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# A rare case of primary Ewing's sarcoma presenting in the posterior nasal cavity with extension into the sphenoid sinus and a review of the literature

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ABSTRACT

Ewing's sarcomas/primitive neuroectodermal tumors (PNET) are a group of tumors characterized by small round cells that rarely affect the bone and soft tissues of the nasal cavities. Herein, we describe the case of a 38-year-old man who presented with a one-year history of epistaxis, headache, hyposmia, anosmia, night sweats, weight loss, hearing loss, dysphagia, and nasal blockage. A computed tomography (CT) scan and magnetic resonance imaging (MRI) studies revealed a lesion in the posterior nasal cavities with extension into the sphenoid and ethmoid sinuses. Histological examination of a biopsied specimen obtained via flexible fiberoptic endoscopy showed small dark round cells with scanty cytoplasm arranged in solid nests and pseudorosette formations. The diagnosis of Ewing's sarcoma was confirmed by the positive reactive pattern of the neoplastic cells for CD99 by immunohistochemistry studies and also identification of a chromosomal translocation of *FLI1* in PCR studies. Cervical lymph nodes and bone marrow involvement were also observed. The patient was treated by adjuvant chemotherapy including vincristine, endoxan, and adriamycin for 6 cycles followed by radiotherapy (eleven courses). Unfortunately, before completion of treatment, the patient died due to disease progression and brain metastasis.

## Introduction

Ewing's sarcomas/primitive neuroectodermal tumors (PNET) are rare tumors characterized by small round cells that mainly affect bone and soft tissues in children and young adults [1,2]. The most common bones affected by the neoplastic cells are the pelvis, femur, humerus, ribs, and clavicle bones [2,3]. However, bone involvement in patients over 30 years old is rare, and involvement of the nasal cavity by Ewing's sarcomas/PNET tumors is extremely rare and only a few case reports have been published [4–6]. Herein, we present a case of Ewing's sarcoma arising in the nasal cavity with extension into the ethmoid and sphenoid sinuses with secondary bone marrow involvement.

## Case presentation

A 38-year-old man came to our otorhinolaryngology clinic with a one-year history of epistaxis, headache, hyposmia, anosmia, night sweats, weight loss, hearing loss, dysphagia, and nasal blockage. On physical examination, numbness of both cheeks was evident. Laboratory tests revealed anemia with red blood cell count $3.03 \times 10^6$/ml (normal range, 3.6–6.1), hemoglobin level of 7.5 mg/dl (normal range 11.5–18.8 g/dl), hematocrit, 26.5% (normal range 34–54%), and platelet count 143 × 103/ml (normal range, 165–415).

An axial view of a neck computed tomography (CT) scan with intravenous contrast showed a soft tissue mass with punctuate calcification in the ethmoid and sphenoid sinuses with extension into the nasal cavities. On the sagittal view, involvement and destruction of the clivus was evident, and on the coronal view, erosion of the medial wall of the left orbit was seen (Fig. 1A, B). Mucosal thickening and retention cysts in both maxillary sinuses with left osteomeatal complex obstruction (OMC) was present. Multiple hypodense lymph nodes (necrotic) were seen on both sides in the anterior and posterior cervical lymphatic chains, the largest one being around 23 × 25 mm in zone 2 on the left side. A cranial MRI, without contrast, using axial, sagittal, and coronal views on T1-weighted images (T1WI), showed a heterogeneous mildly hyper signal nasopharyngeal mass measuring about 51 × 56 × 30 mm that was occupying both sides of the ethmoid and sphenoid sinuses and superior part of the nasal cavities. Erosion of the medial wall of the left orbit and clivus was evident (Fig. 2A, B, C). Obstruction of bilateral Eustachian tubes caused mastoiditis and bilateral OMC caused retention

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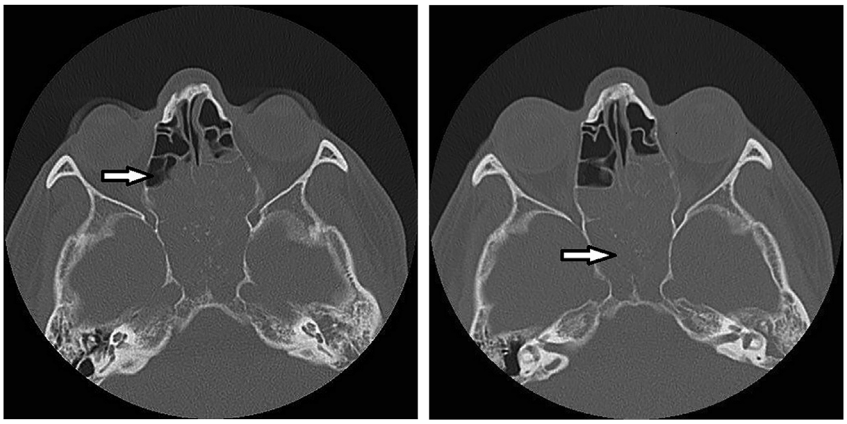

**Fig. 1.** A: Sagittal view CT scan demonstrates destruction of the clivus. B: The coronal view shows erosion of the medial wall of the left orbit.

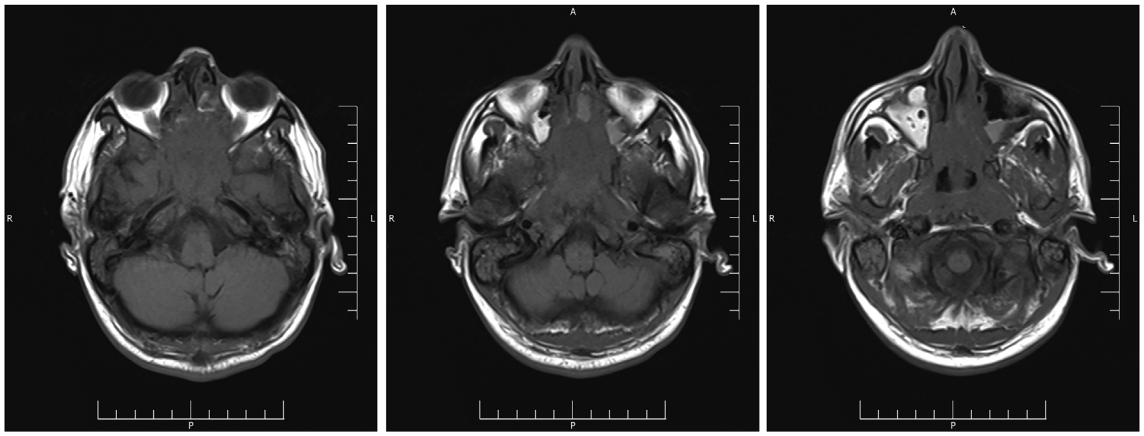

**Fig. 2.** A, B, C: T1-weighted image (T1WI), shows a heterogeneous mildly hyper signal nasopharyngeal mass that is occupying both sides of the ethmoid and sphenoid sinuses and superior part of the nasal cavities. Erosion of the medial wall of the left orbit and clivus is evident.

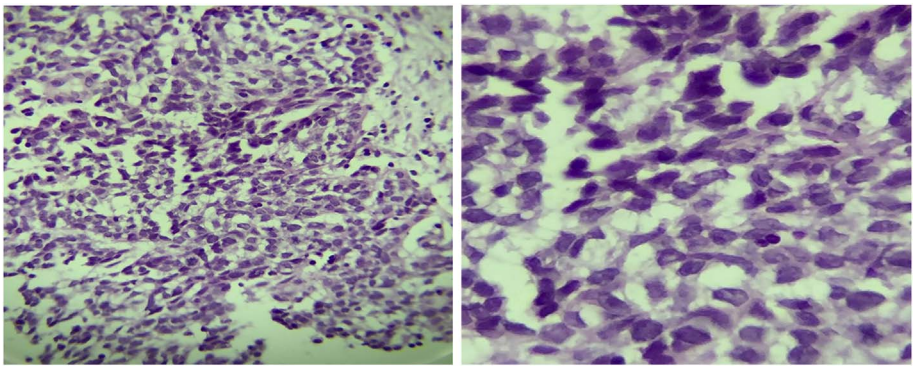

**Fig. 3.** A, B: Small dark round cells with scanty cytoplasm that are arranged in solid nests and pseudo rosette formations within fibrovascular connective tissue, (H&E staining, magnification ×10, ×20).

of secretions in both maxillary sinuses. Multiple hypo signals on T1W1 and hyper signal lesions on T2W1 were seen in the upper zone of the neck bilaterally, suggestive of necrotic lymph nodes. The patient underwent flexible fiberoptic endoscopy, but because of the widespread extension of the mass, only biopsy without complete resection was performed. Histologically, the tumor consisted of small dark round cells with scanty cytoplasm that were arranged in solid nests and pseudo rosette formations with vascular connective tissue between the neoplastic nests (Fig. 3A, B). Immunohistochemical (IHC) studies revealed diffuse positivity for CD99, and Vimentin, and patchy positivity for S-100 protein (Fig. 4A, B, C). The neoplastic cells were negative for pan-cytokeratin, NSE, leukocyte common antigen, synaptophysin, and chromogranin A. On periodic Acid-Schiff (PAS) staining, the neoplastic cells had positive cytoplasmic staining results that confirmed the glycogen content of the cytoplasm. In molecular studies (PCR) chromosomal translocation of *FLI1* was identified. Thus, a diagnosis of primary Ewing's sarcoma/PNET of the nasal cavity was confirmed. For staging of the tumor, bone marrow aspiration and a biopsy were performed. Histopathological and cytological examinations revealed marrow involvement by neoplastic cells (Fig. 5). Cerebrospinal fluid cytology was negative for neoplastic cells.

## Treatment and follow-up

Because the tumor had an intracranial extension with bone marrow metastases, surgical resection was not feasible. Adjuvant chemotherapy followed by radiotherapy was selected. A chemotherapy protocol composed of vincristine (2 mg, D1), endoxan (1500 g, D1), and adriamycin (50 g, D2) was administered for 6 cycles. After chemotherapy, the patient was scheduled to undergo radiotherapy (54 Gy) for eleven