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Carcinosarcoma Maxillary Sinus: A Rare Case Report with Review of Literature

Sanudev Sadanandan Vadakke Puthiyottil[1] Mohamed Jabir K.[1] Arathi Edayattil[1] Suprya N. K.[2] Suma R.[3] Amina Abdurahman[4]

[1] Department of Medical Oncology, Government Medical College, Kozhikode, Kerala, India
[2] Department of Pathology, Government Medical College, Kozhikode, Kerala, India
[3] Department of Otorhinolaryngology, Government Medical College, Kozhikode, Kerala, India
[4] Department of Radiation Oncology, Government Medical College, Kozhikode, Kerala, India

Address for correspondence Mohamed Jabir K., MBBS, DNB, Department of Medical OncologyGovernment Medical College, Kozhikode 673008, Kerala, India (e-mail: jabz007.mj@gmail.com).

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Abstract

Carcinosarcoma of maxillary sinus is an extremely rare type of cancer in the head and neck region. It is an aggressive tumor compared with other head and neck malignancies.

An 85-year-old male evaluated for left nasal obstruction, swelling at the root of the nose, and epistaxis. On evaluation with nasal endoscopy, computed tomography (CT) and magnetic resonance imaging showed an enhancing proliferative soft tissue mass, involving the left nasal cavity, with destruction of the left frontal bone, extending up to the medial wall of the orbit. Biopsy showed a biphasic tumor with malignant squamous and sarcomatoid areas. He underwent medial maxillectomy and histology report was consistent with carcinosarcoma, with positive margins. Repeat CT showed gross residual disease. The patient was treated with palliative radiotherapy in view of advanced age, multiple comorbidities, and poor performance status and poor general health. He tolerated the treatment well. Surgery with or without adjuvant radiation is the mainstay of treatment. There is no role of adjuvant chemotherapy as of now. Chemotherapy can be tried in neoadjuvant or palliative setting. Radiotherapy can be delivered for postsurgical palliation of local recurrence. Palliative radiotherapy can be offered when no other options are available or not tolerated, or to relieve symptoms, or to treat the metastases, or in case of recurrent disease. There is no evidence for role of immunotherapy or targeted agents in the neoadjuvant or adjuvant setting. Molecular studies may pave way to the future therapeutic options for this rare variant.

Keywords

- carcinosarcoma
- maxillary sinus
- case report
- sarcomatoid carcinoma
- pseudosarcoma

Introduction

Carcinosarcoma of maxilla is a rare tumor which has a characteristic feature of both epithelial and mesenchymal components. Localized disease can be managed with surgery and adjuvant radiotherapy (RT)/chemoradiation. Palliative radiation and chemotherapy can be offered for metastatic or unresectable disease. There is very minimal evidence for the use of immunotherapy or any other targeted treatment available for this rare variant. Molecular studies may give

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way for newer dimensions in the treatment of these patients. Here, we present a case of carcinosarcoma of maxilla with brief review of literature.

Case Report

An 85-year-old male with a history of cerebrovascular accident, diabetes mellitus, hypertension, and chronic obstructive pulmonary disease on medications presented with complaints of left nasal obstruction for 1 month, swelling at the root of the nose for 2 weeks, and epistaxis for 3 days to the ear, nose, and throat outpatient department (OPD). Clinical examination showed a 4×2.5 cm soft swelling in the left side of the root of the nose, absent fogging on the left side, and a mass visible in the left nasal cavity with purulent discharge. There was no cervical lymphadenopathy. Systemic examinations were within normal limits. Nasal endoscopy showed a grayish friable proliferative growth filling the left nasal cavity and reaching up to the anterior areas, firm in consistency, sensitive to touch, and bleeds on touch. Computed tomography (CT) scan imaging showed an enhancing soft tissue mass approximately 7.2×4.3 cm centered on the left nasal canal with extension to the bilateral frontal and ipsilateral ethmoid sinuses, with destruction of anterolateral outer table of left frontal bone reaching to the subcutaneous plane and medial wall of the left orbit. Magnetic resonance imaging (MRI) done showed altered intensity expansile mass lesion noted arising from the left nasal cavity, measuring $9.7 \times 2.3 \times 7.3$ cm, showing heterogeneous T2 hyperintensity, T1 hypointensity, short tau inversion recovery hyperintensity, and no blooming in gradient echo necrotic areas noted within showing suppression in fluid-attenuated inversion recovery. The lesion was extending to bilateral frontal and ipsilateral ethmoid sinuses. Anteriorly occupying and invading the left nasal vestibule posteriorly reaching the nasopharynx, medially abutting and indenting the nasal septum, laterally up to the medial wall of the orbit with focal breach, up to osteomeatal unit with no obvious extension into the maxillary sinus. Biopsy (June 2023) showed a biphasic tumor with malignant squamous and sarcomatoid areas. Sarcomatoid areas showing liposarcomatous, osteosarcomatous, and undifferentiated components (►**Fig. 1**). Immunohistochemistry (IHC) revealed that p63, p40, and CK were positive in atypical squamous cells, vimentin was positive in sarcomatous areas, and CD31 and CD34 were highlighted in the blood vessels. The morphology and IHC was consistent with carcinosarcoma. Patient underwent medial maxillectomy in August 2023 and histology report was consistent with carcinosarcoma and all margins were positive. With this report, he was referred to the medical oncology department for further management.

On examination, his Eastern Cooperative Oncology Group performance status (ECOG PS) was 2. Geriatric assessment with (G8) tool revealed a score of 12, which required a full geriatric evaluation for the patient. Cancer and Aging Research Group score was 7, which placed the patient in mid-

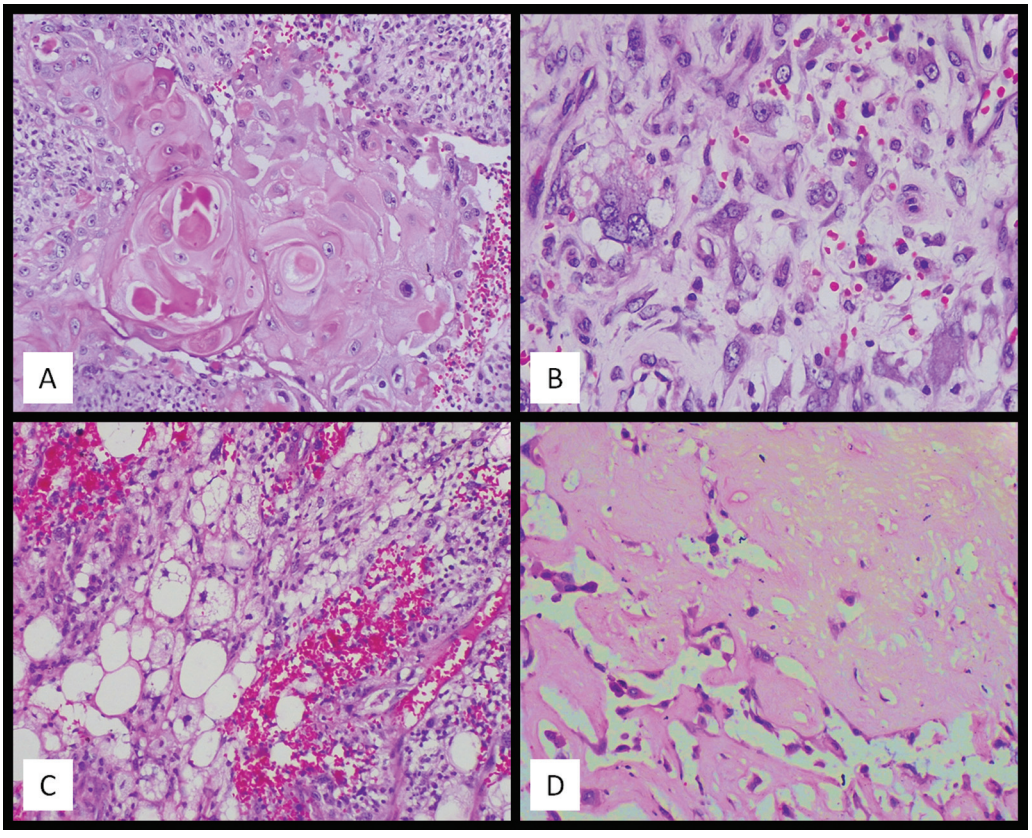

Fig. 1 (**A**) Hematoxylin and eosin (H&E), epithelial component showing well-differentiated squamous cell carcinoma. (**B**) H&E, mesenchymal component showing malignant spindle cells with tumor giant cells. (**C**) H&E, mesenchymal component showing osteosarcomatous area. (**D**) H&E, mesenchymal component showing liposarcomatous area.

risk category. There was a swelling of size 4 × 3 cm at the root of the nose. A surgical scar of size 6 cm in the right malar region. There was no cervical lymphadenopathy noted. Repeat contrast-enhanced CT (CECT) scan of the head, neck, thorax, and abdomen was taken, which showed enhancing soft tissue mass of approximately 7.2 × 4.3 cm centered on the left nasal canal with extension to the bilateral frontal and ipsilateral ethmoid sinuses, with destruction of anterolateral outer table of the left frontal bone reaching to the subcutaneous plane and medial wall of the left orbit. The case was discussed in multidisciplinary tumor board and clinicopathology meet and decided to treat with palliative RT in view of advanced age, multiple comorbidities, ECOG PS 2, and poor general health. He completed hypofractionated RT, 45 Gy in 15 fractions. He tolerated the treatment well without much side effects. We suggested adjuvant chemotherapy but the patient's relatives were not willing for the same. Next-generation sequencing (NGS) could not be performed due to financial constraints. The patient was reviewed in our OPD in June 2024, clinical examination revealed no deterioration in general health and performance status and the disease was clinically in remission.

Discussion

Carcinosarcoma is a rare and unique variant of carcinoma. The term carcinosarcoma was first described by Virchow in 1864, which is composed of both epithelial and mesenchymal components. It may be otherwise called pseudosarcoma, sarcomatoid squamous cell carcinoma, pleomorphic carcinoma, spindle cell carcinoma, or collision tumor.[1] In the upper respiratory region, the most common locations include the larynx, hypopharynx, esophagus, trachea, oral mucosa, and nasal cavity. Its incidence is very low in the maxilla. Incidence is highest in males; mean age: sixth decade. Commonly identified risk factors are the history of alcohol abuse, smoking, and radiation exposure. Different hypotheses were postulated for the carcinogenesis of this variant. Two primary hypotheses are "convergence hypothesis" and "divergent hypothesis." The convergence hypothesis proposes that a tumor with different cell types comes from two separate stem cell lines. The divergent hypothesis suggests that components have the same monoclonal origin, with the epithelial cell being transformed and dedifferentiated into sarcomatoid cells. Recent electron microscopic and immunohistochemical investigations favor divergent hypothesis that could explain the presence of two different cells in this variant.

Further investigations utilizing polymorphic microsatellite markers in both components revealed shared patterns in the loss of heterozygosity, suggesting late patterns of divergent evolution.[2] It is believed that the morphological change of cells from squamoid to a spindled type has been caused by an impaired intercellular adhesion complex cadherin catenin. The most common gene mutation that has been identified is believed to be p53. Carcinosarcoma may be a sarcomatous metaplasia of squamous cell carcinoma, based on the expression of antigen positivity of both epithelial and mesenchymal markers in transitory cells.

Clinically, the signs and symptoms are often nonspecific, can be oral, nasal, orbital, facial, and auditory.

Nasal symptoms may encompass unilateral nasal blockage, loss of smell, nosebleeds, nasal drainage, and speech that lacks nasal resonance. Oral symptoms may manifest as a widened alveolus, toothache, obvious mass on the palate, and presence of ulceration or nonhealing wounds. Orbital symptoms may include edema of the eyelid, proptosis, excessive tearing, double vision, or loss of vision. Facial symptoms may involve a loss of nasolabial fold, facial asymmetry, an evident mass on the cheek, cutaneous fistula, facial swelling, reduced sensation, pain in the cheek due to the invasion of the infraorbital nerve, and extension into the nasopharynx—causes hearing loss, blockage or dysfunction of the Eustachian tube, and accumulation of serous effusion. Posterior extension of the tumor can cause severe, deep-seated pain due to skull base invasion, trismus due to pterygoid muscle invasion and cranial neuropathies.

Blood investigations should be done during the workup, including complete blood count, renal and liver function test, and viral markers. Nasal endoscopy is done to assess the character of swelling, extent of the disease, and sinus involvement and to get a tissue biopsy. X-ray of paranasal sinuses may show opacities filling the sinuses. CT and MRI will show the local extension of the disease and nodal involvement. CECT is relevant to identifying invasion and/or destruction of the bone including the skull base. MRI with Gadolinium (Gd) contrast is superior in identifying the cortical involvement of bone, extension to soft tissue, orbit, intracranial extension or nerve involvement, and precise determination of perineural invasion. Positron emission tomography-CT scan may help to assess the nodal and distant metastases.

On microscopic examination, it shows both epithelial and mesenchymal components. The epithelial component demonstrates well to poorly differentiated squamous cell carcinoma with cells arranged in sheets, cords, and bundles. It may also show adenocarcinomatous characteristics. The mesenchymal component may be chondrosarcoma, fibrosarcoma, leiomyosarcoma, osteosarcoma, or liposarcoma, with hypercellularity with marked pleomorphism, and large nucleus. IHC with cytokeratin AE1/AE3, vimentin, S-100, and antismooth muscle actin can be helpful in differentiating carcinomatous and sarcomatous components. K_i-67 reactivity was observed diffusely in areas containing spindle cells.[1] But a negative expression of epithelial markers does not rule out the diagnosis of carcinosarcoma, as there could be loss of squamous marker with the loss of squamous differentiation.

Pathological differentials include other benign and malignant conditions like variants of squamous cell carcinoma, Pleomorphic ex-adenoma, juxtaoral organ of Chievitz, adenoid carcinoma, malignant fibrous histiocytoma, fibrosarcoma, malignant melanoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumor, leiomyoma, and leiomyosarcoma. Molecular studies are a way through for future treatment approach. There are very few studies done so far in this rare variant of squamous cell carcinoma (►**Table 1**). Ansari-Lari et al observed a high incidence of strong and diffuse p53

Table 1 Summary of molecular studies done in carcinosarcoma

No	No of patients	Site/s	Technique	No. of genes	Conclusion	Author, year
1	1	Maxilla	NGS	50	• Somatic mutation in MET proto-oncogene • Increased copy number of PIK3CA	Ando et al, 2015[6]
2	1	Maxilla	NGS	539	• PDCD6-TERT fusion - 44.81% • SF3B1 - 32.94% • CDKN2A - 28.52% • TERT - 8.20%	Zhang et al, 2022[5]
3	12	Head and neck sites	NGS	539	• TP53 –– 11 (92%) • CDKN2A - 8(58%) • CDKN2B - 6 (50%) • FAT1 - 4 (33%) • PIK3CA, MUC16, PRKDC, MYC, CDK4 - 3 (25%) • Median tumor mutation burden (TMB) - 4.34 per megabase (0.71–14.71)	Chen et al, 2021[4]
4	10	Head and neck sites	FISH	1	ALK positive - 20%	Kim et al, 2015[18]
5	23	Head and neck sites	PCR	1	P53 mutation - 78%	Ansari-Lari et al, 2002[3]
6	36	Lung	WES	48	• TP53 (60%), KRAS (20%), PIK3CA (20%), MET (20%), NOTCH (10%), STK11 (10%), RB1 (10%) • Crizotinib showed marked effects on cell viability and decrease in downstream AKT and MAPK activation	Liu et al, 2016[15]
7	15	Esophagus	NGS	432	• TP53 - 100% • RTK - 67% • PI3KCA - 67% • MMR genes and proofreading gene POLE - 33%	Lu et al, 2018[16]

Abbreviations: FISH, fluorescence in situ hybridization; MET, mesenchymal-epithelial transition; NGS, next-generation sequencing; PCR, polymerase chain reaction; WES, whole exome sequencing.

expression in carcinosarcoma in the head and neck, compared with the squamous cell carcinoma counterpart.[3] According to Chen et al, the most frequently mutated gene in the carcinosarcoma is *TP53*.[4] Zhang et al conducted a molecular study by NGS in a locally advanced undifferentiated sarcomatoid carcinoma of right maxillary sinus found a PDCD6–TERT fusion gene.[5] Ando et al conducted a genetic analysis by examining 50 genes using NGS on a patient with maxillary carcinosarcoma and identified a somatic mutation in the mesenchymal-epithelial transition factor (MET) proto-oncogene in both components of the tumor.[6] Furthermore, they also discovered that despite the absence of any identified mutations in the screened PI3K-AKT pathway genes, both components exhibited a slight elevation in the copy number of PIK3CA. This observation implies a potential involvement of the PI3K-AKT pathway in the development of oncogenesis. There is a 20% incidence of ALK translocation in carcinosarcoma in the head and neck region.

As this tumor is aggressive, a multimodal type of therapy is needed. Surgery remains the primary treatment. It depends on which walls of the maxillary sinus are involved. Shah et al[7] categorized maxillectomies into limited, subtotal, and total maxillectomies. Limited maxillectomy means the removal of one wall, either resection of the floor or medial wall of the maxillary sinus. The subtotal maxillectomy includes removal of at least two walls, along with the palate. Total maxillectomy means resecting the entire maxilla. Total maxillectomy is the preferred modality done in most of the case studies (►**Table 2**). Radical maxillectomy is a term reserved for excision of the pterygoid plate along with the maxillectomy in advanced maxillary sinus cancers with invasion of infratemporal fossa to provide adequate margins and reduce the local recurrence rates. If periorbita is involved, maxillectomy with resection of orbital floor must be performed; may include orbital exenteration. If pterygoid plates or posterior wall are involved, resection must include those sites.[7]

Reconstruction in surgery is determined by the type of surgical defect present. This may involve regional soft tissue with bone flaps, free flaps, bone flaps with soft tissue or

Table 2 Summary of case reports published after 2000

No	Age/sex	Stage	Treatment	Outcome	Author, year
1	47/M		Partial maxillectomy + radiation therapy	Local recurrence, death after 1 year	Furuta et al, 2001[23]
2	54/M	T3N3M0	Radiation therapy + chemotherapy	Death after 4 months	Howard et al, 2007[24]
3	60/M	T3N0M0	Total maxillectomy + radiation therapy + chemotherapy	Local recurrence	Moon et al, 2013[25]
4	52/M	T4aN0M0	Total maxillectomy + radiation therapy + chemotherapy	Local recurrence (soft palate)	Alem and AlNoury, 2014[26]
5	61/M	T4aN0M0	Total maxillectomy with a modified radical neck dissection	Sternal metastasis within 1 month and died	Cheong et al, 2014[27]
6	55/M	T4aN0M0	Radical radiotherapy	No response, died after 4 months	Li et al, 2023[28]

Abbreviation: M, male.

bone, as well as a combination of soft tissue flaps and alloplastic implants. Examples of regional soft tissue flaps include temporalis myofascial flaps, facial artery musculomucosal flaps, buccal pad of fat flaps, and reverse submental flaps. Free soft tissue flaps consist of radial forearm flaps, rectus abdominis flaps, anterolateral thigh flaps, and deep inferior epigastric perforator flaps.

Adjuvant RT is indicated in T3/T4 tumors, perineural, vascular, or lymphatic invasion, close or positive margins, or extranodal extension. Volumetric modulated arc therapy/intensity-modulated RT is the current standard of radiation therapy in the head and neck site, usually started 3 to 6 weeks after surgery. In postoperative setting, the dose to the primary site is 55, 60, and 66 Gy in 1.8 to 2 Gy/fraction for tumors with free margin >1 cm, <1 cm margin, and with positive margins, respectively. Elective neck irradiation is routinely not recommended unless patient is having positive nodes. For patients receiving preoperative RT, the dose is 50 Gy in 1.8 to 2 Gy/fraction; radical RT dose is 70 Gy in 35 fractions delivered over 7 weeks.

For definitive therapy, gross tumor volume includes the gross tumor disease. The clinical target volume (CTV) encompassed the maxilla, floor of the maxillary sinus, medial aspect of the orbit, pterygomaxillary space, infratemporal fossa, ethmoid sinuses, and nasal cavity except around the ipsilateral optic apparatus. CTV for adjuvant therapy involves covering the resection cavity and modified to encompass the entire affected anatomical compartment or sinus, while making modification to anatomical boundaries to avoid bone and/or air without signs of invasion. Elective neck irradiation was not routinely performed for clinically node-negative disease. Planning target volumes are created by expanding the CTV structures by 5 mm autoexpansion. The organs at risk include the mandible, spinal cord, brainstem, optic nerve, eyeball, optic chiasm, and lens. Acute toxicities include oral mucositis, pharyngitis, dermatitis, rhinitis, and conjunctivitis. Chronic toxicities include fistula formation, cataracts, retinopathy, epiphora, nasal obstruction, cellulitis, fibrosis, and osteoradionecrosis of the orbital wall.[8]

Adjuvant chemoradiation is indicated when margin is positive after surgical resection or in case of lymph nodes with extracapsular spread. Concurrent chemotherapy is added to radical RT in unresectable nonmetastatic disease. Most common dose is cisplatin at 40 mg/m^2 weekly during the course of radiation.

There is no role of adjuvant chemotherapy, immunotherapy, or targeted therapy as of now. As there is no sufficient data on carcinosarcoma maxilla, chemotherapy regimens are extrapolated from squamous cell carcinoma of the head and neck. In neoadjuvant setting, chemotherapy regimens may include platinum, with taxanes or fluorouracil (5-FU), either as doublet or triplet. A study with induction chemotherapy in advanced squamous cell carcinoma of paranasal sinuses, with either a two-drug combination (taxane + platinum, taxane + 5-FU) or three-drug combination (taxane + platinum with ifosfamide or 5-FU), $>67\%$ of the patients achieved at least a partial response.[9] A retrospective analysis conducted on carcinosarcoma of unknown primary revealed that the predominant treatment regimen consisted of gemcitabine and docetaxel, proven effective for both epithelial cancers and sarcoma. Subsequently, a combination of gemcitabine or taxane with a platinum agent was often utilized.[10] Common toxicities include anemia, neutropenia, thrombocytopenia, vomiting, febrile neutropenia, sepsis, electrolyte imbalance, diarrhea, and renal dysfunction.[11] There is no evidence for role of immunotherapy or targeted agents in the neoadjuvant setting. Radiologic response is evaluated after two cycles with CECT scan or Gd-enhanced MRI and scored according to the Response Evaluation Criteria in Solid Tumors criteria.

In the palliative setting, there is no role for surgery. RT might be an option for postsurgical palliation of local recurrence. Palliative RT can be offered when no other options are available or not tolerated, or to relieve symptoms (pain, bleeding, ulceration), or to treat the metastases, or in case

of recurrent disease. Dose of RT can be 30 Gy in 10 fractions, 20 Gy in 5 fractions, or 8 Gy in 1 fraction. For the newly diagnosed, T4b or unresectable nodal disease or unfit for surgery or metastatic at presentation, in patients with poor general health, palliative RT or single-agent systemic therapy is an option. Pembrolizumab and nivolumab with chemotherapy were approved for the palliative treatment of maxillary sinus tumors.[12,13] Cetuximab, platinum (cisplatin, carboplatin), 5-FU, and taxanes in different combinations are the preferred systemic therapeutic agents other than immunotherapy in palliative setting.[14]

A study done by Chen et al analyzed patients with NGS and identified TP53 as the most frequently mutated gene.[4] TP53 is the most common mutated gene in carcinosarcoma of nonhead and neck sites.[15,16] TP53-mutated carcinomas and sarcomas respond better to bevacizumab and pazopanib, respectively.

Multiple inhibitors, including MAT2A inhibitors, PARP inhibitors, and c-MYC inhibitors, have been administered to patients with gene mutations related to deoxyribonucleic acid (DNA) repair and cell cycling. PI3-kinase pathway was activated in a proportion of patients, which could be targeted by PI3K/mTOR/AKT or MEK inhibitors, which are currently in ongoing clinical trials

Several inhibitors, such as MAT2A inhibitors, PARP inhibitors, and c-MYC inhibitors, have been administered to patients who have gene mutations associated with DNA repair and cell cycling. A proportion of patients showed activation of the PI3-kinase pathway, which could be targeted by PI3K/mTOR/AKT or MEK inhibitors. These inhibitors are currently being tested in clinical trials.

Furthermore, patients suffering from carcinosarcoma of the head and neck may also experience promising benefits through the utilization of clinical inhibitors targeting histone modification (Wee1 inhibitor) and chromatin remodeling (histone deacetylase inhibitor).[4] Zhang et al conducted a molecular study in a locally advanced sarcomatoid carcinoma of the right maxillary sinus and found a PDCD6–TERT fusion gene.[5] In recent years, a retrospective cohort analysis has found that pan-cancer TERT variation can benefit from immune checkpoint inhibitors.[17] The patient was treated with adjuvant chemoradiotherapy combined with immune checkpoint inhibitor (camrelizumab [anti-PD-1 inhibitor]), found good curative effect. Ando et al found that each of the epithelial and mesenchymal components, the conserved domain of the mesenchymal-epithelial transition factor (MET) proto-oncogene, showed a somatic mutation within, suggesting the possibility of treating MET mutation-positive carcinosarcomas with c-MET inhibitors.[6] Kim et al have tried with crizotinib in a patient with carcinosarcoma of the maxillary sinus, resulting in tumor shrinkage with 4 months of progression-free interval and clinical improvement.[18] An ongoing study assesses the antitumor activity of crizotinib in a variety of malignancies with alterations in ALK and/or MET pathways.[19] Carcinosarcoma, being a biphasic cancer, could potentially indicate the occurrence of epithelial-mesenchymal transition (EMT). Chemoresistant cell lines are enriched with EMT markers, may benefit patients with EMT-targeted drugs, though it is an area of active research.[20–22]

Disease progression is characterized by recurrences and metastases, seen in 33% cases, including local recurrence (20.5%), lymph nodal metastases (42%), and distant metastases to the lung and soft tissue (5.1%). The tumor size, location, depth of invasion, and stage of the disease are the important factors in determining prognosis. Local recurrences and metastases frequently occur and often result in fatality. The 5- and 10-year overall survival of sinonasal carcinosarcoma were 45 and 37.5%, respectively, with disease-specific survival (DSS) of 48.5 and 37.5%, respectively. Local recurrence eventually occurs and survival periods were from 2 to 40 months.[2] The sinonasal carcinosarcoma cohort exhibited significantly lower 5- and 10-year DSS rates compared with both the nonsinonasal carcinosarcoma cohort and the carcinosarcoma cohort at all other head and neck sites.[29] The limited accessibility of the sinonasal tract may contribute to the challenges faced during surgical interventions, unlike in nonsinonasal regions. Additionally, carcinosarcoma tends to exhibit aggressive and infiltrating behavior in the sinonasal area, whereas in nonsinonasal regions like the larynx and pharynx, it often presents a polypoid growth pattern, resulting in more favorable surgical outcomes.[30]

Conclusion

Carcinosarcoma maxillary sinus is an extremely rare and a very aggressive tumor. The exact biology is still unknown, though many hypotheses have been proposed. There are very limited data to direct the direct treatment guidelines except in uterine carcinosarcoma. Prognosis is poorer compared with squamous cell carcinoma maxilla, carcinosarcoma of other head and neck sites, or nonhead and neck sites. Resectable nonmetastatic disease is managed by radical maxillectomy and adjuvant radiation or chemoradiation. Metastatic disease is managed by palliative chemotherapy and/or radiation therapy. Further treatment options are limited. With the continuous approval of more targeted therapies, molecular studies are set to play a significant role in providing treatment options for patients with limited choices. Nevertheless, further research is imperative in this field.

Declaration of Patient Consent

We have obtained written consent from the patient. The full statement is attached with the form submitted with the manuscript.

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Conflict of Interest

None declared.

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