Mesenchymal Chondrosarcoma of the Parotid Gland

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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. It is characterized by a multinodular architecture, abundant myxoid matrix, and malignant chondroblast like cells arranged in cords. The tumor is an entity from chondrosarcomas of bones, commonly found in the soft tissues of the lower extremities at 80%. There are very limited reports of this tumor in the head and neck, especially in the parotid gland. We report a rare case of a parotid mesenchymal chondrosarcoma in a 55 years old man and discuss the pathogenesis, clinical presentation and management of such a rare tumour.

Key words: Chondrosarcoma, mesenchymal, parotid

Introduction:
Mesenchymal Chondrosarcoma is a rare, malignant type of cancer of the cartilage first described by Lichtenstein and Bernstein in 1959.¹ About 66% of Mesenchymal Chondrosarcomas occur in bone, while the remaining cases occur in extra skeletal sites.² 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 metastatizing pleomorphic adenoma 3. True malignant mixed tumor (carcino-sarcoma). Carcinoma ex-pleomorphic adenoma represents about 99% of these cases.⁴

Case Report:
A 55 years old man presented to Matin Specialized Hospital, Bogra with a history of...
swelling around the right inferior part of the ear. The swelling had been slowly progressing over 12 months. He did not report any facial pain, neck pains or any facial weakness. Physical examination of the neck revealed a 6 × 4.5-cm slightly mobile oval-shaped and nonpulsatile parotid mass in the right infra auricular region. (Fig.-1) It was firm in consistency except in its central part which was cystic in consistency with skin discolouration and cranial nerve examination was normal. Examination of the neck revealed no neck nodes. FNAC showed benign disease. Under the diagnosis of benign parotid tumour, a superficial parotidectomy was planned but after exploration the tumour involved both the lobes so total parotidectomy was performed with preservation of facial nerve branches. (Fig.2) The tumour mass was removed with adequate margins. (Fig.-3) The post operative outcome was uneventful and the patient recovered fully. (Fig.-4) The facial nerve was intact. Histologic features were consistent with a mesenchymal chondrosarcoma. (Fig-5). The patient also underwent radiotherapy post operatively. There has been no evidence of recurrence 9 months after the operation.
**Discussion:**
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. 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 extra-skeletal 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. 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.

**References:**