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Case Report

A Rare Malignancy of Head And Neck Region: Sinonasal Undifferentiated Carcinoma

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ABSTRACT

Sinonasal undifferentiated carcinoma is a rare malignancy of the head and neck region. Its diagnosis and treatment are difficult due to its rare and aggressive tumor nature and the complex anatomy of its localization. A 70-year-old male who presented with symptoms caused by this rare tumor was reported. The patient presented with pain on the left side of the head and vision loss in the left eye for 1 month, and his endoscopic biopsy was reported as undifferentiated carcinoma. This case report aimed to discuss the diagnosis and treatment of sinonasal undifferentiated carcinoma.

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Introduction

Sinonasal undifferentiated carcinomas (SUIC) are rare malignancies of the head and neck region, constituting less than %3 of all head and neck tumors with an incidence of 0.556 cases per 100.000 people per year [1]. SUIC presents unique treatment challenges for surgeons due to their rarity, difficult anatomical localization, histological heterogeneity, and tendency for recurrence. SNUC usually involves the elder individuals of the population with a slight male predominance (2-3:1) [2, 3].

SUIC is different from other sinonasal malignancies as it is hypothesized to arise from the Schneiderian epithelium or the nasal ectoderm of the paranasal sinuses [4]. It's a highly aggressive tumor extending beyond the sinonasal tract to the skull-base, orbit, and intracranium. Following diagnosis, treatment protocols usually include a combination of multi-disciplinary approaches occurring of surgery, radiation, and/or chemotherapy, which are decided depending upon various clinicopathologic factors. The prognosis of SUIC is poor, reporting 5-

year overall survival (OS) rates of 46% to 67% according to prior large population-based studies [5-7].

Case Report

A 70-year-old man presented to the emergency room with loss of vision in the left eye and a left-sided headache with a history of one month. He indicated that these complaints were minor but increased day by day. He had a cataract operation on both of his eyes 2 months ago and he also stated he realized he got double vision after the operation. The first emergency room consultation was neurology because of possible etiologies for headaches such as temporal arteritis that came to their minds in the first place. The diffusion MRI scan reported "a soft tissue mass which located at the left temporal zone of brain anteromedially, showing extension to the cavernous sinus, left ethmoidal cells and superior orbital fissure, contacts with left ICA, lobulated and sized 54x40 mm; with diffuse hyperintense signal enhancement at left temporal lobe subcortical white matter in FLAIR sequences" (Figures 1A & 1B).

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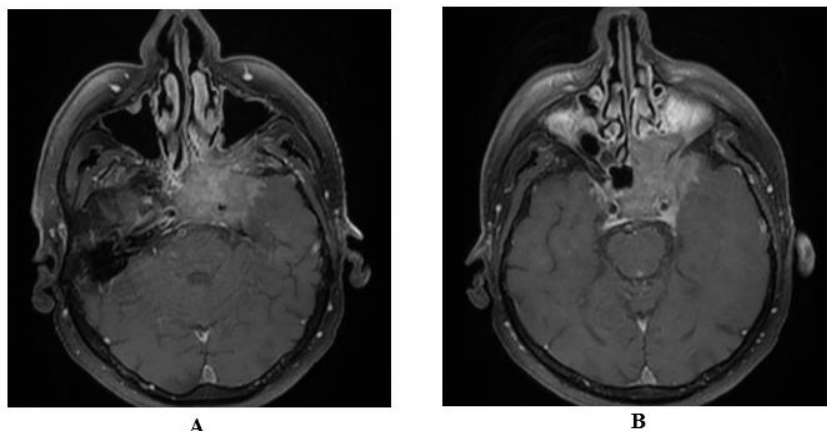


Figure 1: A & B) Contrast-enhanced cranial MRI findings from first admission to hospital reveals the mass originated from left sphenoid sinus.

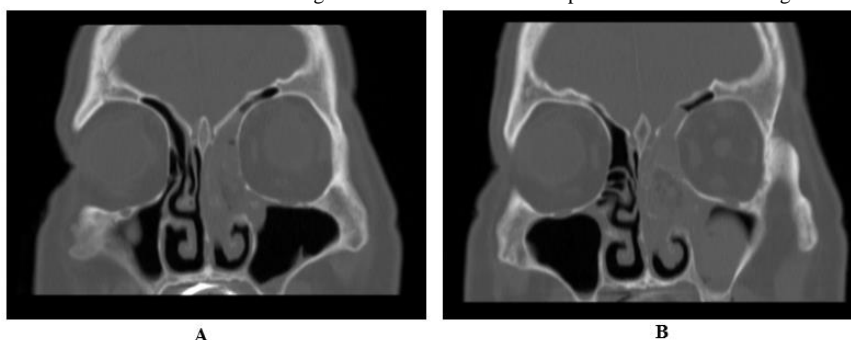


Figure 2: A & B) Preoperative paranasal CT imaging showing extension of tumor.

During the physical examination of the patient, he was conscious and oriented. There was mild proptosis, loss of vision in his left eye, and also he noted that he got continuous pain on his left side of the head, left eye, and behind it. His cranial nerve evaluation showed the 1-3-4-6th cranial nerves were affected. Only purulent rhinitis was noted and no masses were seen on the nasal cavity or nasopharynx. Other ENT examinations were normal.

After the evaluation was completed, the patient was hospitalized for further assessments and an endoscopic sinus surgery was planned for biopsy. Preoperative facial and nasopharynx MRI revealed an irregular, distinctly contrasting, lobulated, and infiltrative 44x37 mm sized mass extended to the left minor ala of sphenoid at skull base level, posterior ethmoid cells, left optic nerve, retroorbital fat mass, left temporal lobe, and the middle cranial fossa. The patient underwent surgery under the precautions of COVID-19. During the operation left maxillary sinus ostium and left grand lamella were seen infiltrated by tumor similar to

preoperative paranasal CT imaging (Figure 2A & 2B). The frozen section diagnosis was reported as “undifferentiated carcinoma”. Afterward, the operation ended without any other intervention. Three days later patient's vision complaints are increased and consulted to ophthalmology department. They stated there was no pupil reflex on the left and minimal on the right eye, no vision on left and 7/10 on the right eye, no movement in the left eye with conjunctival edema and hyperemia.

Later on, the patient discussed with our center's neurosurgery department. They had the conclusion about the inoperability of patient neurosurgically. On the same day, pathology results were reported as “sinonasal undifferentiated carcinoma” (Figure 4). After then the patient was referred to the medical oncology department. However, the general condition of the patient deteriorated rapidly within days due to progression of mass, which also could be seen on MRI scans later (Figures 3A & 3B) and he died due to respiratory failure.

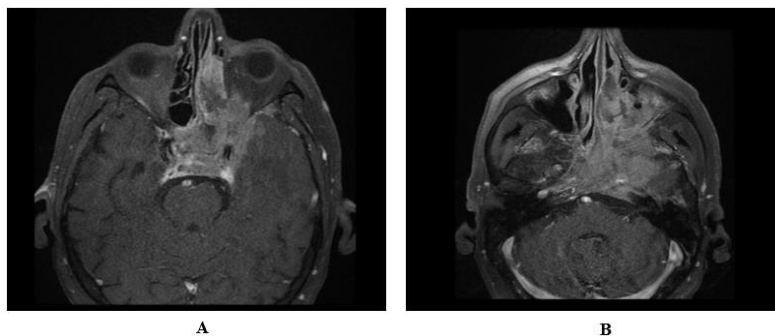


Figure 3: A & B) Second contrast enhanced MRI imaging , note the progression and extension of mass in 2 weeks.

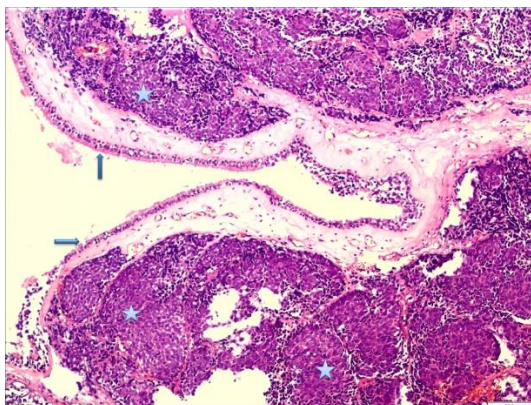


Figure 4: Histopathological image of tumor. Arrows represent normal sinus mucosa, stars represent solid islands of tumoral infiltration; (H&E x40).

Discussion and Conclusion

SUIC is a rare and extremely malignant tumor of the sinonasal cavity. Treatment outcomes are very poor. Patients who receive aggressive multimodality treatments have improved survival. The surgery seems to be an important part of the treatment. The use of neoadjuvant chemotherapy may be associated with improved outcomes and can be used to guide the subsequent treatment selection [8]. It is controversial whether RT should be given preoperatively or postoperatively. However, it seems to be an indisputable fact that it is a tumor that requires trimodal treatment [9]. Due to its silent, progressive and invasive nature with the patient's poor description of symptoms can cause a delay in the diagnosis of the tumors in the COVID-19 era. Our aim, in this case, is to increase the awareness of these relatively rare tumors and make a contribution with our clinical data to the ENT community.

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