

Malignant Chondroid Syringoma: A Case Report with Review of Literature.

Veenita Yogi*, OP Singh, and Vivek Tiwari.

Department of Radiation, Oncology, Gandhi Medical College, Bhopal, Madhya Pradesh, India.

Case Report

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*For Correspondence

HIG 315, Bagmugalia Katara
Hills, Bhopal, Madhya Pradesh
462043, India.
Tel.No.0755-4050380

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ABSTRACT

Among malignant tumour of the skin, sweat gland origin tumours are rare. Malignant chondroid syringoma probably is one of the rarest subtypes and poorly understood. It lacks distinctive clinical features, often delaying initial diagnosis leads late therapeutic management. The wide surgical excision, adjuvant radiation therapy with or without chemotherapy as well as patient education is critical in facilitating long term survival.

INTRODUCTION

Adnexal carcinomas are rarely seen tumour. Sebaceous gland carcinoma accounts for less than 1% of all skin cancer. Sweat gland carcinoma is very rare, accounts for fewer than 0.01% of malignant epithelial neoplasm of the skin. Its eccrine variant (chondroid syringoma) malignancy is very rare and appears to behave in an aggressive manner. It is associated with poor prognosis. Malignant chondroid syringoma is more commonly seen among female population. It occurs most often in head & neck, trunk and extremities. Tumour appears as a firm intradermal or subcutaneous, painless and slowly growing nodule with varying histopathological appearance from benign to malignant. The wide surgical excision and adjuvant radiotherapy has been the mainstay of treatment. Adjuvant chemotherapy with significant response has yet not been defined.

Case Report

A 67-years-old male, presented with the c/o multiple lobulated painless lesions over left side of face, which often became painful. It was progressive in nature and within two months presented as 10X12 cm size post auricular lobulated mass. The lesion was mobile, nontender and firm in consistency with involved overlying skin. The patient underwent surgical excision of the lesion of the same temporo-parietal region three yrs back at the local hospital. After one year of surgery, lesion reappeared. Then he was operated again with surgical reconstruction (spiral PMMC), and again lesion reappeared in the form of multiple lobulated mass around skin graft [Figure 1]. Microscopically it is characterized by nodules of cells composed of epithelial and mesenchymal like cells separated by connective tissue. The cells are seen in cords and acinar arrangements, with a rich myxomatous and cartilaginous areas, scattered mitosis, variable pleomorphism with presence of necrosis [Figure 2]. Immunohistochemistry revealed positive staining for cytokeratin, S-100 protein and calponin, SMA and P53 were negative. Haemogram and biochemistry profiles were reported to be within normal limit. MRI revealed heterogeneous enhanced, soft tissue signal intensity mass in left external auditory canal and glenoid fossa of temporal bone extending into infratemporal region. Condylar process of mandible was displaced inferiorly and anteriorly by the mass. Inferiorly mass was extended upto parotid region & anteriorly upto pre styloid compartment along with medial aspect of the ramus of mandible and infiltrating lateral pterygoid muscle. Temporal muscles were not properly visualized because infiltrated by mass. Mass appears to be extending upto mandibular canal. Posteriorly it was eroding the posterior cortex in external auditory canal and extending into mastoid air-cells. Medially mass was extended into middle ear cavity and encasing ear ossicles. No obvious cervical adenopathy was noticed [Figure 3]. CT scan of chest showed bilateral nodular opacities in lungs involving the basal and apical segment of left lower lobe, anterior segment of right upper and medial segment of right middle lobe. Minimal

fibrotic changes in basal segment of right lower lobe with minimal pleural thickening on right side. Subcentimeter sized mediastinal lymph nodes in pretracheal and subcarinal region. Subcarinal nodes show calcification [Figure 4].Based on the histopathology, immunohistochemistry, local recurrence, invasion to surrounding structures and pulmonary metastasis the diagnosis of malignant chondroid syringoma was confirmed. After confirming the diagnosis patient was given one course of chemotherapy consisting of vincristine, adriamycin, cyclophosphamide & dacarbazine for five days. Patient responded well and lung opacities were disappeared, but he didn't turn up for next course of chemotherapy. After two months he again came with progressive growth which did not respond with the above regimen. Then nanotaxel 300mg with cisplatin 50mg for three days was given. There was marked regression of lesion, but after 15 days he developed brain infarct and died.

Figure 1: Chondroid syringoma-patient's photograph, Legend- Multiple lobulated mass around skin graft



Figure 2: Chondroid syringoma-histopathology slide, Legend –Chondroid tissue and epithelial elements H and E x 400

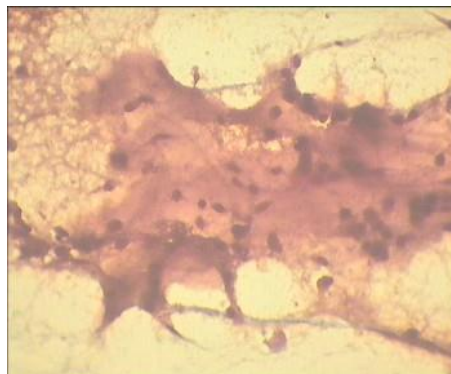


Figure 3: Chondroid syringoma- MRI of tumour, Legend- MRI of tumour (transverse and coronal) showing extension of disease.

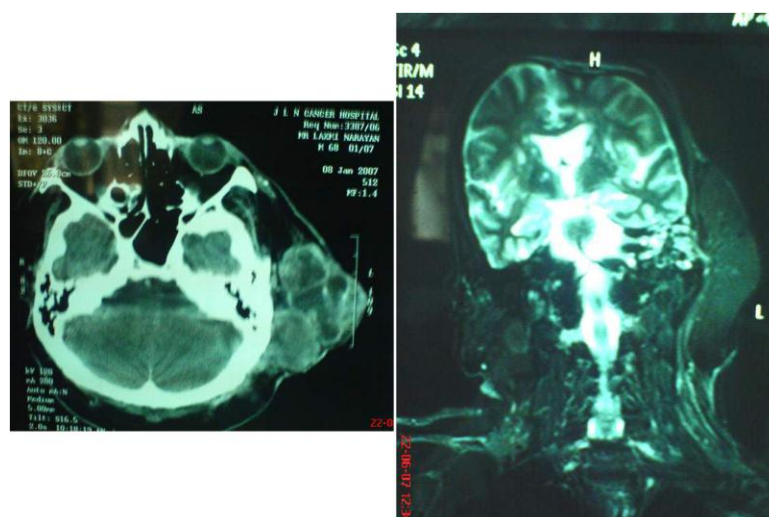
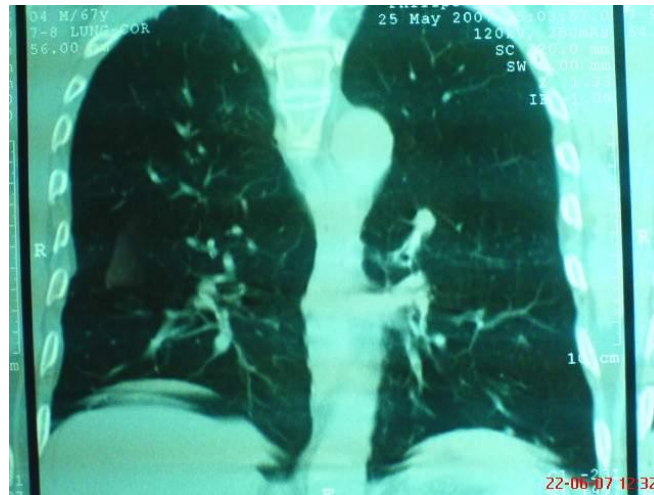


Figure 4: Chondroid syringoma-CT scan chest, Legend – CT scan showing bilateral nodular opacities.



DISCUSSION

Malignancy of adnexal structure, eccrine (sweat gland) carcinoma is rare accounting for fewer than 0.01% of malignant epithelial neoplasm of the skin [4]. Eccrine (sweat gland) carcinoma arises most often in the skin of the head and neck, trunk and extremities and usually present as slow growing painless papules or nodules [2]. It is usually diagnosed in between 50 and 70 years of age [3]. Chondroid syringoma is occurs more common in women. Histologically chondroid syringoma resemble carcinomas of the breast, bronchus and kidney and thus may difficult to differentiate from cutaneous metastasis. Its histologic appearance vary greatly and it may even have benign appearance [4,5].

Microscopically, the chondroid syringoma comprises of nests of atypical cells with few mitotic cells, which partly formed gland like structures and areas of myxoid degeneration. Gland like structure composed of epithelial and mesenchymal like cells, myxomatous and cartilaginous areas [5,6].

On immunohistochemistry chondroid syringoma is positive for alcian blue staining and they did not stain after they were digested with hyaluronidase, were prominent in the matrix among tumour cells for cytokeratin(AE1+AE3), S-100 protein, neuron specific enolase and glial fibrillary acidic protein(GFAP). GFAP is characteristic of malignant chondroid syringoma, 39% of cases with this staining positive have metastatic lesions and 22% died of this malignant tumour. Intraluminal cells are CEA positive. The stromal cells are cytokeratin negative and sporadically positive for vimentin. Positive chondroid areas are S-100 protein and vimentin positive [6,7].

Chondroid syringoma behave more aggressively than do other skin carcinomas. According to Wick MR et al in a series of 14 patients with this tumour who underwent surgical excision with negative margin, 11 had at least one local recurrence and 5 had recurrence in regional nodes or distant sites. One patient died of an uncontrolled local relapse and four patients died of distant metastases 2 months to 10 years after diagnosis. In contrast to the benign counterpart, which is common in the head and neck region, the malignant variety occurs predominantly on the trunk & extremities [3,8].

For eccrine carcinomas wide excision of the primary lesion is accepted as the standard treatment in patients with no lymphadenopathy at diagnosis. Postoperative radiation therapy to the surgical bed with 2 to 3-cm margins is indicated, particularly for eccrine carcinoma because of the high incidence of local recurrence after surgery alone. The role of elective treatment of regional lymphadenopathy is controversial. Treatment for patients with palpable lymphadenopathy consists of wide local excision, lymph node dissection and postoperative irradiation of the surgical bed. In radiation therapy for adnexal carcinomas, the target volume consist of the primary tumour bed with 2 to 3-cm margins and the draining lymphatics. Radiation is administered, preferably with electrons in 2-Gy daily fractions. For adnexal carcinomas, a dose of 60 Gy is generally administered in the postoperative setting.[9]

The role of adjuvant treatment is not very clear since there was no difference in survival. However for cytoreduction which can provoke growth and role of radiotherapy may be enhanced and to prevent metastasis chemotherapy drugs before or after surgical excision can be tried followed with radiotherapy to tumour bed. Our patient responded very well with chemotherapy but he died because of brain infarct.

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