CASE REPORT

A huge pleomorphic adenoma of the submandibular salivary gland

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SUMMARY

The most common tumour of salivary gland is pleomorphic adenoma (PA). They are benign, painless, can grow into big tumours but usually do not affect nerves or lymph nodes. PA most commonly occurs in the parotid gland but it may involve submandibular, lingual and minor salivary glands also. They can attain giant proportions and weigh several kilograms. We report a giant PA arising in the submandibular gland and treated by complete surgical excision without any complication. A female patient presented with a tumour in the submandibular region and front of neck with a history of more than 18 years. The weight of the resected mass was 4.35 kg. Patient's fear of surgery and lack of awareness were the main reasons for her long-standing swelling. Such giant PAs of the submandibular gland are very rare in medical literature.

BACKGROUND

The purpose of this article is to emphasise that although the pleomorphic adenoma (PA) in parotid salivary gland is a well-reported entity where they can attain very large sizes, it is rare to find huge PA in submandibular salivary gland which can lead to unsightly swelling from the submandibular area to anterior part of chest.^{1,2}

We report a case, which to the best of the author's knowledge is one of the largest PA of the submandibular gland. That we can find such large tumours even in modern times is astonishing. This is probably the first huge PA of submandibular salivary gland in a woman which has got a MRI picture.

Figure 2 Preoperative picture—front view.

CASE PRESENTATION

A 47-year-old woman, resident of Allahabad, visited the Department of ENT and Head & Neck Surgery, SRN Hospital, Allahabad, and sought medical attention for a huge swelling of left submandibular area and in front of the neck since 18 years. It was slowly progressive in nature, had discharge and ulceration from the advancing tip of the swelling since 4 months. She was taking ayurvedic medicine from some local doctor for the same but size rapidly increased in the last 2 years. There was no history of difficulty in breathing or swallowing. The patient was otherwise healthy with no other significant medical history or finding except having marginally increased thyroid-stimulating hormone (TSH) level (5.79 U/mL) for which she was taking thyroxine hormone 50 µg daily.

On examination, approximately 32 cm × 22 cm × 14 cm swelling was present in left submandibular area and in front of neck (figures 1 and 2), extending from left mastoid tip to submental area on the right side, originating from left submandibular and submental area, hanging over the chest

Figure 1 Preoperative picture of the tumour.

Figure 3 MRI of the tumour.

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Figure 4 MRI of the tumour.

and reaching upto the xiphisternum, firm to hard in consistency, multinodular and non-tender.

Advancing tip of swelling had multiple ulceration (may be due to ischaemic necrosis) with foul smelling discharge with the different stages of healing and scarring. Overlying skin was hyperpigmented. Oral cavity was within normal limit and movement of the tumour mass could be elicited and palpated bimanually intraorally. Submandibular lymph nodes could not be palpated due to the tumour mass. No other nodes were palpable.

INVESTIGATIONS

All the routine investigation were within the normal limit except TSH marginally raised.

Incisional biopsy was done in the ENT operation theatre. Histopathological examination of the tissue showed features of PA with fibrosis.

MRI: contrast-enhanced MRI shows huge heterogeneously enhancing heterointense mass lesion in the anterior neck possibly arising from left submandibular space demonstrating lobulated solid cystic areas and necrotic areas with displacement of adjacent structures but no significant infiltration/erosion of bone (figures 3 and 4).

Figure 5 Immediate postoperative stitch line.

Figure 6 Size of the resected tumour.

DIFFERENTIAL DIAGNOSIS

Some common differential diagnosis can be thyroid swelling, submandibular lymph gland pathology, tumour of the tail of parotid gland, Hodgkin's disease, non-Hodgkin's lymphoma, metastatic disease and dermoid.

TREATMENT

Complete resection of the tumour was done along with the submandibular salivary gland as the tumour was arising from the

Figure 7 High-power view showing myoepithelial cells in chondromyxoid stroma.

Figure 8 Postoperative scar after 3 months.

gland, with preservation of the majority of the overlying healthy skin and the marginal mandibular nerve. Primary closure of the wound was done (figures 5 and 6). Weight of resected tumour was 4.35 kg.

The patient recovered well and was discharged satisfactorily. Postoperative period was uneventful and the incision wound healed well.

Postoperative histopathology of the resected tumour showed partially encapsulated tissue consisting of dual population of

Figure 9 Postoperative scar after 3 months.

Table 1 An overview of different case reports of pleomorphic adenoma (PA) of submandibular gland.							
Name of the journal	Year	Site of origin/sex, age of patient	Size/volume/weight of the tumour	Modality of investigation (CT/ MRI)	Histological diagnosis	Treatment	Outcome
BMJ Case Reports	2017	Left SSG female, 47	32 cm×22 cm×14 cm, 9856 cm ³ ; 4.35 kg	MRI	Huge pleomorphic adenoma of the left SSG	Excision	Improved with no complication
Pakistan Journal of Otolaryngology, 30: 90–92	2014	Right SSG male, 70	32 cm×38 cm×29 cm, 35 264 cm ³ ; 7.6 kg	FNAC, CT scan	Carcinoma ex- PA of right submandibular gland	Excision	Sent for further management for carcinoma
Scholar Journal of Applied Medical Sciences (SJAMS), 1: 432–434	2013	Right SSG male, 52	4.6 cm×2.8 12.88 cm ² ; weight—NA	USG NECK and CT scan—axial and coronal views, HPE	PA of right submandibular gland	Right submandibulectomy	Improved
Cranioaxillary Trauma and Reconstruction, 5: 185–188	2012	Left SSG female, 20	16 cm×15 cm×12 cm, 2880 cm ³ ; weight —NA	OPG and CT	PA of left submandibular gland	En-bloc resection of the tumour	Improved
BMJ Case Reports	2011	Left SSG male, 60	11.5 cm×7.5 cm×7.5 cm; 647 cm ³ ; 2.24 kg	CECT	PA of submandibular salivary gland	Excision	Improved
Kulak Burun Bogaz Ihtis Derg, 20(4): 210–213	2010	Right SSG female, 7	3 cm×2 cm×2 cm, 12 cm ³ ; weight —NA	FNAC, contrast enhance MRI	PA of right submandibular gland with potential of malignant transformation	Right total submandibular gland excision	Improved

CECT, contrast-enhanced CT scan; FNAC, fine needle aspiration cytology; HPE, histopathological examination; NA, not available; OPG, orthopantomogram; SSG, submandibular salivary gland; USG, ultrasonogram.

predominantly plasmocytoid myoepithelial cells arranged in sheets along with few clusters of oval to round epithelial cells. The cells population is benign looking and is suspended in abundant chondromyxoid stroma. No significant atypia/pleomorphism/mitosis/necrosis noted (figure 7). Histology is consistent with PA (submandibular gland).

OUTCOME AND FOLLOW-UP

Postoperative period was uneventful and the patient was discharged satisfactorily. A follow-up period of 5 months has been uneventful (figures 8 and 9).

DISCUSSION

According to available literature, PA is the most common salivary gland tumour and is mostly found in parotid gland although large tumours have been described in submandibular salivary gland also. Sporadic case reports of giant submandibular gland adenomas have been reported by Belli *et al*, Shultz *et al* and Rognini *et al*.^{3 4} Although they have reported such tumours in predominantly male sex, ours is a female patient.

The tumours are smooth, painless and do not produce any symptoms till they attain huge size when they can show pressure symptoms. CT scan and MRI are the gold standard radiological tools.⁵ The tumour is usually well demarcated from the surrounding tissue by a pseudocapsule due to compression of the surrounding parenchyma and fibrosis. The tumour has small protrusions (pseudopodia) which can be a cause of recurrence. When they attain giant proportions they can develop heterogeneous appearance due to areas of haemorrhage, necrosis, cysts and calcification.

Histomorphologically, it is characterised by a variegated architecture comprising epithelial elements admixed with a mucoid, myxoid, fibrohyaline or chondroid stroma.⁶

Table 1 shows some recent case reports of PA.

Most of the reported cases have used CT and ultrasound as modalities of investigation. There are few cases where MRI has been done, but the MRI picture is not given.

Our case is probably the only case which has MRI picture of the giant PA of submandibular salivary gland in a female patient. As is evident from the chart, the weight of PAs of submandibular salivary gland varied from few grams to 2–3 kg. Our case weighed 4.35 kg with a volume of 9856 cm³, while the volume of other cases reported varied from 12 cm³ to 2880 cm³. The

Learning points

- ▶ Giant pleomorphic adenoma (PA) of submandibular gland is very rare, which is astonishing to find one in modern times.
- ▶ PA can attain huge dimensions if left untreated due to the lack of awareness or fear of surgery.
- ▶ Even huge adenomas can be successfully surgically excised without any morbidity and sequelae.
- ▶ It is the most common tumour of salivary gland and are known to undergo malignant transformation.

case where the reported volume was 35 264 cm³ with a weight of 7.6 kg turned out to be carcinoma ex-PA.²

Therefore, our case is the largest reported PA of the submandibular salivary gland in a female patient with MRI picture and histopathological diagnosis, treated by surgery without complications.

Contributors AR has operated the case with the help of all coauthors. MS and PS helped in assisting the surgery while SS helped in researching the topic, planning and writing of the case report. MS also outlined the importance of reporting this case as many doctors of our time would never see such a big pleomorphic adenoma.

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