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Mesenchymal Chondrosarcoma of the parotid. A case report

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ABSTRACT

Mesenchymal Chondrosarcomas of the parotid are extremely rare tumours . They are broadly classified under extra skeletal form of mesenchymal chondrosarcomas and account for less than 1% of all sarcomas.(1) We report a rare case of a parotid mesenchymal chondrosarcoma and discuss the pathogenesis, clinical presentation, and management of such a rare tumour.

CASE REPORT

An 81 year-old woman presented to the otolaryngology department with a history of swelling around the right anterior part of the ear. The swelling had been slowly progressing over 8 months. She did not report any facial pain or neck pains. Physical examination of the neck revealed a 5 × 3.5-cm mobile oval- shaped and non-pulsatile parotid mass in the right infra auricular region. (Fig. 1)

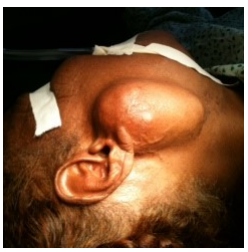

Figure 1

There were no skin changes above the mass and cranial nerve examination was normal. Examination of the neck revealed no neck nodes.

Under the diagnosis of benign parotid tumour, a total parotidectomy was performed with preservation of facial nerve branches. (Fig.2)

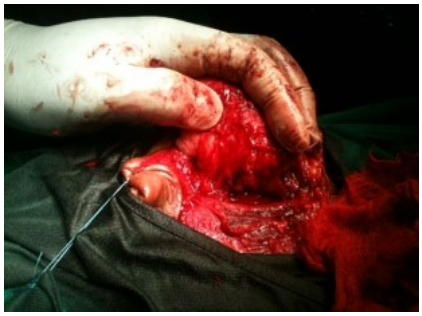

Figure 2

The tumour mass was removed with adequate margins. The post operative outcome was uneventful and the patient recovered fully. The facial nerve was intact. (Fig. 3)

Histologic features were consistent with a mesenchymal chondrosarcoma. Entire examination of the tumour disclosed no component of pleomorphic adenoma. A post operative chest x ray was done which was normal. The patient also underwent radiotherapy post operatively. There has been no evidence of recurrence 9 months after the operation.

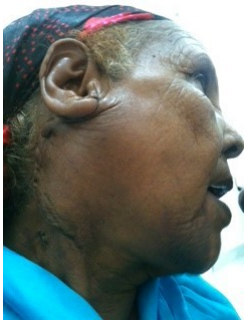

Figure 3

DISCUSSION

Mesenchymal Chondrosarcoma is a rare, malignant type of cancer of the cartilage first described by Lichtenstein and Bernstein in 1959.(2) About 66 % of Mesenchymal Chondrosarcomas occur in bone, while the remaining cases occur in extra skeletal sites.(1) Mesenchymal Chondrosarcomas originate from chondroblasts, which are cartilage precursor cells that have failed to develop into mature chondrocytes. Immunohistochemistry may play a key role in distinguishing mesenchymal chondrosarcomas from other small round cell tumours. The chondrosarcomas may stain for vimentin, muscle specific actin, S100,Proliferating cell nuclear antigen and Ki 67. (3) They are broadly classified under malignant mixed tumors. Malignant mixed tumors are classified into three distinct histologic types – 1. Carcinoma ex-pleomorphic adenoma 2. Benign metastasizing pleomorphic adenoma 3. True malignant mixed tumor (carcinosarcoma). Carcinoma ex-pleomorphic adenoma represents about 99% of these cases. (4) A true malignant mixed tumor is a very rare tumor that is composed of both malignant epithelial and malignant mesenchymal elements.

The commonest malignant epithelial component is squamous cell carcinoma while the commonest malignant mesenchymal component is chondrosarcoma. A true malignant mixed tumor represents about 0.04% to 0.16% of salivary gland tumours and 0.4% of malignant salivary gland neoplasms. (5) About 65% of cases occur in the parotid gland.

The occurrence of mesenchymal chondrosarcomas may be slightly higher in females than in males, and no identifiable risk factors have been found for the development of this tumour.

It is believed that Mesenchymal chondrosarcomas generally occur in younger patients in extraskeletal locations and in bone in older patients. Therefore this particular case is a very rare case indeed.

Initial work up for this condition includes a full history and physical examination. If the condition is initially suspected, a chest x ray is included in the initial workup. Definitive management includes surgical resection of the parotid tumour followed by adjuvant chemotherapy and radiotherapy. Patients with mesenchymal chondrosarcoma do not generally have metastatic disease at presentation although there is a paucity of literature due to the rarity of the tumor. (6)

Due to the possibility of recurrence, follow up is recommended every three months for the first one year, with scans of the original area. Most patients are known to relapse late in the disease. Patients are usually left to determine a follow up schedule for scans with which both they and their specialist are comfortable. In conclusion, the parotid gland can be a site of occurrence of de novo primary chondrosarcoma.

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