

Case Report

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A rare case report of sinonasal undifferentiated carcinoma of paranasal sinuses

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ABSTRACT

Sino nasal malignancies account for only 0.2-0.8% of all malignancies and undifferentiated carcinoma is rare malignant tumour of sinonasal tract, with extremely poor prognosis. We report a case of sinonasal undifferentiated carcinoma which we managed by surgery followed by post-operative radiotherapy with concomitant platinum-based chemotherapy. Although the overall survival is about 20% at 5 years and there are frequent recurrences combined modality treatment is the best management option available at present.

Keywords: Sinonasal undifferentiated carcinoma, Maxillectomy, Blue small round cell tumors

INTRODUCTION

Sino nasal malignancies are rare malignancies with an incidence of 0.5-1 per 100 000 per year and account for only 0.2-0.8% of all malignancies and 3% of upper aero digestive tract neoplasm.^{1,2} Mostly develop in the fifth and sixth decades of life.³ Sinonasal undifferentiated carcinoma was described relatively recently by Frierson et al.⁴ It is otherwise known as anaplastic carcinoma and is hard to distinguish from high-grade olfactory esthesioneuroblastoma. It is a highly aggressive and invasive tumour produces rather subtle symptoms initially despite its extensive nature.⁵ Sino nasal undifferentiated carcinoma is believed to originate from Schneiderian epithelium or from the nasal ectoderm of the paranasal sinuses.⁴ It typically presents as a rapidly enlarging tumor mass involving multiple (sinonasal tract) sites, often with an evidence of extension beyond the anatomic confines of the sinonasal tract.

CASE REPORT

A 42-year-old female presented to ENT OPD with complaint of left sided nasal obstruction and bleeding from left nasal cavity since past 3 months. On left side nasal obstruction was insidious in onset progressive in

nature. History of left sided nasal bleed, spontaneous, intermittent, 2-3 episodes in a week self-limiting in nature. Patient had associated symptom of blood-tinged foul smelling anterior nasal discharge and posterior nasal discharge. There was associated occasional dull aching pain on left side of face and excessive watering from left eye also.

On DNE- left nasal cavity-A single firm friable yellowish mass was present in left nasal cavity between the septum and middle turbinate arising from middle meatus, which bled on touch, probe could not be passed superiorly and laterally. Deviated nasal septum to left with septal spur inferiorly was noted.

Visual acuity and extra ocular movements were normal, Bilateral pupillary reflex (direct and consensual) were equally reactive to light.

Non contrast computer tomogram (NCCT NOSE PNS) both coronal and axial cuts. Findings were, soft tissue density filling left nasal cavity, anterior and posterior ethmoids, left maxillary sinus, blocking left osteomeatal complex, erosions were noted at the level of horizontal and vertical lamella of cribriform plate, left lamina erosion was noted but no involvement of periorbita.

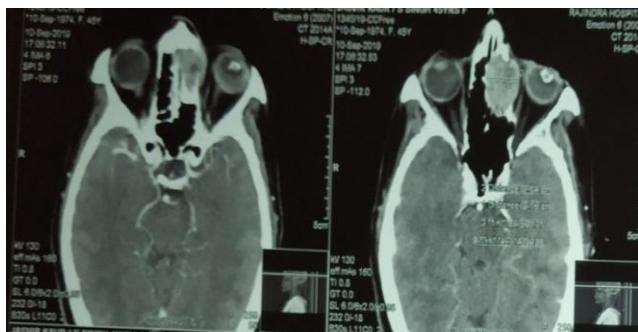


Figure 1: NCCT nose PNS axial sections of soft tissue density material filling left nasal cavity ethmoids and eroding lamina papyracea.

Patient underwent diagnostic biopsy under GA, tissue was sent for HPE.

HPE showed tumor cells with cylindrical basaloid morphology lying in papillary, ribbon like pattern with central necrosis. Cells exhibit marked nuclear atypia and brisk mitotic activity. Tumor is devoid of significant keratinisation. HPE features are those of Non keratinizing transitional cell carcinoma of nasal cavity

In view of malignant nature of lesion, patient was planned for excision via lateral rhinotomy approach followed by post op radiotherapy. Pre-operative MRI was done and was s/o tumor extending intracranially but extradural.



Figure 2: MRI of heterogeneous mass which is hypointense on T2WI axial sections with multiple hyperintense areas suggestive of necrosis in ethmoid and frontal sinus extending into left orbit with intact periorbita.

Under IPPV via oral intubation, cleaning and draping was done lateral rhinotomy incision was made dissection continued in subperiosteal plane, anterior maxillary wall was exposed, medial maxillectomy was carried out, tumor was found to be extending from left ethmoid sinus, left frontal sinus and contralateral frontal sinus by eroding interferential septum. However, periorbita was intact but was removed as margin, superiorly tumor was followed in frontal sinus which was removed, frontal sinus mucosa was stripped, upper part of septum was removed, floor of ipsilateral and contralateral frontal sinus removed (modified Lothrop procedure). Sphenoid sinus was identified and was found to be clear of disease.

Excised tissue and margin sent for HPE. Haemostasis was secured, post-operative period was uneventful.

HPE showed invasive tumor arranged in diffused sheets and in perivascular pattern. Tumor cells showing moderate to marked anaplasia with vesicular to hyperchromatic nuclei with prominent nucleoli with high atypical mitotic activity. Tumor giant cells were also seen. Features suggestive of undifferentiated malignant tumor.

Immunohistochemistry was reactive for CD56, CK and CDX-2, suggestive of sino-nasal undifferentiated carcinoma.

Patient then underwent external beam radiotherapy 66 Gy in 30 fractions in 6 weeks by volumetric modulated arc radiotherapy. PET CT was after radiotherapy and was suggestive of FDG avid soft tissue thickening left ethmoid sinus but no metabolically active lymph nodes. Re-surgery was not possible was hence planned with chemotherapy with cisplatin and etoposide. Patient was symptomatically relieved.

Later patient developed cutaneous fistula communicating from left nasolabial fold to nasal cavity.



Figure 3: Left medial maxillectomy being performed via lateral rhinotomy approach.

DISCUSSION

Sinonasal undifferentiated carcinoma of para nasal sinuses is extremely rare accounting for <3% of all head and neck tumors and 0.2-0.8% of all malignancies.^{1,2}

It presents with rapid onset of symptoms such as nasal obstruction, nasal discharge, proptosis, diplopia, diminution of vision, epistaxis, and facial pain resembling any benign lesion, resulting in delay of appropriate treatment.⁴ Therefore, it becomes important to distinguish it from other sinonasal lesions because of its destructive behavior and dismal clinical outcome.

It is one of the important differential of “blue round cell tumors” in the sinonasal tract along with squamous carcinoma, nasopharyngeal carcinoma, lymphoma, melanoma, olfactory neuroblastoma, hemangiopericytoma and small cell neuroendocrine carcinoma. Thus, immunohistochemical studies are

crucial for differentiating between these entities. Radiologic findings show a more anterior involvement of the nasal cavity and ethmoids by sinonasal undifferentiated carcinoma as compared to nasopharyngeal carcinoma.⁶

Multimodality therapy (surgery, radiotherapy and chemotherapy) has generally been demonstrated to be the most effective approach in the treatment of squamous cell carcinoma and sinonasal undifferentiated carcinoma. Due to its extension outside the natural anatomical boundaries of the nasal cavity and PNS, results in morbid surgery which include maxillectomy, orbital exenteration and craniotomies.⁷⁻⁹

If operable surgery followed by post-operative radiotherapy with concomitant platinum-based chemotherapy is the most common approach. In large volume tumours initial non-surgical treatment with (chemo)radiotherapy or chemotherapy alone followed by chemoradiotherapy appears to give better results.^{10,11} Now a day's treatment with proton beam radiotherapy where available is also coming into picture, with the possibility of reducing radiation-related morbidity.¹²

Similarly, our case also underwent surgical resection followed by postoperative radiotherapy and chemotherapy sessions.

CONCLUSION

Sino nasal undifferentiated carcinoma is rare malignant tumour of sinonasal tract, with extremely poor prognosis. The overall survival is about 20% at 5 years. There is frequent recurrence with metastasis to lymph nodes and distant sites. Combined modality treatment is the best management option which is Surgical excision followed by adjuvant chemotherapy or radiotherapy.

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