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Case Report

A Rare Case of Sino-Nasal Teratocarcinosarcoma (Response to Induction Chemotherapy)

Muhammad Haris Rauf, Asma Asghar, Ali Zafar Sheikh, Anis-ur-Rehman, Kashif Siddique
Department of Radiology, Shaukat Khanum Memorial Cancer Hospital & Research Centre, Lahore Pakistan

ABSTRACT

We are presenting a rare case of sino-nasal teratocarcinosarcoma. A 43-year-old male patient presented with repeated episodes of epistaxis and headache for 3 months. On examination, the ENT consultant noticed a mass in the right nasal cavity for which biopsy was performed and CT scan advised. Sinonasal teratocarcinosarcoma was diagnosed on biopsy and immunohistochemistry. CT Face followed by PET/CT showed a FDG-avid soft tissue density mass in the right nasal cavity extending locally with a standardised uptake value of 7.4 with no distant metastasis. Surgical debulking is the first line of treatment as per the literature; however, in this case, the multidisciplinary tumour board initiated induction chemotherapy of 02 cycles of CISP/DOC owing to marked local extension of the tumour. Subsequent CT scan of the face shows a good treatment response with shrinkage of the sinonasal mass. After successful chemotherapy, the patient is now scheduled for radiotherapy and subsequently follow-up to assess the response.

Keywords: Sinonasal teratocarcinoma, Epithelial and mesenchymal elements, Biopsy and immunohistochemistry, Adjuvant chemotherapy, Chemotherapy cycles CISP/DOC.

How to Cite This Article: *Rauf MH, Asghar A, Sheikh AZ, Rehman AU, Siddique K. A Rare Case of Sino-Nasal Teratocarcinosarcoma (Response to Induction Chemotherapy). Pak Armed Forces Med J 2024; 74(3): 892-894 DOI: https://doi.org/10.51253/pafmj.v74i3.12104*

INTRODUCTION

Sinonasal teratocarcinosarcoma (SNTCS) is an extremely rare malignant growth that arises in the nasal cavity. It is a morphologically heterogeneous tumour with distinct histology characterised by the presence of epithelial (skin and lining of internal organs), mesenchymal (bone, cartilage and fat) and primitive neuroepithelial elements.[1] The overall incidence of SNTCS is extremely low, with fewer than 150 cases reported in the literature. [2] It represents approximately 3% of all head and neck malignancies and less than 1% of all cancers. It tends to affect the adult population with a mean age of 50 years, predominantly seen in males, with male to female ratio of 8:1. It is highly aggressive tumour with predominantly local extension is seen however intra-orbital extension is also reported in 20% of cases. To date, management protocols mentioned in the literature include debulking surgery followed by adjuvant radiotherapy, but the incidence of local recurrence is 38%.[3] We are presenting a rare case of sino-nasal teratocarcinosarcoma.

CASE REPORT

A 43-year-old young male presented with complaints of nasal obstruction, headaches, and recurrent episodes of epistaxis for 03 months. On examination by an ENT specialist (outside hospital), a lobulated growth is observed in the right nasal cavity. Biopsy was performed on the right sinonasal mass, and imaging was suggested to determine its extent. Biopsy specimens were evaluated at the Shaukat Khanam Memorial Cancer Hospital & Research Centre (SKMCH&RC) in Lahore Pakistan (Figure-1).

Immuno/ Histochemical Stain(S):

SMARCA4:	Loss of expression in tumor cells
Synaptophysin:	Positive in neuroepithelial component
p63:	Positive in squamous component
SNOMED:	T-21000 M-8000/3
Diagnosis:	Right side sinonasal mass: Teratocarcinosarcoma, fragmented.

Figure-1: Immunohistochemistry Report of the Patient

A contrast-enhanced computed tomography (CT) scan of the face was performed at the Department of Diagnostic Radiology (PET/CT Hybrid Imaging Unit), SKMCH & RC, Lahore Pakistan, and showed heterogeneously enhancing soft tissue density mass with epicentre in the right nasal cavity extending into the right maxillary, ethmoidal, and sphenoidal sinuses. Posteriorly, it extends into the nasopharynx with an associated mass effect and involvement of the left nasal cavity (Figure-2: A, C & D). No locoregional lymphadenopathy was noted. A PET scan was ordered to see any distant metastasis, which showed an FDG-avid soft tissue density mass in the right nasal

Correspondence: Dr. Muhammad Haris Rauf, Shaukat Khanum Memorial Cancer Hospital and Research Centre, Lahore Pakistan
Received: 22 Apr 2024, revision received: 13 May 2024; accepted: 30 May 2024

cavity extending into the ipsilateral maxillary antrum and ethmoid air cells, sphenoid sinus, frontal sinus, and nasopharynx with a standardised uptake value (SUV) 7.4 Figure-2: (B). No FDG-avid locoregional lymph nodes with avid distant osseous or visceral metastases were observed.

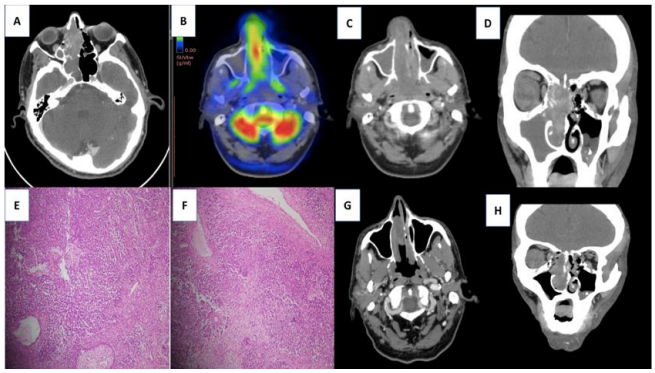

Figure-2: A,C & D: Right Nasal Cavity Mass extending into ipsilateral sinuses, nasopharynx & left nasal cavity

B: PET scan showing FDG Avid mass in right nasal cavity (SUV:7.4)

E&F: Histopathology slides from the biopsy of lesion showing primitive neuroepithelial elements, atypical squamous nests, atypical spindled cells with areas of myxoid changes

G&H: Post treatment scans after 6 weeks of Induction Chemotherapy showing marked interval decrease in size of Rt Sino nasal tumor resulting in restoration of nasopharyngeal air column and complete pneumatization of Rt. maxillary sinus.

In the literature, the widely accepted treatment for this tumour is surgery (de-bulking) followed by adjuvant radiotherapy[3]. In the multidisciplinary team meeting, initial treatment with induction chemo-therapy was suggested to determine the response. Two cycles of induction chemotherapy Cisplatin/ Docetaxel were administered, and a follow-up CT scan was performed after 6 weeks, which showed markedly good treatment response in terms of shrinkage in size of the right sino-nasal mass with complete pneumatization of the right maxillary sinus noted. On the basis of a good response to induction chemo-therapy, Radiotherapy was planned in a multi-disciplinary meeting, and follow-up was proposed with imaging.

DISCUSSION

Sino-nasal teratocarcinosarcoma (SNTC) is a rare malignant tumour of the anterior skull base that comprises benign neural elements and malignant epithelial and mesenchymal components.[1] The carcinoma can be adenocarcinoma or squamous carcinoma, and the mesenchymal component includes smooth, skeletal muscle, spindle, bone, and cartilage features.[4] The common sites of involvement as per decreasing order of frequency include nasal cavity's superior aspect (79% of all cases approx.), paranasal sinuses, orbits, nasopharynx, pharyngeal wall followed by skull base, cribriform plate, and anterior cranial fossa respectively. [2] SNTCS has a particularly low overall incidence, with 150 reported cases in the literature so far. [5] It represents approximately 3% of all head and neck malignancies and less than 1% of all cancers. It tends to affect the adult population with a mean age of 50 years and a male-to-female ratio of 8:1. Owing to its rarity and lack of established therapies, it is challenging to determine the ideal treatment strategies to ensure complete cure with no recurrence. Because of the tumour's aggressive nature and propensity to recur, even after radical surgery and adjuvant radio-therapy, a close follow-up is strongly recommended.[6] For recurrent and metastatic disease, chemotherapy is also included in the treatment regimen. However, even after using bimodal or trimodal protocols, there is no substantial evidence that such treatment will increase the overall cure rate and survival. The recurrence rate was 38% with a mean recurrence time of 19.5 months. After surgical excision, 3- and 5-year survival rates are 30% and 20%.[1]

SNTC is a rare, highly aggressive tumour with distinct histology. It has a high incidence of local recurrence after surgical excision alone. Therefore, adjuvant chemoradiotherapy should always be considered to control its recurrence. Response to induction chemotherapy may also be helpful in reducing the size of locally invasive tumours in which a surgical approach is difficult.

Conflict of Interest: None.

Authors Contribution

Following authors have made substantial contributions to the manuscript as under:

MHR: Conception, study design, drafting the manuscript, approval of the final version to be published.

AA & AZS: Data acquisition, drafting the manuscript, critical review, approval of the final version to be published.

AR & KS: Data acquisition, critical review, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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