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CARCINOSARCOMA OF THE OROPHARYNX : A CASE REPORT AND REVIEW OF LITERATURE

Oncology

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KEYWORDS:

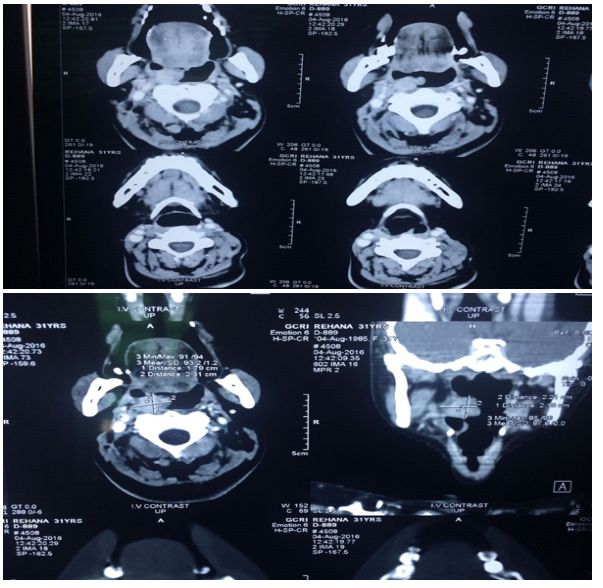
Introduction

Carcinosarcoma is a malignant mixed tumor that contains both carcinomatous and sarcomatous elements. Carcinosarcoma with typical malignant features are extremely rare in the oral cavity, it has been reported in oropharynx, tongue, gingiva, floor of mouth and paranasal sinuses. The carcinomatous component is usually an adenocarcinoma. The sarcomatous component is made up of either undifferentiated spindle cells or pleomorphic cells that are capable of mesenchymal differentiation into bone (osteoid sarcoma), cartilage (chondroid sarcoma), or muscle (rhabdoid sarcoma). Carcinosarcomas are characterized by high rates of recurrence, metastatic spread and mortality. We describe here a case of recurrent carcinosarcoma of the oropharynx with review of literature.

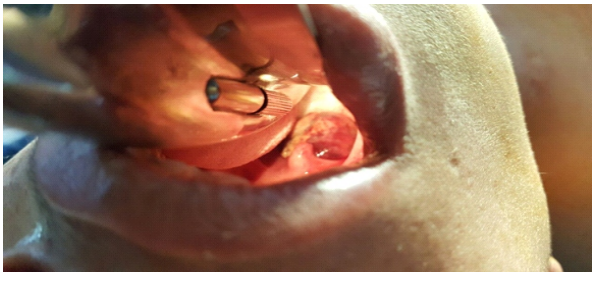
Case Report

A 31-year old women presented in our oncosurgical department with chief complaints of throat pain, muffled voice and progressive dysphagia for last three months with previous history of carcinosarcoma of oropharynx which had been resected ten years back. She had weight loss due to poor appetite and diminished food intake. Earlier, the tumor was resected with the right lateral pharyngotomy approach, wide local excision and bilateral level II-IV neck dissection was done and she received post operative radiotherapy with total dose of 60 Gy/30#. The patient was disease free for ten years and had been on regular follow up until she had similar complaints again. At presentation, she was not in any form of airway embarrassment. Clinically, there was no evidence of cervical lymphadenopathy. Examination of the oral cavity revealed a polypoidal mass with smooth surface located on the right lateral oropharyngeal wall with an extension to the soft palate and uvula pushed towards left side. Right tonsillar fossa appears free. Direct laryngoscopy was done which confirmed the clinical findings and biopsy was taken. Both vallecula, epiglottis and posterior pharyngeal wall was normal. The biopsy report showed high grade malignant tumor, and IHC favors of carcinosarcoma positive for vimentin and cytokeratin AE1. Contrast enhanced computed tomography scan suggest homogeneously enhancing polypoidal mass of size 23 x 21 mm, arising from the right posterolateral wall of oropharynx with associated luminal narrowing. No evidence of lymphadenopathy noted on both sides of the neck. In view of patient past history, and current clinicopathological findings, a diagnosis of recurrent carcinosarcoma involving right oropharynx was made and the patient was thoroughly investigated to exclude any distant metastases. A routine chest X-ray showed bilateral lung fields clear. She also underwent a barium swallow, which was unremarkable. Ultrasonography of the abdomen and Technetium Tc99 methylene diphosphonate (MDP) whole body bone scan also excluded any intra-abdominal and bony metastasis respectively. The patient was subsequently planned for surgery, a tracheostomy was done for ventilation during surgery and to avoid tumor manipulation by endotracheal tube during induction. A lip split incision was given and median mandibulotomy done to gain access to the tumor. Wide local excision with adequate margins done and split skin graft placed at the excision site. She recovered uneventfully and was discharged on nasogastric tube feeding. The patient was on regular follow up for last 3 months and she has started accepting orally.

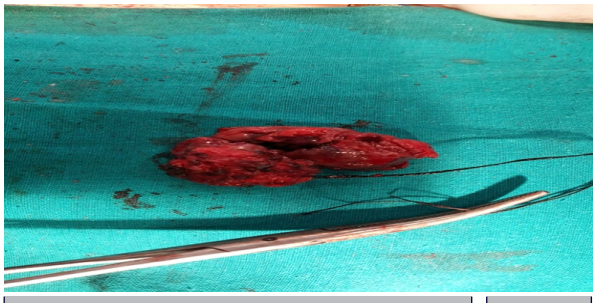
CECT IMAGES:



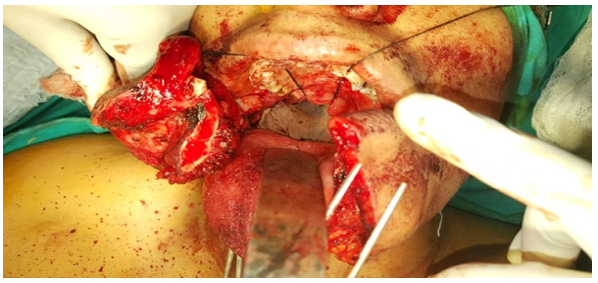
INTRAOPERATION IMAGES:



laryngoscopic view of the tumor



Specimen of excised carcinosarcoma



post operative image after excision of tumor with skin graft

DISCUSSION

In the head and neck, the most common sites of primary carcinosarcomas are the larynx, particularly the glottis, and the oral cavity. Less common sites include the oropharynx, hypopharynx, nasal cavity, and sinuses.[1] Carcinosarcoma is a biphasic neoplasm, having both the carcinomatous and sarcomatous components. The sarcomatous component predominates in most carcinosarcoma. The sarcomatous components most often manifested chondrosarcoma, followed by osteosarcoma, fibrosarcoma while the carcinomatous component manifested undifferentiated carcinoma, SCC or high-grade ductal adenocarcinoma.

Two antithetical hypotheses have been advanced to explain the histogenesis of carcinosarcomas: one proposes a multiclonal origin arising from two or more stem cells; the other a monoclonal origin from a single totipotential stem cell that differentiates in separate epithelial and mesenchymal directions[1]. Recent immunohistochemical and chromosomal analyses appear to have settled this argument in favor of the monoclonal hypothesis.[1,2]. The current case shows positive IHC markers for vimentin and cytokeratin AE1 that favours for carcinosarcoma.

The primary treatment modality for carcinosarcoma is surgical excision along with neck dissection for palpable neck nodes.[6,7]. Radiotherapy as a primarily treatment is considered to be ineffective,[6–8] although adjuvant irradiation may be beneficial in patients who have positive surgical margins or who have positive nodal metastasis [6]. The role of chemotherapy is still unclear.

The prognosis for patients with carcinosarcoma is similar to that for patients with conventional squamous cell carcinoma. Survival depends largely on the location and stage of the tumor; patients with glottic spindle cell carcinoma have a relatively good prognosis, while those with a spindle cell carcinoma of the oral cavity or paranasal sinuses have a poor prognosis. Reported 5-year survival rates range between 63 and 94%; overall mortality is 30 to 34%.[3] Factors although reported controversial yet influence the long term survival includes: The gross morphology and size of the tumor, the differentiation of the carcinoma component, the depth of invasion, and prior treatment with radiation.[12,11.]

CONCLUSION

The case is presented to highlight the following facts: Carcinosarcoma are rare aggressive tumours while orophayrnx subsite involvement is rare in the head and neck region. Surgical excision with adjuvant radiotherapy is the preferred method of treatment. Our present case showed a recurrence in the oropharynx ten years after surgery, therefore careful follow-up is necessary because of the poor prognosis in many cases.

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