

CASE REPORT

Ewing's Sarcoma of the sinonasal tract: Case Report.Waqas Javaid¹, Maria Asif², Mehak Ahmad³, Ayesha Fayyaz⁴, Aamna Durrani⁵, Syed Muhammad Waqas⁶

ABSTRACT... Ewing sarcoma (ES) is a highly malignant, poorly differentiated neuroectodermal neoplasm that predominantly arises within the medullary cavity of long bones in children and adolescents. Extraosseous Ewing sarcoma (EES) affecting the head and neck region is exceedingly rare, accounting for only 1% to 4% of all cases, with primary sinonasal tract localization representing a severe diagnostic and therapeutic challenge.

Key words: Ewing Sarcoma Family of Tumors (ESFT), Extraosseous Ewing Sarcoma (EES), Sinonasal Malignancy, Small Round Blue Cell Tumors (SRBCT).

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INTRODUCTION

Primary Ewing's sarcoma of the sinonasal tract is an exceedingly rare and highly aggressive small round blue cell malignancy/ neuroectodermal malignancy. It accounts for less than 1-4 % of all head and neck tumors.¹ Because it rarely presents in epithelial tracts, it is frequently misdiagnosed as other small round blue cell tumors, such as olfactory neuroblastoma, lymphoma, or rhabdomyosarcoma. Patients who present with metastatic disease have decreased five-year survival rates, which range from 21% to 70%.² Treatment options include multimodal approach i.e. chemotherapy, radiation, and surgery.^{3,4}

Below are two distinct case reports from the ENT department of Sir Ganga Ram Hospital Lahore, highlighting the diagnostic challenges, varied clinical presentations, and critical need for multidisciplinary management.

Case Report 1

A 30-year-old female presented to the ENT Emergency department of Sir Ganga Ram Hospital Lahore in September 2025. She presented with a 6-month history of profuse, recurrent epistaxis from the left nasal cavity anteriorly and posteriorly along with progressive left-sided nasal obstruction for the last year.

Anterior rhinoscopy and endoscopic evaluation revealed a large, fleshy, polypoidal mass completely filling the left nasal cavity and slightly protruding from the left anterior nares. The mass was friable and bled easily upon touch.

Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) of the nose paranasal sinuses demonstrated a soft tissue mass completely opacifying the left nasal cavity and ethmoid sinuses, causing local remodeling of the nasal septum and medial maxillary sinus wall without obvious intracranial or intraorbital extension.

The patient underwent a complete endoscopic excision of the left nasal mass under G/A. Histopathological examination and immunohistochemical (IHC) profiling confirmed a diagnosis of Ewing's Sarcoma of the sinonasal tract (tumor cells positive for Cyclin D-1 and NKX2.2 and exhibit focal positivity for FL1-1. They were negative for cytokeratin AE1/AE3, Neurofilament, NSE, MYO-D1, Desmin, LCA, CD3, CD20 and TdT with proliferative index about 30 – 40%. Microscopic examination revealed diffuse, dense infiltrate of small malignant cells with minimal cytoplasm, arranged in sheets / tuberculae.

Following surgical excision, the patient was immediately referred to the oncology department

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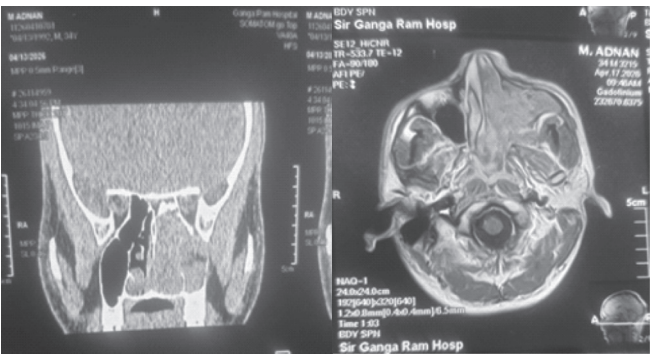


for adjuvant therapy. She successfully completed her prescribed systemic chemotherapy regimen and is scheduled for radiotherapy. On her ENT follow-up, she is symptom free as reports no episode of epistaxis and nasal obstruction after surgical excision and nasal endoscopy is unremarkable.

Case Report 2:

A 33-year-old male presented to the ENT Outpatient Department (OPD) of Sir Ganga Ram Hospital Lahore in March 2026. His primary complaint was persistent, progressive left-sided nasal obstruction for the last 1.5 years, no history of epistaxis.His medical history was significant for a nasal surgery performed 2 years prior at a private hospital in other city. The historical biopsy report from that procedure suggested a diagnosis of olfactory neuroblastoma. However, the patient did not seek the recommended oncological referral or follow-up at that time.

Clinical examination revealed an extensive polypoidal mass completely filling the left nasal passage. Urgent CT and MRI mapping showed a destructive soft tissue lesion of the left sinonasal tract causing erosion of pterygoid plates with intracranial extradural extension. The patient was discussed with neurosurgical department and they advised no neurosurgical intervention required.



Comparative Summary of Cases

Clinical Feature	Case 1	Case 2
Age / Gender	30 Years / Female	33 Years / Male
Presenting symptom	Profuse, recurrent epistaxis (6 months)	Nasal obstruction (1.5 years)
Date & mode of presentation.	September 2025 (Emergency)	March 2026 (OPD)
Previous Surgical History	None	Yes
Initial diagnosis	None	Olfactory Neuroblastoma
Definitive Diagnosis	Sinonasal Ewing's Sarcoma	Sinonasal Ewing's Sarcoma
Current Status	Completed chemotherapy ,scheduled for radiotherapy, symptom free	Referred to Oncology post-excision

Patient underwent endoscopic excision biopsy of the left sinonasal mass and there was no complication per and post operatively in terms of bleeding, CSF leak.

Strikingly, the definitive histopathology classified the lesion as Ewing's Sarcoma rather than olfactory neuroblastoma as CD99, NKX2.2 positive in tumor cells, Synaptophysin weak focal positive in tumor cells and on microscopy sections revealed a malignant neoplasm showing sheets of round blue cells with fine chromatin, scanty cytoplasm and small nucleoli.

Postoperatively, the patient and his family were thoroughly counselled regarding the critical prognosis of the disease, the correction of the previous diagnosis, and the absolute necessity of aggressive multimodal therapy. He was referred to medical and radiation oncology for immediate treatment where he is scheduled for chemotherapy.

DISCUSSION

These two cases highlight the diverse clinical behavior of extraskeletal Ewing's sarcoma arising in the sinonasal cavity. Sinonasal Ewing's sarcoma requires a high index of suspicion. It frequently mimics benign polyps clinically or other small round cell malignancies (like olfactory neuroblastoma) histologically, undifferentiated carcinoma, sino-nasal melanoma, lymphoma, mesenchymal chondrosarcoma, small-cell neuroendocrine carcinoma all challenging due to histopathological morphology overlapping in all these cases.⁵

As it is observed in both cases, surgical excision endoscopically is the first step of treatment. The prognosis relies majorly on aggressive adjuvant

chemotherapy and radiotherapy. A major challenge in managing these cases in our clinical setups remain patient non-compliance and loss to follow-up, emphasizing the importance of patient's counselling.

CONCLUSION

Standard treatment requires an aggressive, multi-modality approach, surgery with adjuvant chemo radiation remains the primary treatment option.⁶ Quality of life can be preserved and results can be greatly enhanced by early diagnosis, customized treatment and follow up.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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AUTHORSHIP AND CONTRIBUTION DECLARATION	
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2	Maria Asif: Critical revisions.
3	Mehak Ahmad: Study concept, design.
4	Ayesha Fayyaz: Literature review.
5	Aamna Durrani: Review of manuscript.
6	Syed Muhammad Waqas: Proof reading.