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Case Report

Carcinosarcoma: a rare sinonasal malignancy with atypical behaviour

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ABSTRACT

Carcinosarcoma is an extremely rare neoplasm in the nasal cavity and in the paranasal sinus, with rapid growth and extensive local destruction. We report a case of a 61-year-old man with progressive left-side nasal obstruction and epiphora for six months. Imaging showed a massive mass lesion, mainly located in the left nasal cavity and ethmoid sinus, with no intracranial or orbital extension. Biopsy of the nasal mass revealed a poorly-differentiated squamous cell carcinoma associated with sarcomatous elements. Patient underwent an endoscopic transnasal resection. Definitive histopathologic analysis revealed a carcinosarcoma. Postoperative radiotherapy was, then, performed. After five years of follow-up, there is no evidence of tumor recurrence. This report highlights that even though despite the lack of optimal treatment protocol, long term disease control can be achieved through endoscopic transnasal resection complemented with radiotherapy.

Keywords: Carcinosarcoma, Nasal cavity, Paranasal sinuses, Nose neoplasms

INTRODUCTION

Sinonasal carcinosarcomas (also known as sinonasal sarcomatoid carcinoma) are a rare and aggressive type of cancer that occurs in nasal cavity and paranasal sinuses.[1,2] Histologically, this malignant comprising epithelial and mesenchymal component having sarcomatoid stroma.[3] Sinonasal carcinosarcomas typically have a rapid growth, extensive local destruction and high recurrence rate.[4,5]

Sinonasal carcinosarcomas are most commonly diagnosed in elderly individuals and are slightly more common in men than women.[6] The exact cause of sinonasal carcinosarcoma is not yet known, but it is believed to be associated with exposure to certain environmental toxins such as wood dust, leather dust and textile fibers.[7] Other risk factors include smoking, alcohol consumption, and a weakened immune system.[6,8]

Symptoms of sinonasal carcinosarcomas can be similar to those of other nasal and sinus conditions, and may include nasal obstruction, epistaxis (nosebleeds), sinusitis, facial pain/ pressure, and mass/ growth in the nasal cavity/ sinuses.[3,6] Because these symptoms nonspecific, diagnosis of sinonasal carcinosarcomas can be difficult.[9]

The current literature suggest that multimodal therapy is advised comprised surgery to remove the tumor, followed by radiation therapy and chemotherapy to destroy any remaining cancer cells.[4] However, because this type of cancer is so rare, there is no standard treatment protocol and the information related to its prognosis remain very limited.[1,4,10] Treatment plans are often tailored to each individual case based on the size and location of the tumor, as well as the patient's overall health.[10]

CASE REPORT

A 61-year-old man, first presented to primary care, was referred to our otolaryngology department with a six months history with progressive left-side nasal obstruction and epiphora. There was no history of epistaxis, headaches or diplopia. The patient had no significant comorbidities or history of tobacco use.

Endoscopic examination revealed an exophytic fleshy mass filling the left nasal cavity. No palpable lymphadenopathy was identified. In the ophthalmologic examination, he had a visual acuity of the right and left eye of 20/20 and had no impairment on ocular alignment or ocular movements and a normal ocular fundoscopy.

Computed tomography (CT) and magnetic resonance imaging (MRI) showed a massive mass lesion, mainly localized in the left nasal cavity and ethmoid sinus, with no intracranial or orbital extension (Figure 1 and 2). Biopsy of the nasal mass revealed a poorly-differentiated squamous cell carcinoma associated with sarcomatous elements. No metastasis lesions detected in neck CT.

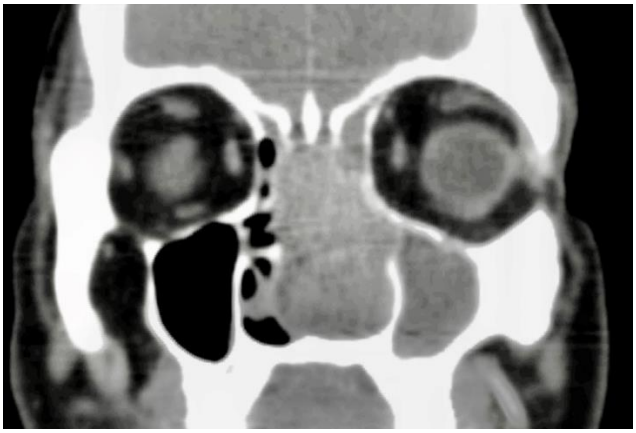

Figure 1: Coronal section of CT demonstrates a massive tumor occupying the left nasal cavity and ethmoid sinus, with no orbital extension.

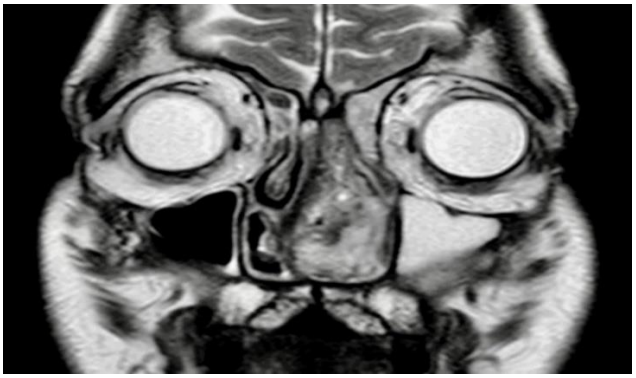

Figure 2: Coronal section of T2 -weighted MRI showing a heterogeneous and well-circumscribed mass lesion in left nasal cavity, with no orbital and intracranial extension.

After tumor board review, the neoplasm was completely resected by sinonasal endoscopic surgery and negative margins were achieve through frozen section analysis. Postoperative histopathologic examination revealed a biphasic tumor with sarcomatous and epithelial components (Figure 3), stained positively for cytokeratin 5/6 (CK 5/6), Cam 5.2, P40, and epithelial membrane antigen (EMA). It was requested an external validation of this analysis by academic institution, and the final pathologic diagnosis was carcinosarcoma. Adjuvant radiotherapy (Volumetric modulated arc therapy-VMAT) was performed with a total of 60 Gy. After five years of follow-up, there is no evidence of tumor recurrence (Figure 4).

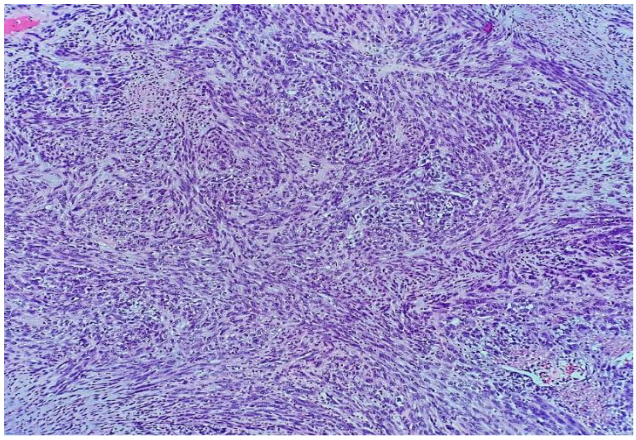

Figure 3: Histopathology features of the specimen, in hematoxylin and eosin stain, demonstrating epithelial and sarcomatous components.

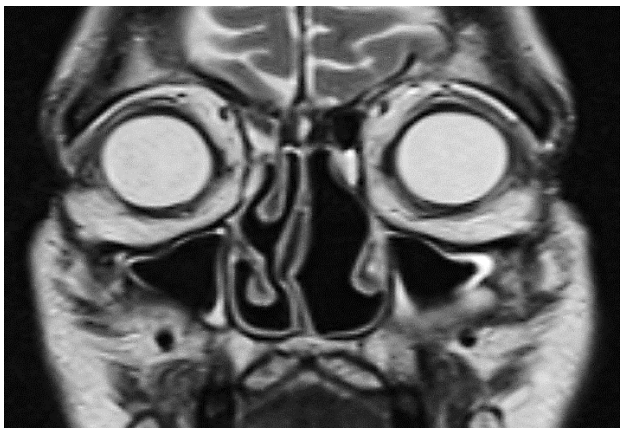

Figure 4: Coronal section of T2 -weighted MRI after five years of follow-up with no evidence of tumor recurrence.

DISCUSSION

Carcinosarcoma is a biphasic variant of sarcomatoid carcinoma that may arise from any squamous epithelium of the body such as respiratory tract, upper gastrointestinal tract, female reproductive organs.[1,2,4,8,9]In the head and neck region, the most common site is in the

salivatory glands.[1,3] This is an extremely rare and aggressive neoplasm in the sinonasal tract.[1-3]

Although the few cases reported in the literature, this tumor tends to present in the sixth decade of life with higher incidence in males with long history of tobacco use.[1,2] Patients usually develop symptoms related to the mass effect of the tumor such as nasal obstruction, epistaxis, facial pain and pressure sensation.[1,11] Similar to other sinonasal tract malignancies, there are nonspecific symptoms to the sinonasal carcinosarcomas.[6]

A recent case-control analysis performed by Patel et al obtained from authoritative source for cancer database in the United States (Surveillance, epidemiology and end results-SEER), found a total of 135 cases of head and neck carcinosarcomas in which, only fifteen cases involved the sinonasal tract.[1] In this analysis, the most frequent sinonasal carcinosarcoma subsite was the nasal cavity (46.7%) followed by maxillary sinus (33.3%). The same study also showed an average five-year disease-specific survival of 48.5%. The main poor prognostic factors are an extensive local destruction and skull base invasion.[2]

Early diagnosis and aggressive therapy are necessary to improve the prognosis.[2,7] Despite that, due to its rarity, there is a lack of available evidence about its best management.[2,7,11] The current literature suggests that the treatment of choice is surgery with adjuvant radiotherapy.[1,2,5,10] The role of chemotherapy in the treatment of this condition remains unclear.[1]

Here, we present a case of extensive sinonasal carcinosarcoma who had a suspicion and early detection, with long term disease control achieved through endoscopic surgical resection complemented with radiotherapy.

CONCLUSION

Carcinosarcoma is rare and aggressive malignancy in nasal cavity and paranasal sinus. Even though despite the lack of optimal treatment protocol, in earlier diagnosis, long term disease control can be achieved through endoscopic surgical resection complemented with radiotherapy.

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