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Unilateral Decreased Visual Acuity as the Initial Manifestation of Ewing Sarcoma : the Key Role of Imaging

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ABSTRACT

Ewing sarcoma is a malignant bone tumor predominantly affecting children and adolescents, most commonly involving long bones and the pelvis. Cranial involvement is rare and may present with misleading clinical signs. We report the case of a 17-year-old patient who presented with progressive decreased visual acuity, followed by unilateral exophthalmia and headache, without prior oncological history. Computed tomography (CT) revealed an aggressive osteolytic lesion centered on the right greater wing of the sphenoid, responsible for cortical destruction, with extension into the surrounding soft tissues and orbit. Histopathology confirmed the diagnosis of Ewing sarcoma. This case highlights the critical role of CT imaging in detecting atypical cranial presentations of Ewing sarcoma and guiding timely diagnosis, biopsy, and multidisciplinary management.

KEYWORDS

Ewing sarcoma, Visual Acuity, Unilateral exophthalmia, Cranial vault, Sphenoid bone, Computed tomography, Case report

MAIN ARTICLE

INTRODUCTION

Ewing sarcoma is an aggressive primary malignant tumor of bone and soft tissue, predominantly affecting adolescents and young adults. It is characterized by rapid growth and early metastatic potential. Cranial involvement is uncommon, accounting for less than 1% of cases, and often presents with nonspecific neurological or ophthalmological symptoms [1,2].

Decreased visual acuity is a rare presenting sign of Ewing sarcoma and may initially suggest inflammatory, infectious, or primary orbital pathology. Radiological imaging, particularly computed tomography (CT), plays a pivotal role in identifying aggressive bone lesions and narrowing the differential diagnosis. We present a case in which decreased visual acuity was the revealing sign of cranial Ewing sarcoma in a previously healthy adolescent.

CASE PRESENTATION

A 17-year-old patient with no significant past medical history presented with progressive decreased visual acuity and unilateral exophthalmia associated with persistent headaches. There were no systemic symptoms, no history of trauma, and no previously known malignancy. Clinical examination confirmed non-pulsatile unilateral exophthalmia without signs of infection.

Given the atypical presentation and progressive symptoms, a non-contrast and contrast-enhanced CT scan of the head was performed.

Radiological Findings

CT imaging demonstrated an aggressive osteolytic lesion centered on the greater wing of the right sphenoid bone, with poorly defined margins and cortical destruction (Figure 1). The lesion was associated with a soft tissue mass extending into the orbit, exerting a mass effect on the lateral rectus muscle, explaining the unilateral exophthalmia (Figure 2).

The mass showed heterogeneous density with moderate enhancement after contrast administration (Figure 2). Intracranial extension was suspected, with compression of adjacent

structures but no evidence of acute hemorrhage. The aggressive bone destruction and associated soft tissue component strongly suggested a malignant etiology.

Based on imaging features, differential diagnoses included Ewing sarcoma, metastatic neuroblastoma, rhabdomyosarcoma, and Langerhans cell histiocytosis (Table 1). The radiological appearance, patient age, and cranial vault involvement favored the diagnosis of Ewing sarcoma.

Diagnostic Confirmation and Management

Following imaging, a biopsy of the lesion was performed. Histopathological examination revealed sheets of small round blue cells. Immunohistochemistry showed strong membranous CD99 positivity, and molecular analysis confirmed the presence of an EWSR1 gene rearrangement, establishing the diagnosis of Ewing sarcoma.

The patient was referred to a multidisciplinary oncology team for further staging and initiation of systemic chemotherapy.

DISCUSSION

Cranial involvement in Ewing sarcoma is rare and represents a diagnostic challenge, particularly when it is the initial manifestation of the disease. In the present case, unilateral decreased visual acuity associated with headache was the revealing symptom in a previously healthy adolescent, emphasizing the importance of radiological imaging in detecting occult malignant bone lesions [2,3,4].

Radiological Importance of Exophthalmia in Adolescents

Unilateral decreased visual acuity and exophthalmia in pediatric and adolescent patients are an alarming clinical sign that requires urgent imaging evaluation. From a radiological standpoint, the primary objective is to determine whether the origin is intraorbital, extraorbital, or secondary to adjacent bone pathology. CT is often the first-line modality due to its availability, speed, and excellent depiction of bone anatomy [1,4].

In this case, CT imaging immediately demonstrated an aggressive cranial vault lesion with orbital extension, shifting the diagnostic focus from primary orbital disease to malignant bone

pathology. This underscores the role of CT as a decisive diagnostic tool in emergency and ophthalmological presentations.

CT Imaging Patterns Suggestive of Ewing Sarcoma

Ewing sarcoma typically appears on CT as a poorly marginated osteolytic lesion with cortical destruction and an associated soft tissue mass. Although periosteal reaction is classically described in long bones, cranial lesions often lack a well-organized periosteal response due to the anatomical characteristics of flat bones [3,4].

The presence of:

- Aggressive bone destruction
- A large soft tissue component
- Orbital invasion causing mass effect

Is highly suggestive of a malignant process. In adolescents, these features should prompt consideration of Ewing sarcoma even in the absence of known primary disease.

Role of MRI and Functional Imaging

Although CT was the revealing examination in this case, MRI plays a complementary role in the complete assessment of cranial Ewing sarcoma. MRI is superior for evaluating intracranial extension, dural involvement, cranial nerve compression, and brain parenchymal invasion. Typical MRI findings include low-to-intermediate T1 signal, heterogeneous T2 hyperintensity, diffusion restriction, and intense post-contrast enhancement [5,6].

Functional imaging, particularly FDG PET-CT, is essential for staging and detection of distant metastases. High FDG uptake reflects the aggressive metabolic activity of Ewing sarcoma and helps differentiate it from inflammatory or benign lesions.

Radiological Differential Diagnosis

The radiological appearance of aggressive cranial bone lesions with orbital extension in adolescents is not specific to Ewing sarcoma. Several entities must be considered, making differential diagnosis a critical component of radiological interpretation (Table 1).

Diagnostic Pitfalls and Radiologist's Role

A major pitfall in such cases is misinterpreting the lesion as a primary orbital tumor or inflammatory process, especially when clinical signs are limited to exophthalmia and headache. Radiologists must maintain a high index of suspicion when encountering aggressive bone destruction adjacent to the orbit.

Prompt identification of malignant imaging features allows:

- Early biopsy guidance
- Accurate staging
- Rapid referral to oncology services

Thus, radiology serves as the gateway to diagnosis in atypical presentations of Ewing sarcoma.

CONCLUSION

This case illustrates the critical role of CT imaging in revealing an uncommon presentation of cranial Ewing sarcoma. Decreased visual acuity and unilateral exophthalmia in adolescents should prompt thorough radiological evaluation, as early detection of aggressive bone lesions significantly impacts diagnostic orientation and patient management. Radiologists play a central role in recognizing these rare presentations and initiating timely multidisciplinary care. Awareness of the characteristic imaging features allows differentiation from inflammatory or benign orbital conditions and prevents diagnostic delay. Furthermore, prompt radiological identification facilitates appropriate staging, guides biopsy planning, and contributes to optimized therapeutic decision-making, ultimately improving clinical outcomes in this aggressive malignancy.

FIGURES:

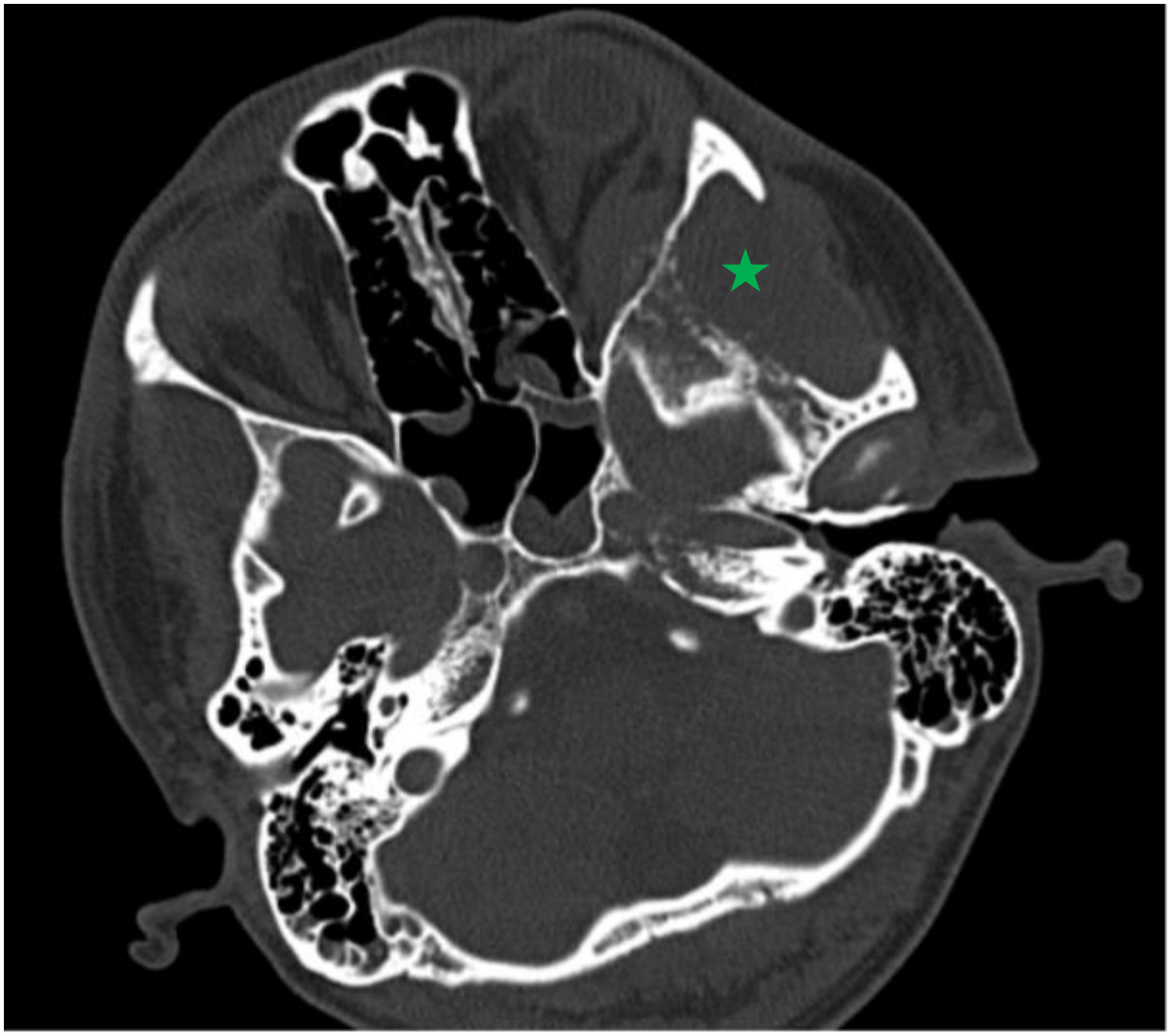

Figure 1: Axial CT scan (bone window) showing an aggressive osteolytic lesion (green star) of the greater wing of the right spenoid bone with orbital extension.

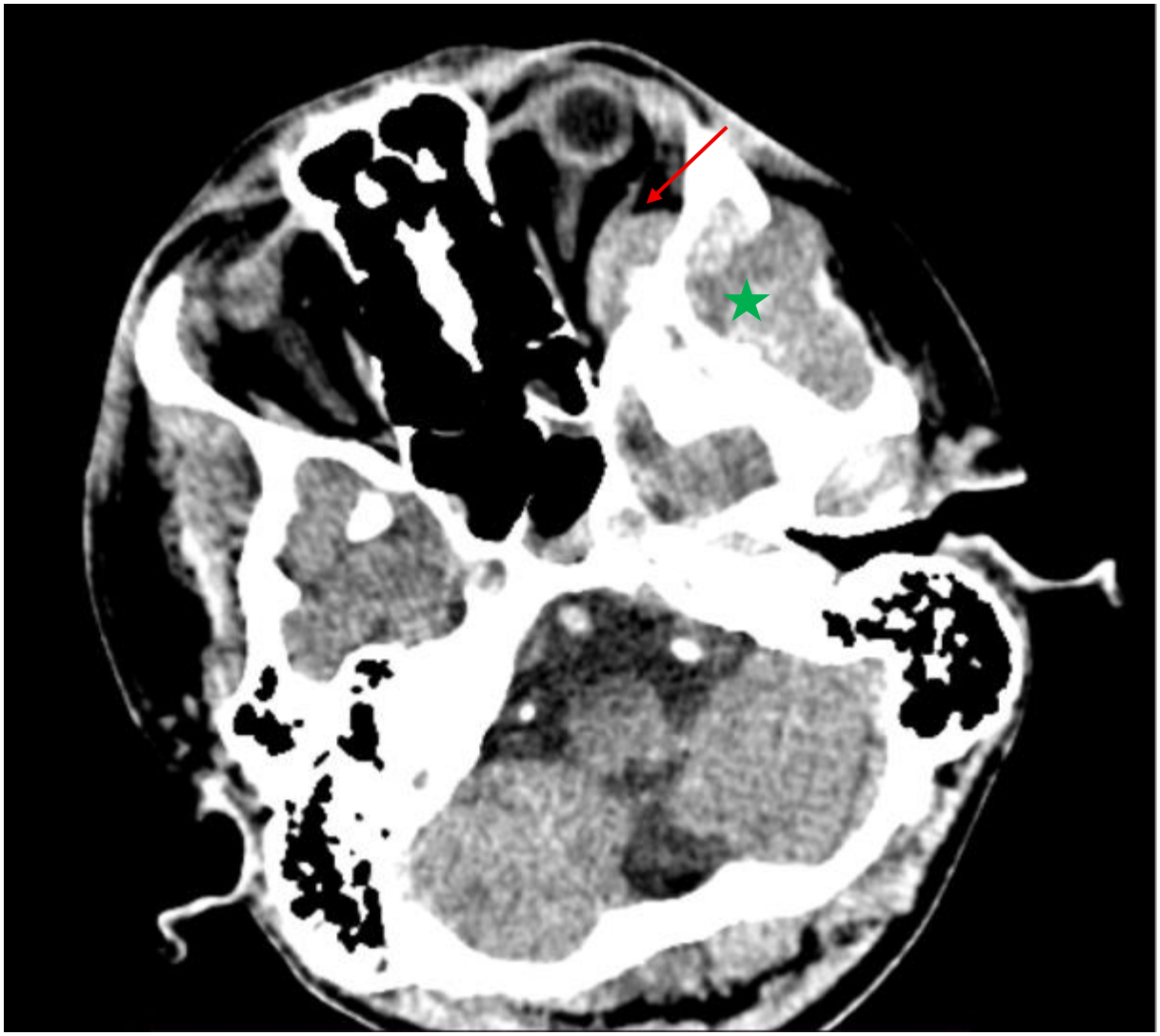

Figure 2 : Axial CT scan (soft tissue window) showing the lesion (green star) causing mass effect on the lateral rectus muscle (red arrow), displacing the globe anteriorly.

TABLES :

Table 1 : Radiological Differential Diagnosis of Aggressive Cranial Lesions in Adolescents

Diagnosis	*Typical Age*	*CT Bone Findings*	*Soft Tissue Component*	*Key Imaging Clues*
Ewing Sarcoma	*Adolescents*	*Aggressive osteolytic lesion, cortical destruction*	*Large, heterogeneous mass*	*Age, aggressive bone loss, CD99 positivity*
Metastatic Neuroblastoma	*<10 years*	*Lytic skull lesions, sutural widening*	*Orbital mass common*	*Younger age, calcifications, known primary*
Rhabdomyosarcoma	*Children*	*Bone erosion rather than destruction*	*Dominant soft tissue mass*	*Primary orbital origin*
Langerhans Cell Histiocytosis	*Children*	*Punched-out lytic lesions*	*Minimal soft tissue*	*Beveled edges, less aggressive*
Osteomyelitis	*Any age*	*Irregular lytic areas*	*Possible abscess*	*Clinical infection signs*
Osteosarcoma	*Adolescents*	*Mixed lytic-sclerotic lesion*	*Variable*	*Osteoid matrix, sunburst reaction*

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