CASE REPORT

Metastatic Pleomorphic Adenoma in the infratemporal fossa and neck following total parotidectomy after 30 years

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SUMMARY

Metastasising pleomorphic adenoma is rare and may occur years after surgical excision of a pleomorphic adenoma (PA). We present a 61-year-old woman with a right infratemporal PA with metastases to the cervical lymph nodes after 30 years following a total parotidectomy. She was treated successfully with a resection of the tumour with combined neck and mandibulotomy approach along with postoperative radiotherapy given subsequently.

INTRODUCTION

Pleomorphic adenoma (PA) is the most common benign tumour affecting the salivary gland. Occasionally, it may undergo a malignant transformation to give rise to a carcinoma ex pleomphic adenoma (CEPA) or carcinosarcoma that is very aggressive in nature. On rare occasions, it metastasizes to surrounding structures. This usually occurs following an incomplete resection of a previous PA and it's called enigmatically as a metastatic pleomorphic adenoma (MPA) because of its benign histological appearance and metastatic capability.[1]

CASE PRESENTATION

A 61-year-old woman presented with a four-month history of throat discomfort and right-sided painless neck swelling. There was no dysphagia, breathlessness, hoarseness or constitutional symptoms. Her past medical history included a right total parotidectomy performed 30 years earlier for a PA based on patient's best recollection of her condition. Her co-morbidities included diabetes, hypertension and dyslipidaemia. Intraoral examination revealed a hard right soft palate bulge seen extending to the lateral pharyngeal wall with a smooth mucosal lining. Neck examination revealed a previous surgical scar, a vague right angle of mandible mass (2.5 x 2.5cm) and level II and III cervical lymphadenopathy. Flexible nasoendoscopy revealed medialization of right lateral pharyngeal wall from the nasopharynx to oropharynx. Fossa of Rosenmuller was normal. Facial nerve function was normal.

Computed tomography (CT) scan showed an irregular mixed solid cystic heterogeneously enhanced mass seen in the right superior tonsillar region (4.1 x 3.5 x 5.0cm) with an enlarged right parapharyngeal lymph node (2.6 x 2.0 x 2.0cm). This caused an asymmetry of the oropharynx and bulging over the right soft palate region. There was also an enhancing mass at the right angle of mandible (2.6 x 2.0 x 2.3cm) (Figure 1) and a few enlarged right upper cervical level II lymph nodes. CT scan of thorax was clear.

Biopsy of the soft palate revealed a PA of the minor salivary glands. The initial impression was a recurrent PA of the parotid gland to the infratemporal space with malignant transformation.

She underwent a resection of the tumour via a combined neck and mandibulotomy approach and a right selective neck dissection (SND) (level II and III). Operative findings were a large multilobulated submucosal well-encapsulated tumour occupying the right oropharynx extending superiorly to the infratemporal fossa and laterally to parapharyngeal space (Figure 1C). The tumour was abuttng the soft palate with no evidence of invasion or erosion to the hard palate or pterygoid plates. The tumour had a good plane of dissection except near the base of her skull where it was adherent to the internal carotid artery and a small amount was left behind. The mass in the infratemporal fossa and angle of mandible were in very close proximity (Figure 1D). There was no semblance of normal salivary gland tissue. A right submandibulectomy and a tracheostomy were also performed.

Histopathological examination revealed that the right infratemporal mass and angle of mandible mass were recurrent PA with metastases in two of 11 cervical lymph nodes. One lymph node (level III) showed extranodal extension. Tumour was partly encapsulated and composed of epithelial, myoepithelial and mesenchymal elements. The myoepithelial cells were arranged in sheets, interconnecting thin trabeculae, spindly and stellate-looking, disposed in lattice-like fashion within a chondromyxoid stroma (Figure 2A). The cervical lymph node showed multinodularity and features similar to the primary tumour (Figure 2B). The epithelial-myoepithelial cells exhibited mild nuclear pleomorphism with fine chromatin and no overt atypia. Mitosis was occasionally seen with no obvious

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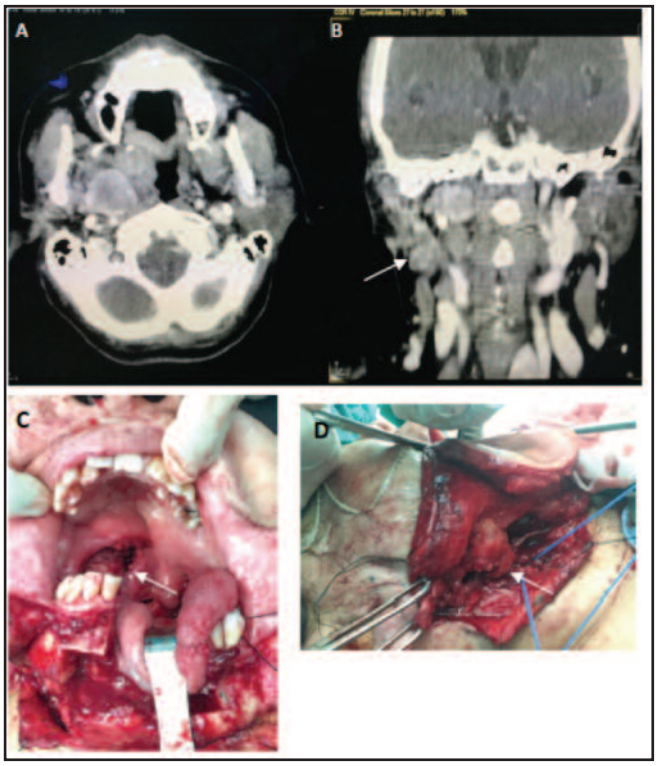

Fig. 1: A. Axial CT scan with contrast showing a right tonsillar and parapharyngeal mass occupying the infratemporal fossa. B. Coronal CT scan with contrast showing the right parapharyngeal mass close to base of skull and mass at the angle of mandible (arrow) with metastases to the right upper cervical lymph nodes. C. Intraoperative view of a multilobulated submucosal well encapsulated tumour occupying the right oropharynx extending superiorly to the infratemporal fossa and laterally to parapharyngeal space (arrow). D. Intraoperative view of the tumour at the right angle of mandible (arrow).

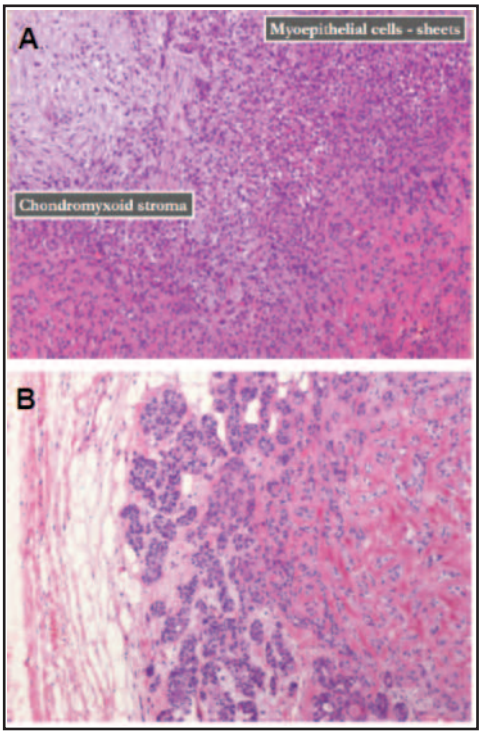

Fig. 2: A. Axial CT scan with contrast showing a right tonsillar and parapharyngeal mass occupying the infratemporal fossa. B. Coronal CT scan with contrast showing the right parapharyngeal mass close to base of skull and mass at the angle of mandible (arrow) with metastases to the right upper cervical lymph nodes. C. Intraoperative view of a multilobulated submucosal well encapsulated tumour occupying the right oropharynx extending superiorly to the infratemporal fossa and laterally to parapharyngeal space (arrow). D. Intraoperative view of the tumour at the right angle of mandible (arrow).

lymphovascular invasion or tumour necrosis. There was no malignancy in the submandibular gland.

Postoperative period was unremarkable. Patient had normal facial nerve function and was decannulated after two weeks. She completed adjuvant radiotherapy and at the six-month follow-up visit, was noted to have no recurrence.

DISCUSSION

MPAs are extremely rare, accounting for 1% of all malignant PAs and is described by the WHO as a "histologically benign PA that inexplicably manifests local or distant metastases". It is thought to be an unrecognized low grade malignancy or a continuum from PA to CEPA. In a recent systemic review, Knight and Ratnasingham, documented only 81 cases in 73 years.[1]

MPA is thought to be a result of tumour spillage during enucleation or incomplete clearance of a previous PA.[2] There is usually a long latency period between initial treatment of PA and the onset of a MPA. This is usually around 14.9 years with a range of 3 to 52 years.[1,3] According to Nouraei et al.'s case-series, MPA is markedly different from the onset of a recurrent PA which is about five years from initial treatment of a primary PA.[2]

PA usually affects the parotid gland 80% of the time and commonly presents as a painless parotid mass between the 4th to 6th decades of life.[1] MPA however, presents as a head and/or neck mass 40% of the time.[2] They could also present with other symptoms due to the propensity of MPAs to metastasize. Most common metastases are to the bones and lungs but other sites have been reported to include kidneys, liver, skin and brain.[1,3]

MPA has similar histologic features to PA where there is an epithelial -myoepithelial component and a metaplastic mesenchymal component consisting of fibrous and chondromyxoid features.[4] There are usually minimal features of malignancy such as cellular atypia, increased mitotic activity, infiltrative pattern of spread and areas of necrosis.[2] Differential diagnosis could include primary bone tumour, chordoma, myoepithelioma, inflammatory mass, chondroid syringoma, adenoid cystic carcinoma or CEPA.

The diagnosis of recurrent PA of the parotid gland versus a minor salivary gland PA was challenging. We considered a few factors prior to arriving to our diagnosis. Firstly, it was the previous history of a parotid PA. Secondly was that the incidence of multifocal PA, which has been reported to occur in 68% of patients in recurrences, meant that soft palate involvement was possible.[5] Thirdly, a new minor salivary gland PA would be very unusual given its low incidence as a primary compared to a parotid PA.[1] And fourthly, the intraoperative findings showed a well-encapsulated tumour that was abuttng the soft palate with no evidence of invasion or erosion to the hard palate or pterygoid plates.

Total surgical resection or metastectomy is the mainstay of treatment with the addition of a SND and postoperative radiotherapy in the case of extranodal spread.[2] This treatment confers a disease free survival rate of between 50 to 63%.[1,2] However, this value may be underestimated, as long-term follow-up has not been discussed in many case reports.

In conclusion, MPA is an intriguing entity with no histological signs to predict its occurrence. The only risk factor is previous history of a PA. Due to its indolent and asymptomatic nature of growth, it may be wise to have an extended period of follow-up for patients who have had surgical removal of their PA.

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