Rare disease

Giant presentation of pleomorphic adenoma in major salivary gland

Manish Gupta,¹ Neena Chaudhary,² Monica Gupta³¹ENT, Gian Sagar Medical College, Chandigarh, India;²Vardhman Mahavir Medical College and Safdarjang Hospital, New Delhi, Delhi, India;³Department of Medicine, Government Medical College and Hospital, Chandigarh, India**Correspondence to** Dr Manish Gupta, manishgupta1217@gmail.com

Summary

Pleomorphic adenoma (PA) is the most common benign salivary gland tumour, accounting for as many as 80% of all such tumours. Although PA most commonly occurs in the parotid gland (85% cases), it may involve submandibular (8%), lingual and minor salivary glands. It is a benign tumour with a slow and continuous growth and without treatment can assume enormous dimensions. This paper describes a giant PA's arising in the submandibular gland and treated by complete surgical excision without sequel. In our case, a male patient presented with a tumour history of more than 4 years and weight of the resected mass was 2.24 kg. Giant PA's of the submandibular gland are very rare in medical literature.

BACKGROUND

Pleomorphic adenoma (PA) presents clinically as a painless, slow growing mass, usually varying from 2 cm to 6 cm when resected.¹ Cases of giant PA have been reported commonly in the parotid gland, presenting as an irregular multi-nodular mass that can weigh more than few kilograms.² However, giant PA's of the submandibular gland are very infrequent and there are just a handful of such case reports.^{3–5} Patient's lack of awareness and the fear of surgery seem to be the main reasons for these long standing lesions.

CASE PRESENTATION

A 60-year-old male presented in our out patient department with painless, gradual progressive swelling in left upper neck for 4 years. There was no history of difficulty in swallowing or respiratory discomfort. The patient gave no history of anorexia, weight loss, per-oral bleeding, and passage of stone or change in size of swelling with meals.

On external examination, a 12 cm × 8 cm × 8 cm sized (figure 1), non-tender, normothermic, firm, multi-lobulated, freely mobile swelling was seen involving left submandibular area vertically extending from the body of mandible superiorly to the hyoid and horizontally from angle of mandible to midline of the neck. The relevant cranial nerve examination was normal. Intraoral examination showed the submandibular duct opening to be normal and submandibular gland was ballottable and bimanually palpable.

INVESTIGATIONS

The computer-enhanced CT revealed 11.5 cm × 7.5 cm, heterogeneous, well-defined, marginated soft tissue mass in left submandibular area (figure 2) suggestive of PA of the submandibular gland.

TREATMENT

The mass was excised in-toto by standard submandibular approach. Perioperative period was uneventful. The resected specimen weighed 2.24 kg. Histology showed a PA with an intact capsule and no indication of malignancy.

OUTCOME AND FOLLOW-UP

There is no recurrence in 3 year follow-up.

DISCUSSION

PA is the most common salivary gland tumour. The main site of occurrence is the parotid gland, affecting patients of any age, more frequently between the fifth and sixth decades of life. Although uncommon, cases of giant PA, most of them involving the parotid gland have been reported. Buenting *et al* reviewed the ten largest PA's ever published in the English language literature, most of them before 1980s.¹ They found a mean tumour weight of 7.81 kg, nine out of ten cases occurred in female, with a mean age of 56.2 years. In 1989, Schultz-Coulon reviewed 31 cases of giant PA's of the parotid gland during the last 140 years.² He found a female predominance (64.5%), with an age range of 20–40 years old, and weight of the tumour between 1 and 27 kg. In three cases (10%), malignant areas were found within the tumours. In most of the cases described, it is considered that lack of information and negligence of the patients is relevant for the long course of an evident clinical mass.

Giant adenomas of submandibular gland are very rare. Belli *et al*, Schulz *et al* and Rognini *et al* each have reported isolated case reports of giant submandibular gland presenting as large neck swellings in the last decade.^{3–5} These neoplasms involve predominantly male sex (male/female ratio: 2/1) and usually occur between 20 and 40 years of age. Signs of malignant transformation may be observed

Figure 1 Clinical photo of left submandibular benign mass.

in the adenomatous epithelial component in a percentage ranging from 1% to 10% of cases.⁵

PA's typically arise as a slow growing, firm mass that is slightly compressible. Almost all are asymptomatic and they are usually brought to the attention of the physician when routine physical examination is performed or when the patient feels or sees a lump. Imaging studies alone can provide a presumptive diagnosis. CT scan findings of PA's are typically smooth, well marginated tumours. The attenuation values of the mass are usually homogeneous and higher than that of the surrounding gland. They typically show delayed contrast enhancement. When very large, they may develop a heterogeneous appearance with areas of necrosis, haemorrhage, cysts and calcification. Giant tumours commonly have a lobulated contour, which strongly suggests the diagnosis.

The PA's are typically well demarcated from the surrounding tissue by a fibrous capsule, which varies both in thickness and completeness. This capsule is a result of fibrosis of surrounding salivary parenchyma, which is compressed by the tumour and is referred to as 'false capsule'. If the fibrous capsule can be completely removed these tumours can be cured with surgery. The tumour also

has small protrusions (pseudopodia) that extend beyond the central mass, caused by variability in the growth rates of the various cell types. This factor contributes to recurrence rates as high as 50%, depending on the type of surgical intervention.

Microscopically, PA's are characterised by a myriad of morphological diversity. Epithelial cells are arranged in sheets and islands showing typical ductal structures and various epithelial and myoepithelial characteristics as spindle, clear, squamous, basaloid, plasmacytoid, oncocytic and sebaceous. The stroma characteristically is mixed with fibrous, chondroid, myxoid or hyaline aspects.

The incidence of malignant transformation in PA's, ranges from 1.9% to 23.3%.⁶ The risk increases in tumours with long time of evolution, recurrences, advanced age of the patient and location in a major salivary gland. Some authors postulated that the risk of malignant transformation increases from 1.6% in tumour with less than 5 years of evolution, to 9.5% for those presenting for more than 15 years.⁶ The classic clinical history of carcinoma ex-PA is of a slow growing mass for many years, with a recent fast growth associated with pain. In our patient there was no such history of sudden fast growth and

Figure 2 Computer-enhanced CT axial cuts of left submandibular benign mass.

there were no evidences of malignancy either clinically or microscopically.

Competing interests None.

Patient consent Obtained.

Learning points

- ▶ Giant PAs of submandibular gland are very rare.
- ▶ The tumour assumes large proportions due to neglect, lack of awareness and fear of surgery.
- ▶ PAs are known to recur and undergo malignant transformation.
- ▶ The tumour and its pseudocapsule must be fully resected to prevent recurrence.

REFERENCES

1. **Buenting JE**, Smith TL, Holmes DK. Giant pleomorphic adenoma of the parotid gland: case report and review of the literature. *Ear Nose Throat J* 1998;**77**:634,637–8,640.
2. **Schultz-Coulon HJ**. [Pleomorphic giant adenomas of the parotid gland]. *Laryngorhinootologie* 1989;**68**:445–9.
3. **Belli E**, Marini Balestra F, Longo B. Giant pleomorphic adenoma of the submandibular gland. Case report. *Minerva Stomatol* 2005;**54**:449–52.
4. **Schulz T**, Pauli R, Didzuneit-Sandhop B. [Cervical giant tumor in a 56-year-old female patient]. *HNO* 2010;**58**:159–61.
5. **Rognini F**, Bruschini L, Re M, *et al*. Giant neck neoplasm. Case report. *Acta Otorhinolaryngol Ital* 2003;**23**:388–90.
6. **Takahama A Jr**, da Cruz Perez DE, Magrin J, *et al*. Giant pleomorphic adenoma of the parotid gland. *Med Oral Patol Oral Cir Bucal* 2008;**13**:E58–60.

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