

DISCUSSION

A wide range of diagnoses enters into the differentials of a nasal mass, varying from inflammatory proliferations, infective lesions to benign and malignant neoplastic conditions. In a child, developmental aberrations have also to be considered. GCTs however, have not, as far as we are aware, been reported in the nasal septum. Although Abrikossoff is commonly credited with the description of GCT[2,3], Ordonez in his

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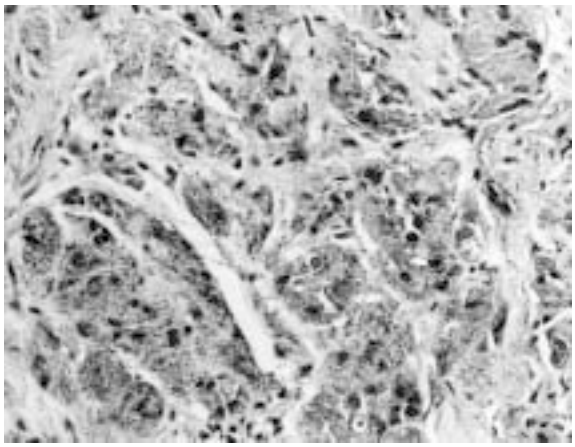

Fig.2 Positive cytoplasmic immunostaining with S-100 protein. (original magnification X200)

recent review referred to even earlier descriptions of this tumour by Virchow and Weber in 1854[4]. This uncommon lesion is interesting because of its characteristic histological appearance and uncertain histogenesis. Initially thought to be of skeletal muscle origin, it is currently believed to have a neural origin through its close association with nerves and the results of histochemical and electron microscopic findings[1,5].

GCT may occur at any age, from 11 months to 104 years[4] but most frequently in the fourth to the sixth decade[1]. Paediatric cases are relatively uncommon[4,6]. These tumours often arise in the head and neck region and in particular the tongue[1,7]. Other areas in which this tumour has been described include the skin, bronchus, stomach and bile duct[1]. In the head and neck region, GCT has also been found in the larynx[1,7,8,9], labial maxilla submucosa[10], soft palate, labial mucosa, uvula, oral floor, gingiva[7], orbit, lacrimal sac and nasolacrimal duct[3] and the parotid gland[11]. Multiplicity is present in 10 to 15% of cases[1,6,7].

This case is unusual in its location as well as in the age of presentation. Despite its common occurrence in the head and neck, this is, as far as we are aware, the first reported case of a granular cell tumour in the nasal septum. In Grotas' review of 17 paediatric cases of GCTs, five of them occurred in the head and neck, including the tongue[2]. The distribution in paediatric cases appears no different from those in adults[2].

Histologically, pseudoepitheliomatous hyperplasia, mimicking carcinoma, commonly seen in GCT was not present in this case. This lack of pseudoepitheliomatous hyperplasia has been affirmed by similar findings by Holland and Har-El in their paediatric cases of laryngeal GCT[8,9]. Given the young age of the patient, a diagnosis of congenital epulis was considered but this latter lesion is seen in the oral cavity of infants and is negative for S-100 protein.

Most GCTs are benign and treatment varies from conservative excision[7] to wide local excision[4]. The former is favoured by some as recurrence is seen in less than 7% of cases thus treated. Sabet reported a GCT that did not enlarge even after incomplete excision[3], Others however, note that those that recur had all been incompletely excised. Malignant forms are rare, occurring in 1 to 2%[1,6] Although most are characterised by a combination of necrosis, spindling, vesicular nuclei with prominent nucleoli, increased mitotic activity, high nuclear cytoplasmic ratio and pleomorphism[12], there are others that appear histologically bland but metastasize[1,6]. Clinically, malignancy is suggested by local recurrence, rapid growth, tumour size larger than 5 cm and metastases[1].

In summary, GCT although uncommon, should be considered in the differential diagnosis of a nasal septal mass.

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