Carcinosarcoma of the parotid gland: A case report and short review of literature
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Citation

Abstract

Background:
Carcinosarcomas are very rare true malignant mixed tumors of salivary gland. Majority is seen in the parotid glands. Chondrosarcoma and osteosarcoma elements are the most common sarcomatous element whereas moderately or a poorly differentiated ductal adenocarcinoma is the most common carcinomatous element seen.

Case details:
We report a parotid lesion in a 47-year-old female, composed of ductal type adenocarcinoma admixed with chondrosarcomatous and malignant spindle cell elements.

Conclusion:
The case is of interest because of its rarity and the initial diagnosis of benign tumor by Fine needle aspiration cytology. This case is a carcinosarcoma, which combines both features of ductal type adenocarcinoma and malignant spindle cell tumors along with chondrosarcomatous elements. In addition, a short review of literature is presented.

INTRODUCTION
Malignant mixed tumour of salivary gland comprises 3 different clinical entities-carcinoma arising in benign mixed tumour (carcinoma ex pleomorphic adenoma), carcinosarcoma and metastasizing mixed malignant tumour. It constitutes approximately 12% of malignant salivary gland tumors, 6.2% of all mixed tumors and 3.6% of all salivary gland neoplasms. Carcinosarcoma is a very rare true malignant mixed tumor of salivary gland, which is an aggressive, high-grade neoplasm with multiple episodes of recurrences and metastasis.

CASE DETAILS
A 47-year-old female presented with history of swelling in right parotid area for 25 years, which was increasing in size for past 1 month. Previous history of operation was not available. The swelling was 5.0x5.0 cm, hard, immobile and slightly tender. The patient had facial palsy at presentation.

Computerized tomography (CT) scan of the swelling showed enhancing hyper-dense lesion in parotid gland with popcorn calcification (Fig.1).
Figure 1
Figure 1: CECT showing enhancing hyperdense lesion in the right parotid gland with popcorn calcification

Figure 2
Figure 2: An area showing chondromatous differentiation. (H&E; X100)

Fine needle aspiration cytology (FNAC) of the parotid gland done previously in another institution was reported as pleomorphic adenoma. Right total parotidectomy was done and the tumor was found to be fixed posteriorly to mastoid and to infiltrate the facial nerve trunk and was resected in piecemeal.

PATHOLOGY

The tumor was received in multiple soft tissue bits, measuring 7.5x7.0x4.0 cm and weighing 40 grams. The external surface of the lesion was nodular and at places well circumscribed. The cut surface showed normal salivary gland along with a gray white lesion, measuring 3.5x3.0x3.0 cm, which had a gritty sensation while taking sections. Regional lymph nodes were not involved. Multiple histological sections showed a lesion comprised of a bimodal population of both epithelial and mesenchymal component. The predominant mesenchymal component was the malignant spindle cells, arranged in interlacing fascicles along with a chondrosarcomatous component (Fig.2).

The other component intermingled with these was the malignant ductal component (Fig.3, 4).

Figure 3
Figure 3: Admixture of glandular and mesenchymal component. (H&E; X100)
Both the components showed pleomorphism, hyperchromasia of nuclei and presence of mitotic figures. However no areas of necrosis identified. The lesion showed entrapped nerve twigs and was also seen infiltrating the normal salivary gland. Multiple sections showed no evidence of pleomorphic adenoma.

DISCUSSION

Kirklin et al of the Mayo Clinic first described mixed carcinoma and sarcoma of the parotid gland in 1951. King in 1967 first used the term true malignant mixed tumor (carcinosarcoma), which means a tumor consisting of both carcinomatous and sarcomatous elements. There are 3 distinct histological types of malignant mixed tumor: carcinoma ex pleomorphic adenoma; metastasizing mixed tumors; and carcinosarcomas, which are true malignant mixed tumors. Carcinosarcoma of the salivary gland is an extremely rare tumor. By definition, it is a biphasic tumor composed of malignant epithelial and malignant mesenchymal elements. In most tumors epithelial and mesenchymal components are admixed with each other, as seen in the present case. Gnepp summarized 43 cases of carcinosarcoma of the salivary glands published in the literature. The majority of these tumors (33%) are from parotid glands followed by submandibular glands (19%) and palate (14%). There is no sex predominance. The mean age at presentation is 58 years with a range of 14 – 87 years. Many cases are seen to arise in a pre-existing benign mixed tumor.

Few articles have described the cytologic features of carcinosarcoma. In the literature, the most common epithelial components are ductal adenocarcinoma or squamous cell carcinoma, whereas the most common mesenchymal components are chondrosarcoma. Other sarcomas that have been reported include fibrosarcoma, leiomyosarcoma, osteosarcoma, malignant fibrous histiocytoma and rarely liposarcoma, rhabdomyosarcoma or the combination of these sarcomas and also myoepithelial malignant proliferation. Also recently the combination of epithelial- myoepithelial carcinoma and pleomorphic sarcoma and large-cell neuroendocrine carcinoma with rhabdomyosarcoma has been described (Table-1).

In our case, the malignant mesenchymal components are undifferentiated spindle cell elements and chondrosarcomatous elements and these are intermingled with malignant ductal component. The differential diagnosis always includes benign mixed tumor and primary versus metastatic sarcomas. Multiple sections taken in the current tumor however did not show any evidence of pleomorphic adenoma.

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tumors of the salivary glands are controversial. According to some authors, carcinosarcoma and pleomorphic adenoma of the salivary gland may share a common precursor, possibly a myoepithelial cell. On the other hand, it has been shown that in some carcinosarcomas both components arise de novo. Immunohistochemically cytokeratin and EMA are epithelial markers whereas vimentin and s-100 are sarcomatous mesenchymal markers. Kwon and Gu, based on their study where both components did not show any immunoreactivity for smooth muscle actin, a marker for myoepithelial cells hypothesized that the primitive mesenchymal cells may have given rise to different kinds of sarcoma. Gotte et al also favored the hypothesis of common stem cell monoclonal origin of carcinosarcoma that could be the myoepithelial cell and an inactivated tumor suppressor gene on chromosome 17 other than p53.

Carcinosarcoma is an aggressive, high-grade neoplasm and requires aggressive therapy, including radical surgery, with and without radiation therapy and chemotherapy. Carcinosarcoma of the parotid arising after irradiation to the resection site of a pleomorphic adenoma is also described. Recurrent disease develops in approximately two thirds of patients and metastases in about half. The routes of metastases are hematogenous and lymphatic. Lung is described as the most common site of metastases. Cervical and hilar lymph node metastases are also common. There is a good correlation between stage and local extension of the tumor and prognosis. Facial nerve palsy and grade of malignancy are important prognostic factors.

CONCLUSION

The case reported in this article is a carcinosarcoma, which combines both features of ductal type adenocarcinoma and malignant spindle cell tumors along with chondrosarcomatous elements. It is reported because of its rarity. Post-operatively the patient received radiotherapy and is doing well after a follow-up of two years.

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