

Carcinosarcoma: A rare case report of a recurrent mass in the neck region

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Abstract

Carcinosarcoma, a biphasic malignant mixed tumor, is an extremely rare neoplasm with >1% incidence. This aggressive malignancy is characterized by the presence of two components admixed with each other, i.e., the epithelial component and the mesenchymal component arising from a monoclonal/multiclonal origin or *de novo*. Most patients usually present between 60 and 65 years of age with no sex predilection. The authors present a case of carcinosarcoma arising as a mass in the neck region of a 14-year-old male. The case is being presented for its rarity of occurrence in the younger age group.

Keywords: Carcinosarcoma, head and neck neoplasm, mixed tumor, recurrent mass, supraclavicular region, young adult

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INTRODUCTION

Carcinosarcoma, an aggressive biphasic malignant neoplasm with both epithelial and mesenchymal components is extremely rare tumor that accounts for about >1% of all malignancies.^[1,2] Way back to history, during early 1989, the total cases documented were up to 20 cases.^[3] In 1993, a slight numerical rise of about 60 cases were documented.^[4] Recent statistical data revised till date revealed up to 73 cases.^[5] This modest rise in the occurrence of this malignancy over decades has raised the curiosity among researchers to explore this tumor in detail and identify the best treatment modalities.

By definition, carcinosarcoma is composed of two components, i.e., epithelial and mesenchymal components

giving a biphasic appearance.^[6] It was first described in 1951 by Kirklin *et al.*^[7] and labeled as “carcinosarcoma” by Virchow in 1864.^[8,9] King Jr. in 1976 was the first to use the term “True Malignant Mixed Tumor” for this malignancy.^[10] Thereafter, various authors described a wide range of terminologies for this baffling malignancy as spindle cell carcinoma, pseudosarcoma and squamous cell carcinoma with pseudosarcoma.^[9] Carcinosarcoma are usually seen in elderly patients (6th–7th decade), although a wide range from 14 to 87 years does exist, with a slight male predilection. It is thought to arise from larynx, hypopharynx, esophagus, trachea, nasal cavity and oral mucosa.^[11-13] Irrespective of its origin, this aggressive tumor has a poor prognosis. Recurrent mass in the neck region in younger age groups are uncommon, and this article presents one such rare case.

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CASE REPORT

An Indian 14-year-old male was addressed to the Department of Head and Neck Surgery, Mahatma Gandhi Cancer Hospital, South Maharashtra, India, with a chief complaint of a tender mass on the right side of the neck of 2 years duration. The patient's medical history revealed a previous open biopsy, which was undertaken in another institute, details of which were untraceable except for the biopsy report that revealed carcinosarcoma. The present mass reported on December 10, 2019, measured approximately about 10 cm × 8 cm in size extending from the right mid-neck at the level of C3 vertebrae region till the right supraclavicular region and was associated with pain. The photographs of the recurrent mass were taken after due consent from the patient for documentation [Figure 1]. On clinical examination, a well defined, lobulated, tender mass that was hard in consistency was felt on palpation. The mass was mobile and not fixed to the deeper structures and planes of the neck region. The overlying skin was tense and taut. Level III and IV lymph nodes were palpable. There were no other systemic symptoms involved or associated.

On contrast enhanced computed tomography (CT) imaging of the right side of the neck, a 7 cm (crainocaudal) × 6.2 cm (anteroposterior) × 4.7 cm (transverse) lobulated heterogeneously enhancing lesion on the subclavicular/clavicular regions, abutting the right sternocleidomastoid and right paraspinal muscles with sharp emarginated borders was seen. An intraluminal hypodense area of 2.3–1.0 cm was noticed within the right subclavian vein suggestive of a tumor thrombus. The lymph nodes were assessed on the long axis, in which one node in the right posterior triangle area revealed 1.5 cm enlargement, indicative of metastasis. On further evaluation, adjacent anatomical structures such as nasopharynx, torus tubarius,

oropharyngeal airway, hyoid bone, epiglottis, thyroid gland, esophagus, larynx, tongue, muscles of mastication and all the major salivary glands/salivary ducts were normal. CT thorax and ultrasonography abdomen were done to rule out any systemic disease or metastasis. Clinical differential diagnosis of malignant tumor of the neck was considered. The patient, being minor, signed consent from his parents was taken before the surgery. They were also informed in brief about the biological behavior of the tumor and the type of surgery planned based on the tumor characteristics.

Sternotomy followed by the opening of the thorax, the right superior major venous system and the “innominate vein” was dissected and prepared. The venous angle was dissected, and an emergency ligation of the subclavian vein was planned before its resection and repair. *En bloc* resection of the tumor was planned and performed that involved a circumferential incision around the tumor skin margin and part of the sternocleidomastoid muscle, Level II, III, IV and lymph nodes were dissected along with the tumor mass. Part of the clavicle, which was involved by the tumor mass, was also resected *en bloc*. The subclavian vein that showed an intraluminal hypodense area on CT was clamped and partly resected, followed by its mobilization and repair of the remaining proximal and distal stumps. Hemostasis was achieved. The thorax was closed meticulously. The resected defect was calculated, and accordingly, a pectoralis myocutaneous flap was harvested from the right side along with the skin paddle, which was rotated 180° and inset done [Figures 2-4]. Histopathological evaluation of the resected mass showed characteristics of biphasic histological pattern with adenocarcinoma and sarcomatous elements displaying atypical large cells with irregular nuclear contour and prominent nucleoli with vascular, lymphatic and perineural invasion. At places,



Figure 1: Depicts well defined, lobulated, hard tender mass on the neck region

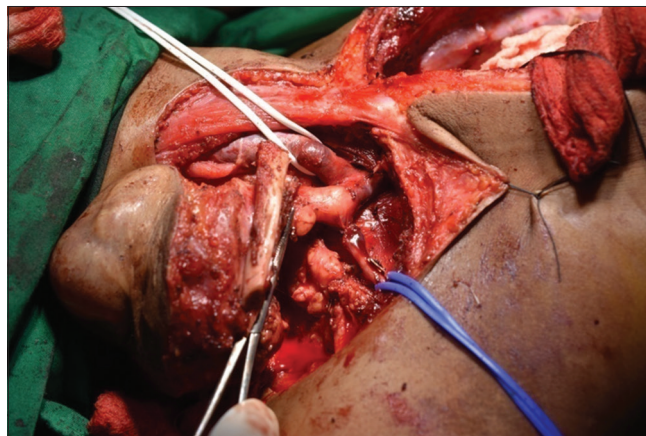


Figure 2: Depicts intra-operative picture showing internal jugular vein retracted by white elastic band and the subclavian vein retracted by blue band

necrosis and calcifications are also evident [Figure 5]. MIB 2 positivity for immunohistochemical analysis was done [Figure 6].

DISCUSSION

According to the World Health Organization and Armed Forces Institute of Pathology, Carcinosarcoma belongs to the group of true malignant mixed tumors, accounting for about <1%.^[1,16,17] Traced back to the historiography of this lesion, the term “malignant mixed tumor” was used for carcinomas arising in pleomorphic adenomas (carcinoma ex pleomorphic adenoma), sarcomas arising in pleomorphic adenoma (sarcoma ex pleomorphic adenoma), true malignant mixed tumors (carcinosarcoma), and the controversial entity “benign metastasizing pleomorphic adenoma.”^[18-20] Finally, the term was coined by King Jr. in 1976. Most commonly, it occurs in larynx, hypopharynx, esophagus, trachea, nasal cavity and oral mucosa among elderly individuals with slight male predominance.^[11-15]

The histogenesis is quite controversial. Initially, it was thought to arise from benign pleomorphic adenoma. In 1993, Gnepp *et al.*^[21] documented 43 cases of carcinosarcoma with a mean age of occurrence being 14–87 years with no sex prediction.^[14,15] Among all these cases, only 7 cases showed the histological evidence of preexisting pleomorphic adenoma and 4 cases did not show any such correlation.^[21] Later, numerous studies revealed the biphasic potential of this tumor, demonstrating the epithelial and mesenchymal components. Beyond the shadow of doubt, three dominant enigmatic theories were finally proposed for this malignancy as follows:

1. The first hypothesis stated that the tumor represents a collision tumor or squamous cell carcinoma with atypical reactive stroma resembling “pseudosarcoma”.^[8,22,23]
2. The second hypothesis stated it to be of epithelial origin, with “de-differentiation” or transformation to a spindle cell morphology (sarcomatoid carcinoma)^[24]
3. The third hypothesis stated it to be of both epithelial and mesenchymal origin. This was further



Figure 3: Depicts postresection of the tumor and repair of the subclavian vein



Figure 4: Depicts reconstruction and inset of the pectoralis myocutaneous flap into the neck defect, final closure

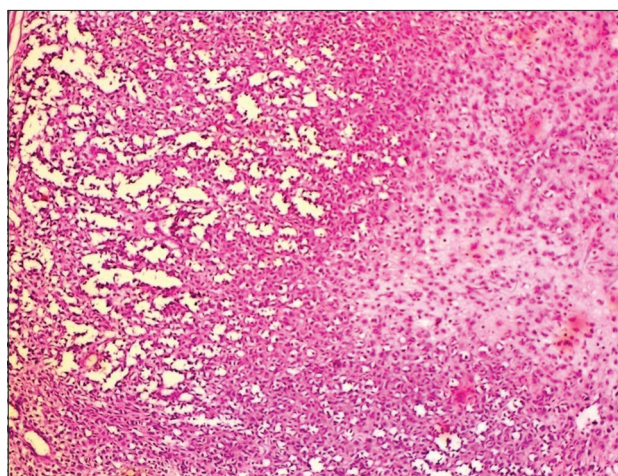


Figure 5: H and E stained section shows large pleomorphic malignant cells demonstrating cellular atypia and vascular invasion

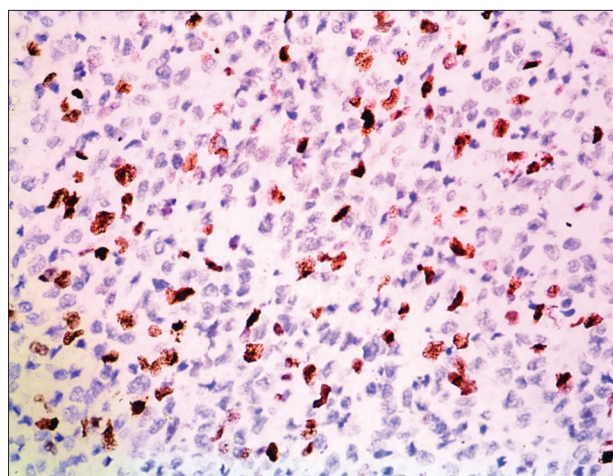


Figure 6: Immunohistochemical photomicrograph showing MIB 2 positivity

authenticated by the following observation of the occurrence of this lesion in exact sites which have the squamous epithelium, its polypoid appearance, direct contact and smooth transition of spindle cells with squamous cells and the immunohistochemical (IHC) profile displaying both markers for epithelial and mesenchymal origin.^[25,26] Hence to summarize the mysterious, baffling mechanism and histogenesis, two antithetical hypotheses called *Convergence hypothesis* and *Divergence hypothesis* were formulated in which the former showed a multiclonal origin arising from two or more stem cells and the later showed a monoclonal origin arising from a single totipotential stem cell that differentiates in separate epithelial and mesenchymal directions.^[9,15]

Being composed of both malignant epithelial component and malignant mesenchymal component, it shows specific diversity on histological examination. The epithelial component demonstrates well-differentiated keratinizing (with keratin pearls evident) or poorly differentiated non-keratinizing squamous cell carcinoma areas (no keratin pearls evident) in which squamous cells are arranged in sheets, cords, and bundles separated by varying amounts of vascular connective tissue. The epithelial component also shows adenocarcinomatous characteristics, as seen in a case study documented by Kwon and Gu as well as in our present case. The mesenchymal component demonstrates chondrosarcoma, followed by fibrosarcoma, leiomyosarcoma, osteosarcoma, and liposarcoma. Sarcoma-like, malignant features such as hypercellularity, marked pleomorphism, and enlarged nuclei are also evident similar to our case.^[4] The differential diagnosis, in this case, could be pleomorphic ex adenoma, squamous cell carcinoma, Juxtaoral organ of Chievitz and adenoid carcinoma.^[9,15,27] The IHC profiling with cytokeratin AE1/AE3, vimentin, S-100 and anti smooth muscle actin can be helpful in differentiating carcinomatous and sarcomatous components.^[28] Molecular and genetic studies conducted by Gotte *et al.* in 2000 inferred that inactivated form tumor suppressor gene on Chromosome 17 and a wild-type allele of the p53 tumor suppressor gene play a role, thus favoring the monoclonal origin. Recently, Xin and Paulino found that Ki67 can a useful prognostic marker and plays an important role in carcinogenesis.^[29] Semczuk *et al.*^[30] in his study evaluated proliferative activity using IHC analysis of carcinomatous and sarcomatous components with a panel of antibodies BCL 2, CD10, COX 2, HER 2, MIB, etc., in uterine carcinosarcoma. The results inferred that the proliferative activity was more in carcinomatous components than the sarcomatous components. The

present case report also showed the same result when immunostained with MIB 2 [Figure 6].

The treatment is a radical surgical resection, which should be combined with radiation and chemotherapy.^[31] Our case was treated by *en bloc* resection of the tumor along with the involved structure followed by adjuvant radiotherapy. In the literature, recurrence occurs in approximately two-thirds of patients and metastases in about half of them. The median period of survival after diagnosis is 10 months in 63%.^[32] In the present case, the rare occurrence of carcinosarcoma in the younger age group creates a hypothesis that needs to be further explored for better understanding of the pathogenesis, which can be correlated with the recurrence and proper treatment plan.

CONCLUSION

The case reported in this article is a carcinosarcoma (CRS), which combines both carcinomatous and sarcomatous components. It is reported because of its rare occurrence in the neck in the younger age group. Although the number of reported cases is less, the combination of radical surgical excision and radiotherapy not always seems to be the treatment of choice for CRS currently.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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