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From Obstruction to Oncogenesis: Ewing's Sarcoma Hidden in the Nasal Cavity

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ABSTRACT

BACKGROUND: Sinonasal tumors in children are uncommon, and the majority are benign. Malignant lesions such as Ewing's sarcoma are exceedingly rare in this location, accounting for only a small fraction of cases in the head and neck. Because symptoms such as nasal obstruction and discharge can mimic those of inflammatory polyps, diagnosis is often delayed.
CASE PRESENTATION: A 16-year-old boy presented with a six-month history of progressive left-sided nasal obstruction, followed by blood-stained mucoid discharge, anosmia, and change in voice. Examination revealed a firm, polypoidal mass filling the left nasal cavity. CT and MRI suggested an angiomatous polyp. The mass was excised endoscopically. Histopathology showed sheets of malignant small round cells with peritheliomatous arrangement, and immunohistochemistry demonstrated diffuse membranous CD99 positivity with nuclear FLI-1 expression, confirming Ewing's sarcoma. Staging with chest CT, PET scan, and bone marrow biopsy excluded systemic spread, apart from reactive cervical nodes. The patient was treated with multi-agent chemotherapy as per the AEWS1221 protocol, along with local radiotherapy. At one year of follow-up, he remains well and disease-free.
CONCLUSION: This case underlines the importance of considering malignant pathology in unilateral nasal polyps in adolescents. Early biopsy is essential for diagnosis, as imaging alone may mimic benign conditions. Combined surgical and oncological management can provide favourable outcomes in sinonasal Ewing's sarcoma.

Keywords: Ewing sarcoma; malignancy; nasal polyp; Adolescent;

INTRODUCTION

Sinonasal tumours in children and adolescents are uncommon and frequently present a diagnostic challenge. Most nasal masses in this age group are benign, such as antrochoanal polyps, juvenile nasopharyngeal angiofibromas, or congenital midline anomalies, but malignant lesions must always be considered when symptoms are unilateral, progressive, or accompanied by bleeding(1). Among malignant small round cell tumours, Ewing's sarcoma and the related primitive neuroectodermal tumours (PNETs) are exceedingly rare in the sinonasal tract, accounting for only 1–4% of all Ewing's

sarcoma cases in the head and neck region(2,3).

Ewing's sarcoma, first described by James Ewing in 1921, is characterised by undifferentiated small round blue cells, strong membranous expression of CD99, nuclear positivity for FLI-1, and often the EWSR1-FLI1 translocation(2). While the skeletal form most commonly involves the long bones and pelvis, extraskeletal variants may arise from soft tissue, including the nasal cavity and paranasal sinuses(4,5).

The clinical presentation in sinonasal sites is typically nonspecific and overlaps with benign entities; common symptoms include nasal obstruction, blood-stained rhinorrhea, epistaxis, anosmia, and facial swelling(1,6). This frequently contributes to delayed recognition and advanced disease at diagnosis. Imaging helps assess the extent but often fails to reliably distinguish benign polyps from malignant tumors. Histopathology and immunohistochemistry remain essential for definitive diagnosis.

We report a rare case of primary Ewing's sarcoma of the nasal cavity in an adolescent male, radiologically suspected as an angiomatous polyp but confirmed on histopathology and immunohistochemistry. The case highlights the importance of early biopsy in unilateral nasal polyposis and reviews the challenges in recognising and managing this rare entity.

CASE REPORT

A 16-year-old male, previously healthy, presented to the otolaryngology outpatient department with complaints of persistent left-sided nasal obstruction of six months' duration. The obstruction was insidious in onset and gradually progressive. Initially, the patient had trouble breathing predominantly through the left nostril, which became more pronounced in the supine position and during expiration. The obstruction worsened in cold weather and improved transiently with medications, including antihistamines and oral antibiotics prescribed at a local facility. Three months before presentation, he also developed nasal discharge from the same side. The discharge was mucoid in consistency, occasionally blood-stained, and non-foul-smelling. These symptoms were accompanied by progressive loss of smell and a noticeable change in voice quality. There was no history of recurrent running nose, itching, sneezing, watering of eyes, or symptoms suggestive of allergic rhinitis. He denied headache, recurrent epistaxis, fever, sore throat, throat clearing, ear pain, or ear discharge.

The patient had never undergone a formal ENT evaluation before this presentation and had only received symptomatic management. There was no history of similar complaints in the past, no history of chronic systemic illness such as asthma or tuberculosis, and no prior surgical procedures. Drug allergies were not reported. His family history was unremarkable.

Upon general examination, the patient was moderately built and well-nourished, oriented to time, place, and person, with stable vital signs. Pulse rate was 80 beats per minute, respiratory rate 25 per minute, and blood pressure 108/64 mmHg. Capillary refill time was less than three seconds. Pallor was present, while icterus, clubbing, pedal oedema, and lymphadenopathy were absent.

On local examination, the external nose was normal (Figure 1). Loss of misting was noted over the left nostril, and the cotton wisp test demonstrated absent airflow on the left side. The cotton swab test was negative bilaterally. Anterior rhinoscopy revealed congested nasal mucosa, a normal inferior turbinate, and deviation of the nasal septum to the right. A polypoidal, irregular, firm mass was visualised occupying and occluding the left nasal cavity, which bled on touch. Blood-stained mucoid discharge was present. Posterior rhinoscopy further confirmed a polypoidal lesion extending into the posterior nares on the left side. Paranasal sinus examination was unremarkable. Oral cavity and ear examinations were within normal limits.

Figure 1. Clinical photograph of the 16-year-old male patient at presentation, showing no external nasal deformity. The image has been anonymized to protect patient identity.

A diagnostic nasal endoscopy was performed, which showed a firm, fixed, irregular polypoidal mass obstructing the left nasal cavity. The scope could not be negotiated beyond the lesion. The right nasal cavity was narrowed due to the deviated septum, though the choana was free. In view of the progressive nature of the symptoms, the firm consistency of the mass, and the presence of bloody discharge, a malignant aetiology was considered.

A contrast-enhanced computed tomography (CT) scan of the paranasal sinuses revealed a soft tissue density lesion completely filling the left nasal cavity. The mass obscured the margins of the turbinates, making them difficult to delineate separately. The nasal septum was noted to be deviated to the right. There was mucosal thickening involving bilateral maxillary, ethmoid, and frontal sinuses, suggestive of chronic sinusitis, along with left-sided polyposis (Figure 2).

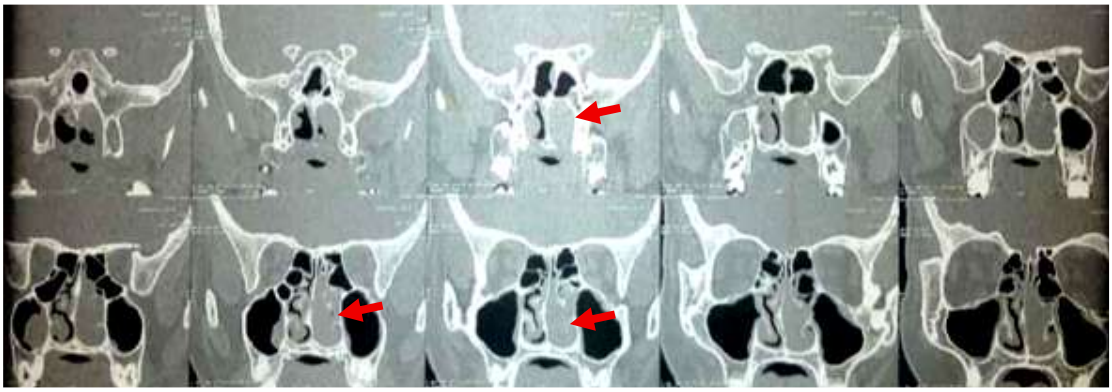

Figure 2. Coronal contrast-enhanced CT scan of the paranasal sinuses showing a soft tissue density lesion completely occupying the left nasal cavity (red arrows). The lesion obscures the margins of the turbinates, causes deviation of the nasal septum to the right, and is associated with mucosal thickening of the bilateral maxillary, ethmoid, and frontal sinuses, consistent with sinusitis.

Subsequent magnetic resonance imaging (MRI) of the nose and paranasal sinuses showed a well-defined lesion measuring 2.5 × 3.7 × 4.3 cm in the left nasal cavity. The mass was hyperintense on T2-weighted images and demonstrated avid contrast enhancement. There was no extension into the choana, nasopharynx, or surrounding

structures. These findings were initially interpreted as being most consistent with an angiomatous nasal polyp.

Given the obstructive symptoms, endoscopic excision of the mass was undertaken under general anaesthesia. Using a microdebrider, the lesion was excised in toto with clearance of at least 1 cm from the visible margins. The immediate postoperative period was uneventful, and the patient recovered without complications (Figure 3).

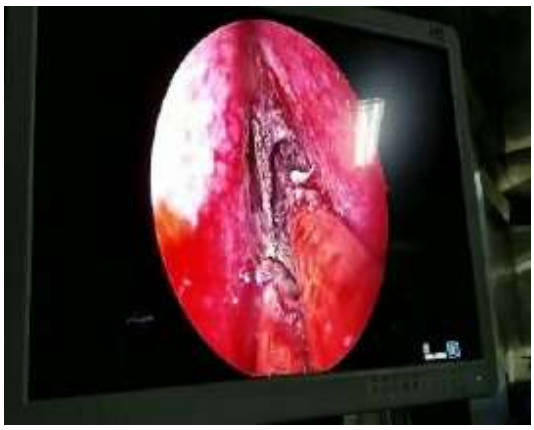

Figure 3. Intraoperative endoscopic view demonstrating excision of the left nasal cavity mass using a microdebrider. The lesion appeared firm and polypoidal, with associated mucosal congestion and contact bleeding.

Histopathological examination of the excised tissue revealed respiratory epithelium focally replaced by stratified squamous epithelium with an underlying stroma infiltrated by sheets of small, round to oval malignant cells. These were arranged in a peritheliomatous pattern and displayed marked anaplastic features. The morphology was suggestive of a small round cell tumour.

To further classify the tumour, immunohistochemistry (IHC) was performed. Tumour cells showed diffuse and strong membranous positivity for CD99 and nuclear positivity for FLI-1, which established the diagnosis of Ewing's sarcoma/primitive neuroectodermal tumour (PNET) of the nasal cavity.

A metastatic workup was initiated, including a chest CT scan, bilateral bone marrow aspirate and biopsy, and a positron emission tomography (PET) scan. All investigations were negative for metastatic spread, except for the presence of mildly PET-avid cervical lymph nodes, which were not enlarged and were considered reactive rather than metastatic.

The case was discussed in a multidisciplinary tumour board, and the consensus was to proceed with multimodality therapy. The patient received systemic multi-agent chemotherapy as per the AEWS1221 protocol (NCT02306161), combined with concurrent local radiotherapy to the primary site.

The patient tolerated the treatment well, with no major adverse events reported during therapy. Regular follow-up, including clinical examination and interval imaging, was performed. At one-year follow-up, the patient remains disease-free, with no evidence of local recurrence or distant metastasis.

DISCUSSION

Sinonasal Ewing's sarcoma is a rare malignancy, with only sporadic case reports and small series available in the literature(7,8). It occurs predominantly in adolescents and young adults, with a slight male predominance. Clinical manifestations are often misleading, and in several reported cases—including ours—the lesion was initially misdiagnosed as an angiomatous polyp or juvenile nasopharyngeal angiofibroma. This highlights the importance of maintaining a high level of suspicion when evaluating unilateral or vascular nasal polyps in the pediatric age group(5,6).

Histopathology typically demonstrates sheets of uniform, small, round, blue cells, and immunohistochemistry is critical for confirmation. CD99 positivity is highly characteristic, while FLI-1 nuclear expression further supports the diagnosis(9). Additional cytogenetic confirmation of the EWSR1 translocation using fluorescence in situ hybridization (FISH) or RT-PCR may be employed where available(2). The main histological differentials include

rhabdomyosarcoma, lymphoma, olfactory neuroblastoma, and sinonasal undifferentiated carcinoma(1).

Management requires a multidisciplinary approach that combines surgery, chemotherapy, and radiotherapy. The feasibility of surgical resection is often dictated by tumour proximity to critical structures such as the orbit and skull base. When complete excision with negative margins is achievable, outcomes are generally superior to non-surgical approaches(6). In our patient, endoscopic resection was possible, followed by systemic chemotherapy according to the AEWS1221 protocol and adjuvant radiotherapy, which aligns with contemporary practice. Chemotherapy remains the backbone of treatment, with vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide forming the standard regimen(2,3).

Prognosis in sinonasal Ewing's sarcoma, though historically poor, has improved with multimodality therapy. Reported outcomes suggest that localised disease in the sinonasal tract may fare better than axial skeletal primaries, partly due to earlier detection and improved access for endoscopic surgery(3). Adverse prognostic factors include age >18 years, tumour size >8 cm, metastasis at diagnosis, and local control with radiotherapy alone. In our patient, disease-free survival at one year reflects the value of timely diagnosis and aggressive multidisciplinary management(6).

In conclusion, sinonasal Ewing's sarcoma is a rare but important differential diagnosis in adolescents with unilateral nasal obstruction and blood-stained discharge. Early biopsy of atypical nasal polyps is essential, and multimodality treatment offers the best chance for long-term survival.

CONCLUSION

Sinonasal Ewing's sarcoma is an uncommon entity that often mimics benign conditions such as nasal polyps, leading to delays in diagnosis. Persistent, unilateral, or hemorrhagic nasal masses in adolescents should always raise suspicion for underlying malignancy. Histopathology with immunohistochemistry remains the cornerstone for diagnosis, with CD99 and FLI-1 serving as key markers. Optimal outcomes are achieved through a multidisciplinary approach that combines surgical excision, systemic chemotherapy, and radiotherapy. Early recognition and timely treatment can significantly improve prognosis, as demonstrated in this case where the patient remains disease-free at one year of follow-up.

Declaration by Authors

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