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Sinonasal Teratocarcinosarcoma with Orbital and Anterior Skull Base Involvement: A Contrast-Enhanced CT-Based Case Report

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Abstract: ***Background:*** ***Sinonasal teratocarcinosarcoma (SNTCS) is a rare and aggressive malignant tumour of the sinonasal tract (SNT) with a prevalence of less than 1% of SNT malignancies. The diagnosis may be delayed since it is a heterogeneous histopathological lesion with non-specific symptoms. Imaging has an important role and adds significant value when used for determining the extent of the disease and directing management.*** ***Case Presentation:*** ***There was a 53-year-old man who had been having progressive bilateral nasal obstruction and epistaxis for 3 months. Computed tomography (CT) revealed a soft tissue-density mass lesion in the right nasal cavity that had heterogeneous enhancement. It severely damaged the sinonasal skeleton, specifically the lamina papyracea, orbital floor, sphenoid bone, and anterior skull base. The lesion involved the orbit and had extra-axial intracranial extension. Histopathological diagnosis showed sinonasal teratocarcinosarcoma. Extensive local invasion of the lesion was noted, and management was a combination of optimal chemotherapy and radiation therapy with external beam radiotherapy (EBRT) and specific drugs- paclitaxel and carboplatin.*** ***Case Summary:*** ***This case illustrates the typical CT appearance of SNTCS, the extent of the tumour and the use of contrast-enhanced CT to assist in the management.***

Keywords: Sinonasal teratocarcinosarcoma, Contrast-enhanced computed tomography, Sinonasal malignancy, Skull base invasion, Orbital invasion

1. Introduction

Sinonasal teratocarcinosarcoma (SNTCS) is a relatively rare and very aggressive malignant tumor of the nasal cavity and the paranasal sinuses. Less than 200 cases have been reported since Heffner and Hyams first described the condition in 1984 [1,3,6,8]. Predominantly, it affects males in the 5th and 6th decades of life and, most importantly, begins in the nasal cavity and ethmoid sinus [1,3,6,9]. The symptoms are generally vague and patients commonly experience progressive nasal blockage, epistaxis, facial pain, headache, anosmia, and/or visual symptoms [3,6,7,8]. These symptoms are common to benign sinonasal disease, which results in a delay of diagnosis, often until it has invaded the adjacent orbit or skull base. Cross-sectional imaging is a big component in assessing SNTCS. Contrast-enhanced CT is of great value in the evaluation of bone destruction, erosion of the skull base and involvement of the sinonasal cavity, while MRI is excellent in evaluating perineural extension and intracranial and orbital extension [3,4,5,6,8,9]. A good radiological evaluation is critical to staging, resectability, and the planning of treatment.

A histopathologically confirmed case of SNTCS in a 53-year-old man with extensive orbital and skull base involvement is presented. This case illustrates the typical contrast-enhanced computed tomography (CT) findings of this rare and aggressive malignancy and underscores the role imaging plays in the diagnosis and management of this disease.

2. Case Report

A 53-year-old man presented with 3 months of nasal obstruction and epistaxis. During the clinical examination, a mass was discovered in the nose, on the right side, and a contrast-enhanced computed tomography (CT) of the brain and paranasal sinuses was performed. CT revealed a heterogeneously enhancing mass lesion of soft tissue density in the right nasal cavity measuring 49×37×52 mm. The lesion destroyed the right superior, middle and inferior turbinates. It involves the nasal septum and extends into the left nasal cavity to a minimum degree. Superiorly, it invades the bilateral ethmoid sinuses along with mucosal thickening of sinuses. There is destruction of the posterolateral and medial wall of the right maxillary sinus with extension into it. Eroded portions of the sphenoid bone and extension into the sphenoid sinus were observed posteriorly. Furthermore, it damages the right lamina papyracea, orbital floor and right frontal process of the maxilla and right nasal bone. Intraorbital extension is seen as loss of fat planes around the right medial rectus, inferior rectus and inferior oblique muscles. The optic nerve on the right side had been pushed laterally. There is a mild proptosis of the right eye and anterolateral displacement of globe is noted. Superiorly the lesion extends into the region of superior orbital fissure. This lesion erodes the base of skull anteriorly and shows that it has overgrown into the right frontal region of the skull with extra-axial intracranial extension.

An aggressive sinonasal malignant tumor with involvement of the orbit and anterior skull base was suspected based upon these imaging findings. Endoscopic biopsy specimen showed

a high-grade poorly differentiated malignant neoplasm. The tumor shows nests of tumor cells with squamoid morphology, admixed with groups of undifferentiated small round cells with hyperchromatic nuclei, scant cytoplasm, and frequent mitotic figures. A primitive mesenchymal stromal component with spindle cells and myxoid matrix is present and there is marked necrosis. Thus, these results were consistent with the diagnosis of sinonasal teratocarcinosarcoma [1,2,3,6,9].

A lesion with such extensive local invasion with involvement of the orbit and anterior skull base was not resectable. Definitive external beam radiotherapy and concurrent chemotherapy with paclitaxel-carboplatin was administered to the patient. Pegfilgrastim was the supportive therapy post-chemotherapy. No follow-up imaging was available at the time of manuscript preparation with which to evaluate the response to therapy.

3. Imaging Findings

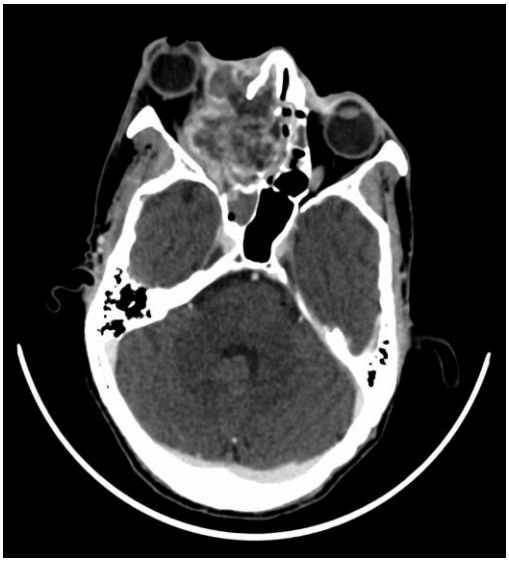

Figure 1: Axial contrast-enhanced CT image showing a heterogeneously enhancing soft-tissue mass centered in the right nasal cavity with destruction of the right nasal turbinates and erosion of the nasal septum

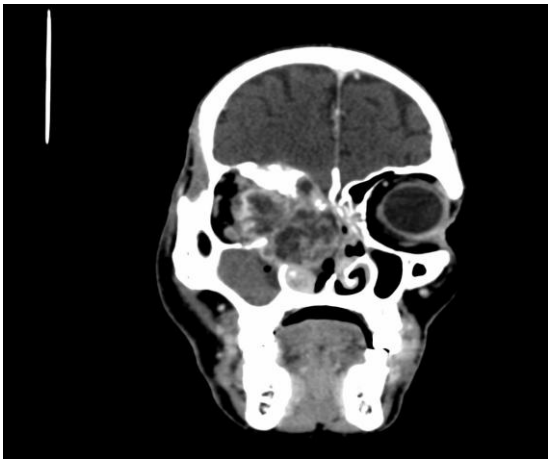

Figure 2: Coronal contrast-enhanced CT image showing destruction of the right lamina papyracea and orbital floor with intraorbital extension involving the medial orbital compartment

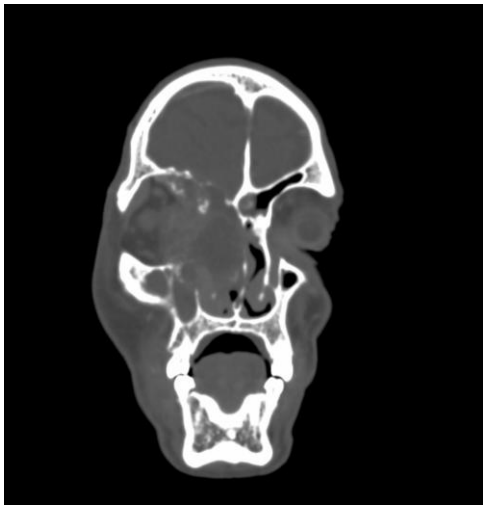

Figure 3: Coronal bone-window CT image demonstrating extensive osseous destruction involving the frontal process of the right maxilla, sphenoid bone, and anterior skull base.

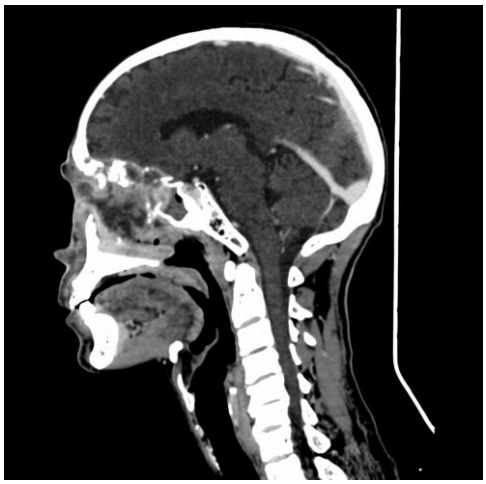

Figure 4: Sagittal contrast-enhanced CT image showing erosion of the anterior skull base with extra-axial intracranial extension into the right frontal region

4. Discussion

The Sinonasal teratocarcinosarcoma (SNTCS) is a very rare and aggressive form of cancer. Diagnosis may be delayed because of rarity and vague clinical appearance, often only being diagnosed when there is marked invasion at a later stage and rapid local spread [1,3,6,8,9]. It originates from the nasal cavity and ethmoid sinus and has a high tendency to invade surrounding structures, such as the orbit, base of the skull and anterior cranial fossa [1,3,6,9]. Imaging is of crucial importance in the evaluation of SNTCS. The use of contrast-enhanced CT is invaluable for defining the primary tumour location, disease spread and aggressive osseous destruction, and MRI is superior to CT for intracranial, orbital, and perineural extension [3,4,6,8,9]. In this case, a heterogeneously enhancing mass was seen in the right nasal cavity and destruction of the turbinates, nasal septum, maxillary sinus walls, sphenoid bone, lamina papyracea, orbital floor and anterior skull base was seen on the CT scan. Intraorbital extension involving extraocular muscles, involvement of the superior orbital fissure and extension into the extra-axial intracranial compartment suggested advanced locoregional disease and had a significant impact on treatment planning. Imaging

differential diagnosis for such an aggressive sinonasal mass includes sinonasal undifferentiated carcinoma, olfactory neuroblastoma, squamous cell carcinoma, lymphoma, and high-grade adenocarcinoma of the nasal cavity or paranasal sinuses [3,6,9]. While most of these entities can show a similar highly aggressive appearance on the imaging studies, if there is extensive bone destruction with invasion of the orbital region or the skull base, then high-grade sinonasal malignancy should be considered. But imaging characteristics are nonspecific, and a definitive diagnosis requires histopathological evaluation. Histologically, SNTCS is characterized by a complex admixture of malignant epithelial, neuroectodermal, and mesenchymal components, often with primitive or teratoid elements [1,2,3,6,9]. Histopathology in our patient confirmed SNTCS with the presence of squamoid epithelial nests, undifferentiated small round cells, and primitive mesenchymal stroma. Immunohistochemistry and correlation with the WHO classification of head and neck tumours further support the diagnosis [2,9] . It has an aggressive clinical course with high local recurrence risk, with a median clinically follow-up period of 41 months. When possible, multimodality treatment using full surgical removal and either radiotherapy or chemotherapy is usually the recommended course of treatment. Total surgical resection is, however, often not possible in cases of more extensive elaboration in the orbit or the base of the skull. In the current scenario, complete resection may not be achievable in cases with extensive invasion of the orbit, skull base, or intracranial compartment, and in such situations, definitive concurrent chemoradiotherapy (CCRT) may be the only viable option [4–6,8]. The recognition of typical CT features becomes important to achieve early accurate staging, assessment of resectability and timely management of the disease.

In our patient, the tumour exhibited extensive local invasion, including orbital and anterior skull base involvement, rendering it unresectable. Definitive EBRT with concurrent paclitaxel-carboplatin chemotherapy was therefore chosen as the primary treatment modality, in line with regimens reported in previous case series and systematic reviews [4–6,8]. Although follow-up imaging was not available at the time of writing, early recognition of the characteristic CT features in this case was crucial for appropriate staging, assessment of resectability, and timely initiation of multimodality therapy.

5. Conclusion

Sinonasal teratocarcinosarcoma is an aggressive rare type of sinonasal cancer, which is often already highly advanced, due to the rather vague symptoms. Contrast-enhanced computed tomography (CT) has a key role within the context of obtaining accurate delineation of tumor extent, advanced proof of osseous destruction, display of orbital invasion and involvement of the skull base, and disease staging and treatment planning. Histopathological examination is still crucial for the definitive diagnosis. Prompt diagnosis of characteristic imaging features and multidisciplinary treatment can enhance the outcome of this rare and highly malignant tumour.

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