Management of Spindle Cell Carcinoma of the Maxillary Sinus: A Case Report and Literature Review

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Conflict of interest: None declared

Patient: Male, 52
Final Diagnosis: Spindle-cell carcinoma of the maxillary sinus
Symptoms: —
Medication: —
Clinical Procedure: Functional endoscopic sinus surgery • total maxillectomy
Specialty: Otolaryngology

Objective: Rare disease
Background: Carcinosarcomas, also known as spindle cell carcinomas, are rare and highly aggressive tumors characterized by dual histologic differentiation of squamous cell and mesenchymal cell tumors. Occurrence of carcinosarcoma in maxillary sinus is very rare, with only 11 cases reported since 1957. The small number of reported cases creates an obstacle to the increased understanding of the behavior, prognosis, and therapeutic management of this tumor.

Case Report: A 52-year-old man presented with a 2-month history of right nasal obstruction. Computed tomography (CT) and magnetic resonance imaging (MRI) showed opacified right frontal, sphenoid, ethmoid, and maxillary sinuses with soft tissue density and expansion of the mass with erosion of the right lateral maxillary wall. Functional endoscopic sinus surgery (FESS) was done and histopathology revealed multiple fragments of nasal mucosa lined by stratified hyperplastic squamous epithelium with an increased degree of dysplasia and pleomorphism and a second spindle cell high-grade neoplastic growth with bizarre giant cells and abnormal mitotic figures. Consistent with carcinosarcoma, immunohistochemistry showed strong positive staining for vimentin in the spindle cell component and strong positive staining for cytokeratin markers in the epithelial cell component. The patient underwent right total maxillectomy with postoperative chemoradiation therapy and survived for 1 year.

Conclusions: Carcinosarcoma of the maxillary sinus is a rare disease with non-specific symptoms; it usually presents in the advance stage and is associated with poor patient prognosis. This case indicates that surgical intervention with postoperative chemoradiotherapy improves patient prognosis and should be considered as the standard therapeutic modality.

MeSH Keywords: Carcinoma • Carcinosarcoma • Maxillary Sinus Neoplasms • Paranasal Sinus Neoplasms

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Background

Carcinosarcomas (also known as pseudosarcoma, pseudosarcomatous squamous cell carcinoma, pleomorphic carcinoma, and spindle cell carcinoma) are biphasic tumors with high-ly aggressive behavior. They are characterized by dual malign-ant histologic differentiation with an epithelial component consisting of focal squamous cell carcinoma and a mesenchymal component with sarcomatoid stroma [1].

Carcinosarcoma may arise in squamous epithelium in any part of the body and has been reported to occur in the upper aerodigestive tract, salivary glands, thyroid, thymus, lung, breast, gastrointestinal tract, hepatobiliary system, genitouri-nary tract, and uterus; however, its occurrence in the sinusosal cavity is extremely rare [2–9]. In this report, we present a case of right carcinosarcoma of the maxillary sinus and briefly discuss the management and follow-up of the case, with a review of the literature on this rare tumor.

Case Report

Following hospital ethics committee approval, a 52-year-old man presented to the ear, nose, and throat clinic of King Abdulaziz University Hospital (a tertiary care hospital in Jeddah, Saudi Arabia) complaining of right nasal obstruction that progressed over 2 months, which was associated with numbness and pain in the right cheek. The patient had experienced intermittent mild epistaxis for 2 weeks prior to presentation. During this period, he was using decongestant nasal drops to relieve the obstruction. He did not report symptoms such as allergic rhinitis symptoms, postnasal drip, decreased hearing, visual problems, or headache. The patient had no significant medical history or any associated comorbid conditions. He lived and was raised in Sanjure City in Indonesia (known to have wood, cement, and steel factories), where he had been working mostly as a tobacco farmer, and he had a history of smoking tobacco since the age of 20 years.

Endoscopic examination revealed a fleshy mass filling the right-side nasal cavity, but there was no cheek fullness. An ear ex-amination revealed a dull tympanic membrane in the right ear. No lymph node abnormalities were detected during a head and neck examination, and the findings of cranial nerve examination were normal. Systemic examination findings were also unremarkable. Computed tomography (CT) of the paranasal sinus (PNS) showed that the right frontal, sphenoid, ethmoid, and maxillary sinuses were opacified, with soft tissue density and expansion of the mass, with erosion of the right lateral maxillary wall (Figures 1 and 2).

The patient underwent functional endoscopic sinus surgery (FESS), which showed a very firm mass involving the right nasal cavity that was eroding the posterior wall of the maxillary sinus. Intraoperative hemorrhaging occurred from the internal maxillary artery and was controlled by bipolar cauterization and packing. Postoperative MRI scans revealed an enhancing mass along the anterior wall of the right maxillary sinus, with an invasion of the lateral wall and extension up to the posterior edge of the infratemporal fossa. Minor enhance-ment was apparent along the lateral pterygoid muscles, but there was no evidence of pterygoid destruction, and the mass involved the inferior wall of maxillary sinus, with a small extension to the palate. There was no evidence of intracranial or orbital extension.
An excisional biopsy specimen of the mass was sent for histopathological analysis. Histopathology revealed multiple fragments of nasal mucosa lined by stratified hyperplastic squamous epithelium, with an increased degree of dysplasia and pleomorphism and a second spindle cell high-grade neoplastic growth with bizarre giant cells and abnormal mitotic figures. Consistent with carcinosarcoma, immunohistochemistry showed strong positive staining for vimentin in the spindle cell component and strong positive staining for cytokeratin markers in the epithelial cell component (Figure 3).

A metastatic work-up revealed negative lab results, and no abnormalities in CT scans of the neck, chest, abdomen, or brain. The clinical TNM staging of the patient was T4aN0M0 (Stage 4a).

The patient underwent right total maxillectomy through a Weber-Ferguson incision and lip splitting. The mass was removed en bloc and was found to also involve the posterior orbital wall in addition to the previously mentioned sites. A surgical obturator was placed with the assistance of the maxillofacial surgical team. Postoperative histopathology of the mass confirmed the diagnosis of carcinosarcoma of the maxillary sinus.

The patient received postoperative chemoradiation therapy consisting of 2 cycles of cisplatin (30 mg/m²) intravenously infused with 500 mL normal saline over 2 h and intensity-modulated radiation therapy (IMRT) over the right maxilla for a total of 70 Gy in 35 fractions. To date, there is no evidence of tumor recurrence in the right maxillary sinus region on MRI scans, 1 year later (Figure 4). However, the patient exhibited soft tissue thickening, nodularity, and enhancement in the remainder of the right side of the soft palate, and recurrence was suspected in the left side of the soft palate, but the patient has refused any further management.

**Discussion**

Carcinosarcomas are biphasic tumors with dual components: an epithelial, squamous cell carcinoma component and a...
sarcomatoid component. This tumor can arise in the squamous epithelium in any part of the body. In the head and neck region, it commonly occurs in the pharynx or larynx [10] and is extremely rare in the sinonasal cavity [2,4]. Management strategies, tumor histogenesis, and the clinical course for such cases are controversial because of the limited number of cases reported in the literature (Table 1).

Two hypotheses for the origin of carcinosarcoma have been proposed. The convergence hypothesis suggests that a heterologous tumor originates from 2 stem-cell lines, whereas the divergence hypothesis suggests that the tumor originates from a single stem cell that differentiates into epithelial and sarcomatous components [2]. A recent study reported epithelial differentiation in some spindle cells in the sarcomatous component, suggesting that this component is derived from the mesenchymal transformation of the carcinoma cells, supporting the divergence hypothesis [11].

Including the current case, 18 cases diagnosed as carcinosarcoma of the maxillary sinus have been reported since 1957 [2,9,11–13]; 6 of them provided insufficient clinical information [14]. Carcinosarcoma of the maxillary sinus is mostly reported in older patients aged 47–80 years. Seven cases involved men and 5 cases involved women. There are no specific symptoms associated with carcinosarcoma of the maxillary sinus, but a history of unilateral nasal obstruction and epistaxis were common symptoms in all reported cases. The non-specific symptoms of carcinosarcoma of the maxillary sinus complicate an early diagnosis; patients often present late, creating a therapeutic challenge and increasing the likelihood of a poor prognosis.

Eight of the 11 previous cases underwent total maxillectomy and almost all had radiotherapy with or without surgery. Patients that were treated with surgery and adjuvant chemoradiation therapy tended to have a better prognosis. In the current case, the carcinosarcoma of the maxillary sinus was managed with a total maxillectomy and postoperative chemoradiation therapy.

Since the number of reported cases of carcinosarcoma of the maxillary sinus is small, it is difficult to report survival,

Table 1. A review of similar cases in the published literature.

| No | Age/sex | Stage | Treatment | Outcome | Author, year |
|----|---------|-------|-----------|---------|--------------|
| 1  | 62/F    |       | Radiotherapy | No marked improvement of the tumor, dead with disease | Meyer, 1957 |
| 2  | 62/F    |       | Radiotherapy | Death in 40 months | Lichtiger, 1970 |
| 3  | 71/M    | T4N0M0 | Preoperative radiotherapy + total maxillectomy + removal of eye | Death due to intracerebral abscess at postoperative period | Feinmesser, 1982 |
| 4  | 65/F    |       | Total maxillectomy + radiotherapy | Local recurrence, death 8 months later | Ampil, 1985 |
| 5  | 57/F    |       | Tumor excision, ethmoidectomy and turbinectomy | Local recurrence 5 months after surgery | Hafiz, 1987 |
| 6  | 60/M    | T3N0M0 | Total maxillectomy + radiation therapy + chemotherapy | Local recurrence, death 2 months later | Sonobe, 1989 |
| 7  | 53/M    | T4N0M0 | Total maxillectomy + craniofacial resection + radiation therapy + chemotherapy | Disease free after 9 months | Shindo, 1990 |
| 8  | 80/F    | T3N0M0 | Total maxillectomy + radiation therapy + 2nd operation | Local recurrence, death 2 months after second operation | Sanabre, 1998 |
| 9  | 47/M    |       | Partial maxillectomy + radiation therapy | Local recurrence, death after 1 year | Furuta, 2001 |
| 10 | 54/M    | T3N3M0 | Radiation therapy + chemotherapy | Death after 4 months | Howard, 2007 |
| 11 | 60/M    | T3N0M0 | Total maxillectomy + radiation therapy + chemotherapy | Local recurrence | Jeong-Ki Moon, 2009 |
| 12 | 52/M    | T4aN0M0 | Total maxillectomy + radiation therapy + chemotherapy | Local recurrence (soft palate) | Current case, 2012 |
recurrence, and mortality rates. An overall mortality rate of 42% at 30 months was reported for carcinosarcoma at different sites [15]. Of the 12 cases discussed here, tumor recurrence occurred in 8.

Conclusions

Carcinosarcoma of the maxillary sinus is a rare disease with non-specific symptoms; it usually presents in the advance stage and is associated with poor patient prognosis. The small number of reported cases creates an obstacle to the increased understanding of the behavior, prognosis, and therapeutic management of this tumor type. This case indicates that surgical intervention with postoperative chemoradiation therapy improves patient prognosis and should be considered as the standard therapeutic modality. Further prospective randomized studies are necessary to confirm such intervention.

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