Case report
Carcinosarcoma of the maxillary sinus: A rare case report
Jawad Hasnaoui, Said Anajar, Mohammed Tatari, Reda Abada, Sami Rouadi, Mohammed Roubal, Mohammed Mahtar

ENT Department, Face and Neck Surgery, Hospital August, 20’1953, University Hospital Centre IBN ROCHD, Casablanca, Morocco

HIGHLIGHTS
• Carcinosarcoma is a rare biphasic neoplasm and a variant of squamous cell carcinoma.
• Carcinosarcoma is an aggressive tumor, rapidly progressive with a poor prognosis.
• Occurrence in nasal cavity is very rare and only few cases have been reported.
• Management for this disease still remains undetermined.

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ABSTRACT
Introduction: Carcinosarcoma is a highly malignant tumor characterized by dual malignant histologic differentiation of epithelial and mesenchymal components. The tumor is extremely rare in the sinonasal tract, with only 13 cases reported since 1957 in the literature.
Presentation of case: We report a case of a 55-year-old man with right-sided face pain revealed a mass in the right maxillary sinus and nasal cavity. A large incisional biopsy from the nasal cavity concluded the diagnosis of carcinosarcoma. The patient was treated with chemoradiation, but no significant effect was obtained. The patient died 4 months after initial examination.
Discussion: In our case, the patient consulted late with a large tumor which darkens his prognosis. According to some authors, this histological type of tumors is known by local recurrences and its lethal metastases. Prognosis is related to location, tumor size, the invasion and stage of disease. Wide surgical excision is the treatment of choice. Radiotherapy can be discussed in inoperable cases.
Conclusion: This is an aggressive tumor, rapidly progressive with a poor prognosis. The management of this rare condition remains undetermined.

1. Introduction
Carcinosarcoma is a highly malignant tumor defined by dual malignant histologic differentiation of the epithelial component and a mesenchymal component having a sarcomatoid stroma. Carcinosarcoma remains an extremely rare and aggressive entity [1,2].

The tumor spreads rapidly in the epithelial tissues of the body such as the lungs, urinary tract, breast and uterus. In the head and neck region, the pharynx and larynx are most frequently involved, followed by the esophagus, the oral cavity and the nasal area. Its occurrence in nasal cavity is very rare and only few cases have been reported in the literature [1]. We report a case of carcinosarcoma of maxillary sinus. This work has been written in accordance with the SCARE criteria [3].

2. Case report
A 55-year-old North-African man with history of ischemic heart disease and high blood pressure was admitted to the hospital for a large painful mass of the right maxillary region. It appeared 3 months before admission and was associated with a right nasal obstruction, right epistaxis, and decreased visual acuity of the right eye with a persistent headache (Fig. 1).

Clinical examination of the nasal cavity showed a polyploid mass from the right middle meatus, bleeding on contact, with a bulging of the lateral wall of the nasal cavity halting advancement...
to the nasopharynx.

CT scan showed a large mass of tissue density in the right maxillary sinus and the nasal cavity with lysis of the orbital floor, the inner and outer walls of the maxillary sinus and the zygoma. It measures 70 mm × 54 mm. The tumor extends into the orbital cavity, without regional and distant metastases. (Fig. 2).

A large incisional biopsy sample (2 × 2 × 1 cm) was obtained from the nasal cavity under local anesthesia with endoscopic guidance.

The biopsy showed proliferation of malignant spindle and round cells with hyperchromatic nuclei and nucleoli. Immunohistochemically, The carcinomatous component was positive for cytokeratin and EMA but negative for vimentin. In contrast, the sarcomatous component was positive for vimentin but negative for cytokeratin and EMA. Because keratins were positive in tumor cells, a diagnosis of sarcomatoid carcinoma was made (Fig. 3).

A total maxillectomy with modified neck dissection and orbital exenteration was indicated for our patient. However, it was considered impossible due to the underlying ischemic heart disease. The echocardiography showed decompensated ischemic heart disease with 25–30% EF and a straight stenosis of the right coronary artery with active stent angioplasty 3 years ago. Following a multidisciplinary staff, the patient was admitted to the oncology department for chemoradiation based on docetaxel, cisplatin, 5-fluorouracil and radiation therapy at a total dose of 70 Gray in 35 fractions.

The evolution of the patient was marked by the degradation of his general condition, asthenia and cachexia, leading to sudden death 4 months after initial examination.

The patient only had one course of chemotherapy and radiation. No other investigations were performed other than a cardiac ultrasound showing an EF of 25%. The patient’s general state did not allow any aggressive treatment and did not tolerate a second course of chemotherapy. He received palliative care.

Clinical deterioration was sudden. Thus we did not discuss further treatment nor investigations.
We believe that the overall death was mainly due to the tumor, as it is known to be very aggressive and with poor treatment outcome. However, his comorbidities were an obstacle to surgery which was his best chance for survival.

3. Discussion

Carcinosarcoma has been described in the gastrointestinal tract, upper and lower respiratory tracts, urogenital tract, breast and skin [4,5]. However, its occurrence in the sinonasal tract is extremely rare [4]. The most common site for carcinosarcoma in the head and neck region is the parotid gland, followed by the submandibular gland [6]. Because of the rarity of primary nasosinusal carcinosarcoma, informations related to its prognosis are very limited in the literature. An American study [7] reported the largest cohort of sinonasal carcinosarcoma patients using the Surveillance, Epidemiology and End Results (SEER) database. This is the first attempt to estimate the long-term survival outcomes for the sinonasal carcinosarcoma patients.

Consistent with cases previously reported in the literature (Table 1), the sinonasal carcinosarcoma cohort had similar characteristics in terms of the age, race, tumor location, histologic grade and treatment. However, unlike previous reports, we found a higher percentage of affected females in this cohort (60.0%). The most frequent sinonasal location was the nasal cavity (46.7%), followed by the maxillary sinus (33.3%). One may surmise that sinonasal carcinosarcoma has a long-term prognosis similar to salivary gland carcinosarcoma, which is the most common site for this neoplasm. On the other hand, the 5- and 10-year DSS rates for the sinonasal carcinosarcoma cohort were found to be much lower than those for the case matched non sinonasal carcinosarcoma and carcinosarcoma at all other head and neck sites cohorts [7]. We believe that this could be due to the poor accessibility of the sinonasal tract for surgical intervention compared to the non-sinonasal regions. Furthermore, carcinosarcoma in the sinonasal region usually shows an aggressive and infiltrating behavior, whereas in the non-sinonasal areas such as in the larynx and pharynx, its growth pattern is often polypoid, leading to better surgical outcomes [8].

The treatment of choice for maxillary sinus carcinosarcoma is

| No | Age/sex | Stage | Treatment | Outcome | Author, year |
|----|---------|-------|-----------|---------|--------------|
| 1  | 62/F    |       | Radiotherapy | No marked improvement of the tumor, dead with disease | Meyer, 1957 [12] |
| 2  | 62/F    | T4N0M0 | Radiotherapy | Death in 40 months | Lichtiger, 1970 [13] |
| 3  | 71/M    | T4N0M0 | Preoperative radiotherapy + total maxillectomy + removal of eye | Death due to intracerebral abscess at postoperative period | Feinsmesser, 1982 [14] |
| 4  | 65/F    |       | Total maxillectomy + radiotherapy | Local recurrence, death 8 months later | Ampi, 1985 [15] |
| 5  | 57/F    | Tumor excision, ethmoidectomy and turbinectomy | Local recurrence 5 months after surgery | Hafiz, 1987 [16] |
| 6  | 60/M    | T3N0M0 | Total maxillectomy + radiation therapy + chemotherapy | Local recurrence, death 2 months later | Sonobe, 1989 [17] |
| 7  | 53/M    | T4N0M0 | Total maxillectomy + craniofacial resection + radiation therapy + chemotherapy | Disease free after 9 months | Shindo, 1990 [9] |
| 8  | 80/F    | T3N0M0 | Total maxillectomy + radiation therapy + 2nd operation | Local recurrence, death 2 months after second operation | Sanabre, 1998 [8] |
| 9  | 47/M    |       | Partial maxillectomy + radiation therapy | Local recurrence, death after 1 year | Furuta, 2001 [2] |
| 10 | 54/M    | T3N3M0 | Radiation therapy + chemotherapy | Death after 4 months | Howard, 2007 [10] |
| 11 | 60/M    | T3N0M0 | Total maxillectomy + radiation therapy + chemotherapy | Local recurrence | Jeong-Ki Moon, 2009 [18] |
| 12 | 52/M    | T4aN0M0 | Total maxillectomy + radiation therapy + chemotherapy | Local recurrence (soft palate) | Hisham B, 2012 [19] |
| 13 | 61/M    | T4aN0M0 | Total maxillectomy with a modified radical neck dissection. | Within 1 month of surgery, the patient developed a sternal metastasis, and he died within a short period of time | Cheong JP, 2014 [20] |
| 14 | 55/M    | T4aN0M0 | Radiotherapy | No significant effect, died 4 months after initial examination | Our case |
surgery with adjuvant radiotherapy [2,4,9]. Given the aggressive
and infiltrating nature of this tumor in the sinonasal region,
multimodal therapy is advised. Some authors think that irradiation
is ineffective, while others consider ‘radiation therapy’ is an
acceptable alternative for inoperable patients [10]. The role of
chemotherapy in the treatment of this tumor remains unclear.

From the known cases (Table 1), the most common treatment
modality was surgery with adjuvant radiotherapy (46.7%), followed
by radiotherapy alone (26.7%) and surgery alone (20.0%). In our
case, the patient consulted late with a large tumor which darkens
his prognosis. He received one session of radio-chemotherapy
because surgery was contraindicated (large extension of the mass
and ischemic heart disease), but no significant effect was obtained.

Prognosis is related to location, tumor size, depth of invasion
and stage of disease. Local recurrences and metastases are typically
lethal. An overall mortality rate of 42% at 30 months was reported
in carcinosarcoma at other sites [11]. Survival periods were from 2–
40 months and local recurrences eventually occurred in most cases.

This article is interesting in terms of the rarity of the case. Un-
fortunately, the patient consulted late. The main limitation of this
case is that we could not perform surgery that could have, poten-
tially, increased his survival chances.

4. Conclusion

Carcinosarcoma of the maxillary sinus is a rare and aggressive
neoplasm with poor survival outcomes compared to carcinosar-
comas in non-sinonasal head and neck regions. Based on our
analysis of historical data, it is clear that the primary treatment
modality for maxillary sinus carcinosarcoma is surgical interven-
tion with adjuvant radiotherapy.

Ethical approval

Patient approval is ok.

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Author contribution

Jawad Hasnaoui: Corresponding author writing the paper.
Mohammed Tatari: writing the paper.
Said Anajar: writing the paper.
Reda Abada: study concept.
Sami Rouadi: study concept.
Mohammed Roubal: correction of the paper.
Mohammed Mahtar: correction of the paper.

Conflicts of interest

All the authors have no personal or financial conflicts of interest
regard this case report.

Guarantor

Jawad Hasnaoui.

Consent

Written informed consent was obtained from the patient for
publication of this case report and accompanying image.

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Dr. Barhmi ismail was involved in the case writing and data
collection for the case report. He was also involved in critical review
and making corrections to the manuscript. Manuscript has been
‘splell-checked’ and ‘grammar-checked’.

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