

Carcinosarcoma Of The Parotid: Parapharyngeal Extension With Facial Nerve Palsy and Airway Obstruction

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Citation

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Abstract

Objective: We report an extremely rare case of carcinosarcoma of the parotid with parapharyngeal extension presenting with facial nerve palsy and airway obstruction. The patient succumbed to the disease despite surgery performed. Here, we discuss the peculiarities of the histopathology, immunohistochemistry, radiology and the surgical aspects of this condition.

Case report: An elderly lady with a rapidly progressing large parapharyngeal mass presented with facial nerve palsy and impending airway compromise. Fine needle aspiration cytology showed presence of sarcomatous components. Distant metastasis was excluded. Subsequently, surgical removal of the mass was performed and carcinosarcoma of the parotid gland was diagnosed. The epithelial component was predominately composed of squamous cell carcinoma whereas chondrosarcoma was the predominant stromal component.

ABBREVIATIONS

VES Tan
SW Yeo
BR Lee
AMM Shahizon

CASE HISTORY

A 60 year-old Indian lady presented to our clinic with a 2-months history of right infraauricular swelling. It was rapidly increasing in size resulting in worsening dysphagia, trimus and a muffled voice. She had considerable weight loss due to her diminishing food intake and poor appetite. She has a past medical history of hypertension and diabetes mellitus.

At presentation, she was not in any form of airway embarrassment. There is a right infraauricular swelling measuring 5cm x 5 cm, firm in consistency and was fixed to the underlying structures. The overlying skin was normal. Clinically, there was no evidence of cervical lymphadenopathy and the facial nerve was intact. Examination of the oral cavity revealed a large submucous swelling located at the right posterior oropharynx, with both the uvula and the tonsil medialised.

A fine needle aspiration cytology (FNAC) from the right

infraauricular swelling showed cytological features consistent with undifferentiated malignancy, most likely a soft tissue sarcoma. Based on contrast-enhanced computer tomography (CECT) scan, this showed a large heterogenous mass with its epicenter at the right parapharyngeal space extending to the parotid space measuring about 5cm x 8 x 7cm in size. A diagnosis of parapharyngeal tumour was made. Magnetic resonance imaging (MRI) showed that the mass is of low to intermediate signal on T1WI and of moderate to high signal on T2WI with thick rim enhancement. A more solid component is seen at its medial aspect. Features are suggestive of an aggressive right parotid gland tumour without any intracranial extension.

The patient was thoroughly investigated to exclude any distant metastases. A routine chest X-ray showed a small oval opacity was noted in the upper zone of the left lung consistent with a focus of consolidation. A CT-guided biopsy later showed only necrotic tissue with no evidence of malignancy with immunohistochemistry (IHC) test which was negative for cytokeratin, desmin and thyroid transcription factor 1 (TTF1). She also underwent a barium swallow, which was unremarkable. Ultrasonography of the abdomen and Technetium Tc ⁹⁹ methylene diphosphonate (MDP) whole body bone scan also excluded any

intraabdominal and bony metastasis respectively.

During the course of admission, she developed right lower motor neuron facial nerve palsy (House Brackmann grade IV). Following that, she developed progressive respiratory distress. In view of the impending airway compromise, tracheostomy was performed while waiting for her operative date. She was planned for excision of the tumour via a transcervical-transparotid approach. Intraoperatively, the superficial lobe of the parotid and the facial nerve were not involved but the deep lobe was enlarged and the tumor was encasing the lower 4 cranial nerves at the base of skull superiorly. A superficial parotidectomy was performed preserving all branches of the facial nerve. The deep lobe of the parotid and parapharyngeal portion was mobilized and removed completely apart from its attachment superiorly at the skull base.

She recovered uneventfully and was discharged well. She was scheduled for a course of adjuvant radiotherapy due to the aggressive nature of the tumor and the positive superior margin. While awaiting adjuvant radiotherapy, she was readmitted for dysphagia. Examination revealed ipsilateral lower cranial nerves (IX, X, XI, XII) palsy in addition to the prior facial nerve palsy. The mass has also grown in size as compared to the immediate post-operative period. She was scheduled for 10 cycles of radiotherapy. Unfortunately, she died after completion of the 5th cycle of radiotherapy.

Figure 1

Figure 1: The carcinomatous component was comprised of squamous cell carcinoma arranged in solid nest. The squamous cell carcinoma displayed polygonal cells with distinct cytoplasmic borders, intercellular bridges, pleomorphic, hyperchromatic, vesicular nuclei and prominent nucleoli. (haematoxylin-eosin, X100)

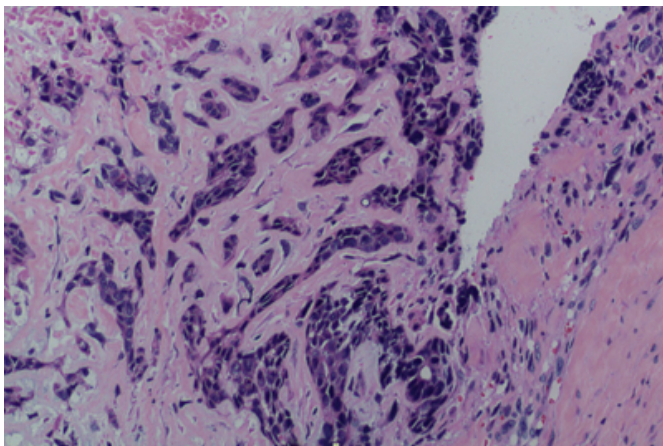


Figure 2

Figure 2: (A) The sarcomatous component was comprised of spindle shaped tumour cells arranged in sheets and displayed markedly abundant eosinophilic cytoplasm with pleomorphic hyperchromatic, vesicular nuclei. (B) In areas, the sarcomatous component displayed chondroid differentiation with tumour cells within the lacunae in a background of hyalinized intervening chondrocytic stroma.

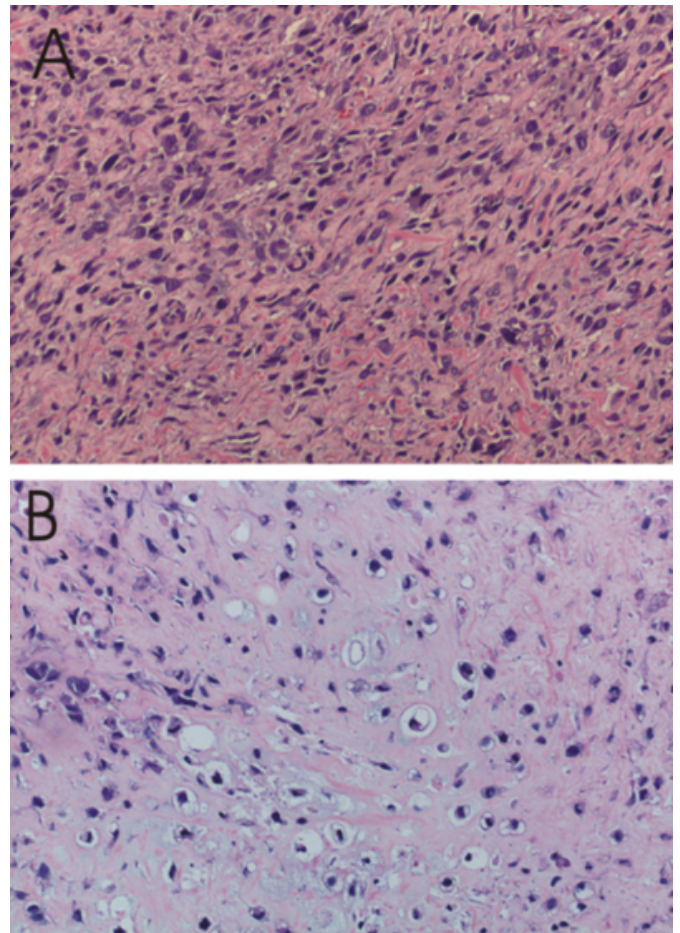


Figure 3

Figure 3: CECT showed a rim enhancing mass in the parotid space with focus of calcification (black arrow) at its inferior aspect

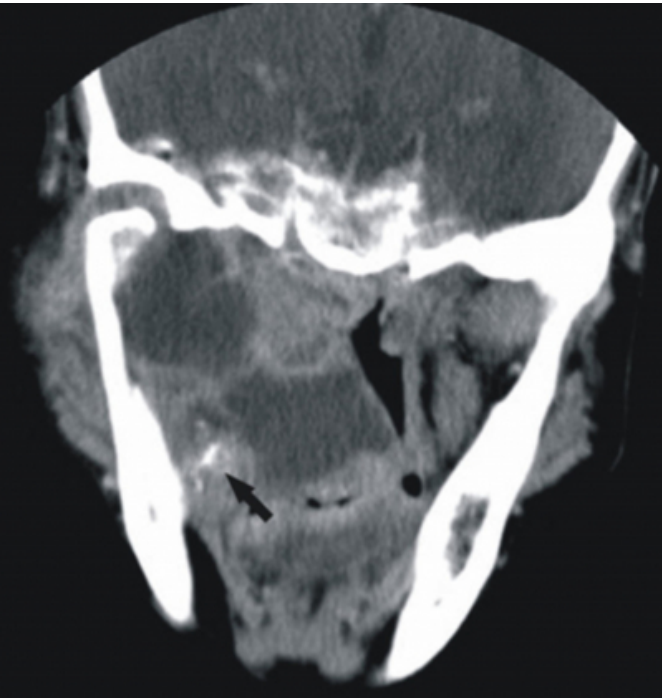


Figure 4

Figure 4: MRI T1WI post gadolinium showing a thick rim enhancing mass with necrotic centre at the parotid space. It is heterogeneous with solid areas displaying intermediate signal intensity on T2WI. The mass has caused anterolateral displacement of the right mandible. MRI clearly delineated the effacement of the parapharyngeal fat as well as medialisation of the right pharyngeal wall. (A) axial section. (B) coronal section.

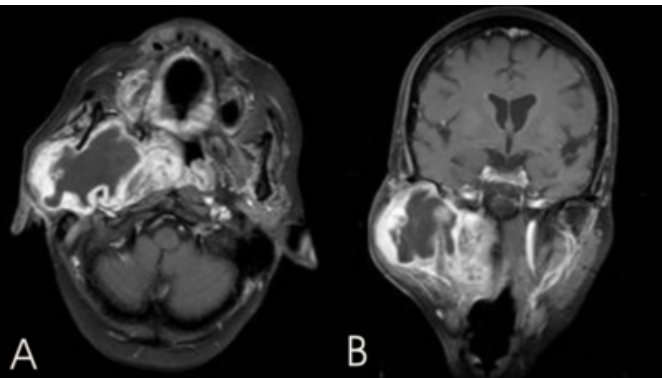


Figure 5

Table 1: Immunohistochemical studies of different component of the tumour

Antibodies	Carcinoma	Sarcoma	PSA Epithelium/ myoepithelium	PSA Mesenchyme
Cytokeratin AE1/3	+	-	+	-
EMA	+	-	+	-
Vimentin	-	+	+	+
S100 protein	Focal+	-	+	-
Smooth Muscle Actin	Focal+	-	+	-
Desmin	-	-	-	-

Plus sign indicates positive, minus sign indicates negative, PSA indicates pleomorphic adenoma, EMA indicates Epithelial Membrane Antigen

DISCUSSION

Carcinosarcoma, also called true malignant mixed tumor (TMMT) of the salivary gland is among the rarest of malignant tumours of the salivary gland, with an occurrence of less than 0.1%¹. Majority arise in the parotid gland, followed by submandibular gland and the palate². In the same review, the prevalence was almost the same in both sexes with the mean age of presentation at 58 years old (range of 14-87 years).

In contradistinction to most case reports in the literature, the tumor with its parapharyngeal extension in this case has grown so large that the patient presented with complications of dysphagia, facial nerve palsy and respiratory distress requiring surgical intervention. The facial nerve palsy testified to its aggressive and infiltrative nature of the tumour. Although most primary parapharyngeal tumors arise from the deep lobe of the parotid gland or minor salivary gland tissue in this area, only 2% of all parotid tumours present as parapharyngeal space tumours³, commonest being the pleomorphic adenoma of the parotid. Therefore, carcinosarcoma was thought to arise from a pleomorphic adenoma, albeit consisting of both carcinomatous and sarcomatous elements², hence the name. In fact, malignant mixed tumor was thought to comprise 3 different clinicopathologic entities: carcinoma ex-pleomorphic adenoma, carcinosarcoma and metastazing mixed tumour⁴. While in carcinoma ex-pleomorphic adenoma, a secondary carcinoma arises from a usually long-standing benign pleomorphic adenoma, a carcinosarcoma can arise within a preexisting pleomorphic adenoma or de novo from a salivary gland⁵. The carcinosarcoma ex-pleomorphic adenoma is commoner than the de novo subgroup⁶. Both the epithelial

and mesenchymal components are malignant in carcinosarcoma, in contrast to the exclusively epithelial malignancy seen in a carcinoma ex-pleomorphic adenoma ⁷.

In the rare group of metastizing mixed tumour, both the primary salivary gland and its metastatic lesion are histologically benign and identical ⁴. As illustrated by the rapid regrowth in this case, carcinosarcoma is aggressive regardless of its origin; however, prognosis remains worse for those that arise in the salivary glands than those arising from the surface epithelium ⁸.

Most of the carcinosarcomas are often grossly infiltrative with poorly defined margins and the cut surface shows solid gray tumour with cystic change, haemorrhage and calcification ^{2,3}. Carcinosarcoma is a biphasic neoplasm, having both the carcinomatous and sarcomatous components. The sarcomatous component predominates in most carcinosarcoma ^{2,4}, probably explaining the reason why the FNAC in this case only demonstrated sarcoma. The sarcomatous components most often manifested chondrosarcoma ⁹, followed by osteosarcoma, fibrosarcoma ² while the carcinomatous component manifested undifferentiated carcinoma, SCC or high-grade ductal adenocarcinoma ^{1,2}. The solid nest of the carcinomatous component showed peripheral arrangement that in areas merges with the sarcomatous component. In this case, the epithelial component was predominately composed of SCC (Figure 1) whereas the stromal component was composed of tissue resembling chondrosarcoma (Figure 2). In some areas, coexisting pleomorphic salivary adenoma was present characterized by cluster of benign epithelial and myoepithelial cells entrapped in abundant hyalinized and cartilaginous matrix. The epithelial elements resembling ductal cells or myoepithelial cells were displayed in duct formations, irregular tubules, strands and sheets of cells. These elements were dispersed within a mesenchyme-like background of loose myxoid tissue containing islands of chondroid. The superior surgical margin was involved and has given rise to a rapid regrowth.

Our immunohistochemistry findings (Table 1) were similar to 2 other reported cases ^{4,10}. This proves that some carcinosarcomas do not show evidence of myoepithelial cell origin or differentiation. Therefore, it is hypothesized that the primitive mesenchymal cells, being able to differentiate along diverse directions, may have given rise to the appearance of different kinds of sarcoma, and in our case, chondrosarcoma.

In terms of radiological investigation, CECT is the initial imaging modality of choice in cases of head and neck pathology. With the advent of multislice CT, it provides fast and reliable reconstructed images. The superiority of CT compared to MRI is the detection of calcification (Fig 3). The presence of calcification is non specific, most commonly seen in pleomorphic adenoma, others include mucoepidermoid carcinomas, vascular malformation and sarcoma ¹¹.

MRI which has superior soft tissue resolution and the ability to detect perineural extension has been used as an adjunct to assess the full extent of the disease. Malignant lesion can be deduced if the lesion display low to intermediate signal on T2WI as it reflects high mitotic ratio and the high nuclear to cytoplasmic ratios ¹². The presence of thick rim enhancement with necrotic centre as well as its signal intensity suggests the aggressiveness of the tumour (Fig. 4), as seen in our case.

A thorough effort was made to exclude metastasis as it would alter our management and her prognosis significantly. Prognosis remains poor for this tumor as some studies have shown zero 5-year survival rates ¹³. The current practice advocates radical surgery with radiation therapy and lymph node dissection for palpable disease ³, although in some instances neck dissection was performed even for N0 neck ¹³. However the role of chemotherapy is still unclear.

CONCLUSION

In conclusion, carcinosarcoma of the parotid gland is a rare tumor which exhibits an aggressive biological behaviour with resultant poor prognosis. Late presentation with parapharyngeal and base of skull extension adds to the complexity of management in this patient. Although the proper management of this condition has not been clearly defined, early radical removal of the tumor must be emphasized.

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