

## CLINICAL CASE

## Extraosseous Ewing sarcoma

Juan Salvador Uribe Olalde,<sup>1</sup> Héctor Guillermo Palomera Gómez,<sup>1</sup> Javier Quillo Olvera,<sup>1</sup> L. Alberto Alcántara Gómez,<sup>1</sup> Luis Eduardo Zambrano Velarde,<sup>1</sup> Ernesto Gómez Limón,<sup>1</sup> Elisa Alexandra González Plascencia,<sup>2</sup> Jorge Paz Gutiérrez,<sup>1</sup> Juan de la Cruz Ramírez Jaimez,<sup>1</sup> Héctor Velázquez Santana<sup>1</sup>.

## ABSTRACT

**Background.** The Ewing sarcoma family includes Ewing bone sarcoma, primitive neuroectodermal tumor, extraosseous Ewing's sarcoma, and Askin tumor (Ewing sarcoma of the chest wall). The most common sites of these tumors are chest and limbs. These malignancies arise from bone or soft tissues. Histologically, tumors are characterized by high cellularity and are small, round, blue and with a tendency to form rosette structures.

**Case report.** We report the case of a 14-year-old patient whose evolution began 3 months before admission with left frontoparietal headache radiating to the orbit. During neurological examination, proptosis and decreased visual acuity were found as well as papillary atrophy. Cranial computed tomography identified a left intraorbital injury and another ipsilateral temporoparietal. Tumor exeresis was performed, reporting positive for primitive neuroectodermal tumor markers, vimentin and CD99.

**Conclusions.** Ewing sarcoma is extremely rare at the orbital level; however, it is prudent to consider this tumor as a differential diagnosis in lesions of this region. Prognosis depends on early diagnostic accuracy.

**Key words:** orbital tumor, primitive neuroectodermal tumor, primary Ewing sarcoma of skull.

## INTRODUCTION

The family of Ewing sarcomas (ES) includes Ewing bone sarcoma, primitive neuroectodermal tumors, extraosseous ES and Askin tumor (Ewing sarcoma of the chest wall).<sup>1,2</sup> In the analysis of 1,631 cases carried out by Jawad et al., there were 290 cases reported of the extraosseous location (18.4%).<sup>3</sup> These types of tumors present themselves mainly during the second and third decades of life, and the most common sites of presentation are the chest cavity and the extremities.<sup>4</sup> ES is a malignant tumor that may arise from the bone and soft tissue.<sup>5</sup> It originates from the postganglionic parasympathetic primordial cells derived from the neural crest.<sup>6</sup> The family of ES is an aggressive form of cancer.<sup>7</sup> Extraosseous ES is defined

as a soft tissue tumor without bone involvement, but with a similar histology<sup>8</sup> because it is characterized by round, blue, small cells derived from the neural crest, giving it its neuroectodermal character. Its coloring is determined by intense basophilia, which is observed with hematoxylin-eosin staining with groupings in chords or Homer-Wright rosette structures). To carry out the definitive diagnosis, immunohistochemical studies positive for vimentin and CD99 are necessary.<sup>9</sup> It is one of the most differentiated histological types of malignant tumors. It appears primarily in bony structures; therefore, its probability in extraosseous structures is very low. Treatment for this type of disease is multidisciplinary and includes surgical management with wide resection of the lesion, chemotherapy and radiation therapy.<sup>10,11</sup>

Because of this, it is considered that the prognosis of these lesions is intimately related to the early diagnosis. Much will depend on the stage at which the lesion is found.<sup>12</sup> In Mexico there are no statistical or anecdotal data from this type of extraosseous tumor. This paper presents the case of a patient with is extraosseous ES that, globally, is very rare. A review of the literature on the subject was also carried out.

<sup>1</sup> Hospital Regional Dr. Valentín Gómez Farías  
Instituto de Seguridad y Servicios Sociales  
de los Trabajadores del Estado, Zapopan, Jalisco, México  
<sup>2</sup> Unidad de Medicina Familiar No. 51  
Instituto Mexicano del Seguro Social,  
Guadalajara, Jalisco, México

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## CLINICAL CASE

We present the case of a 14-year-old male. Three months prior to his admission he presented with sharp, intermittent left frontoparietal headache that occurred predominantly in the evenings. The headache radiated to the ipsilateral orbit and was partially relieved with paracetamol. During the last month, the episodes were more acute and of longer duration and were accompanied by nausea and vomiting of gastrobiliary content, without neurological focus. No convulsive episodes were reported.

Clinically, the patient presented with data of severe malnutrition, severe pain in the left orbit and proptosis. During the ophthalmological examination, decreased visual acuity and atrophy of papilla was observed. Likewise, there was an increase in volume in the left temporoparietal region of ~3 x 3 cm with a soft, painless consistency and fixed to deep planes.

Skull computed tomography with contrast was performed where a circumferential image that occupied the left orbital region was observed (1.7 x 1.6 cm) and was confined to the interior of the orbit (Figure 1).

Due to the presence of a compartment syndrome of the orbital cavity, an orbitofrontal craniotomy and intra-orbital tumor excision was urgently performed. Histopathological study of ES was obtained (Figure 2). After being admitted to the neurosurgery service, plain and contrast computerized tomographies of the skull were carried out (Figure 3) as well as MRI. Imaging studies showed extra-axial and intra- and extracranial biconvex lesions with involvement of the internal as well as external table of the frontotemporal region. Lesions were heterogeneous, predominantly isointense on T1 and T2 with some hyperintensities within the intracranial lesion and with heterogeneous enhancement with intravenous contrast (Figure 4). Spectroscopy showed an increase in the choline peak (Figure 5). Extension studies were performed and consisted of radiological tracing of long bones as well as bone scans in order to identify a primary lesion at another site. However, all studies were negative for some type of primary bone process. With the imaging data and severe progressive clinical deterioration, 5 days after the urgent procedure another surgical treatment was carried out: left temporoparietal craniotomy with excision of the lesion that was invading both the epidural as well as the epicranial regions. Ablation of a soft, yellowish-white colored

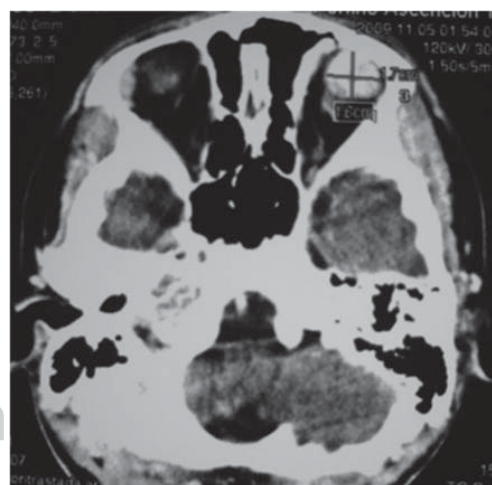
well-delineated, moderately vascularized and irregular in shape tumor was done (Figure 6).

The pathological specimen from the second surgery was sent for histopathological study. Immunohistochemistry was done and resulted in being positive for vimentin and CD99, confirming the diagnosis of extraosseous ES (Figure 7).

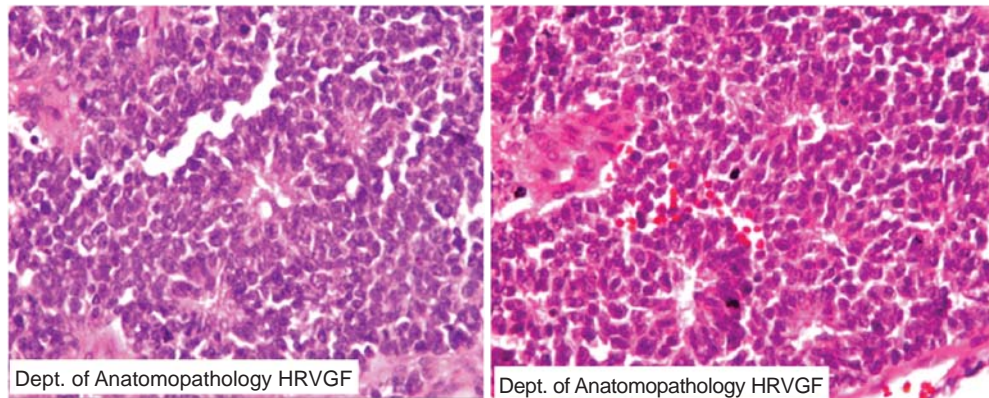
Five days after its removal, a control study was performed and showed a lesion with macroscopic characteristics similar to what was previously seen. The patient had noticeable deterioration in his neurological status. He died 13 days after the second intervention. He presented data of malnutrition, pneumonia and wasting syndrome of rapid evolution.

## DISCUSSION

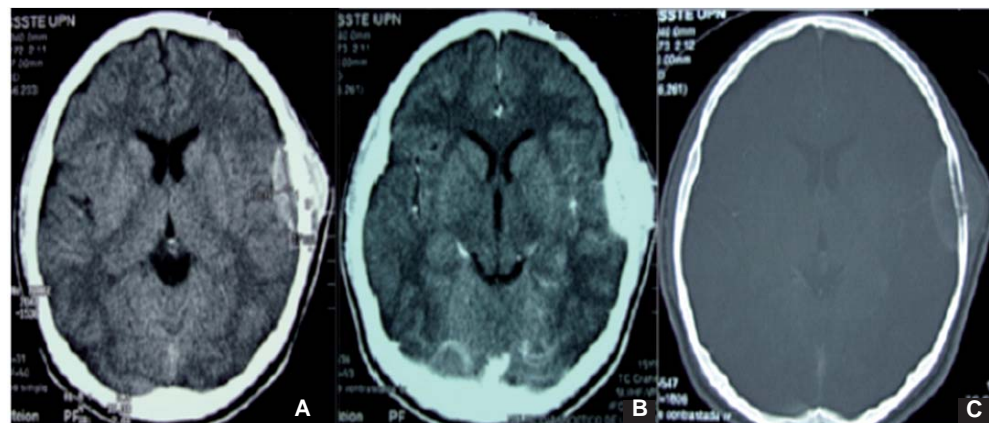
The emergence of extraosseous ES is extremely rare. The importance of early diagnosis is essential in the prognosis of these patients. The treatment is what has been established for those presenting with classic ES. Wide excisional surgery, chemo- and radiotherapy constitute, up to now, the ideal treatment. However, a poorly established diagnosis or one not done in a timely fashion increases the morbidity and mortality of the patients. In this case it is believed that, despite having made an early



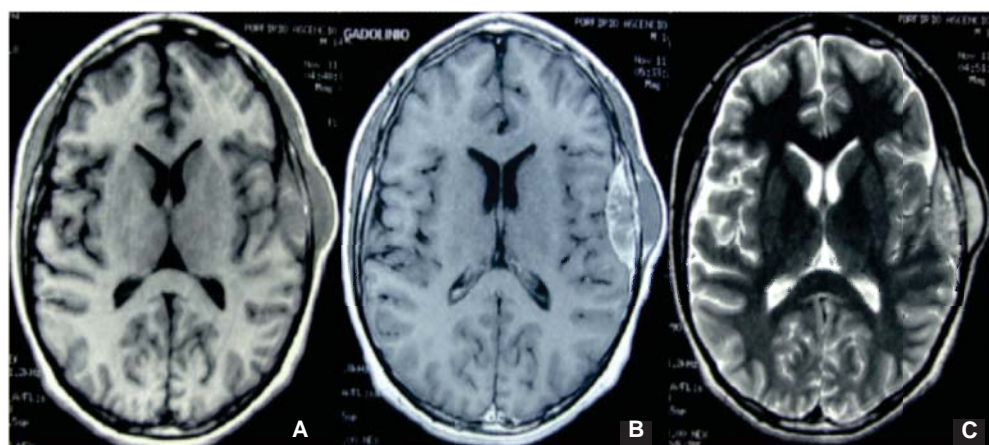
**Figure 1.** Contrast CAT. Left orbital base showing an intraconal tumor with intense and homogeneous enhancement with well-defined contours, causing mass effect, medially displacing the inferior rectus muscle without evidence of remodeling or erosion of the left sphenoid wing.



**Figure 2.** Histopathology of the intraorbital lesion (H&E stain) showing round, small, blue cells with a tendency to form rosette structures and more solid areas. Immunohistochemistry was positive for vimentin and CD99.

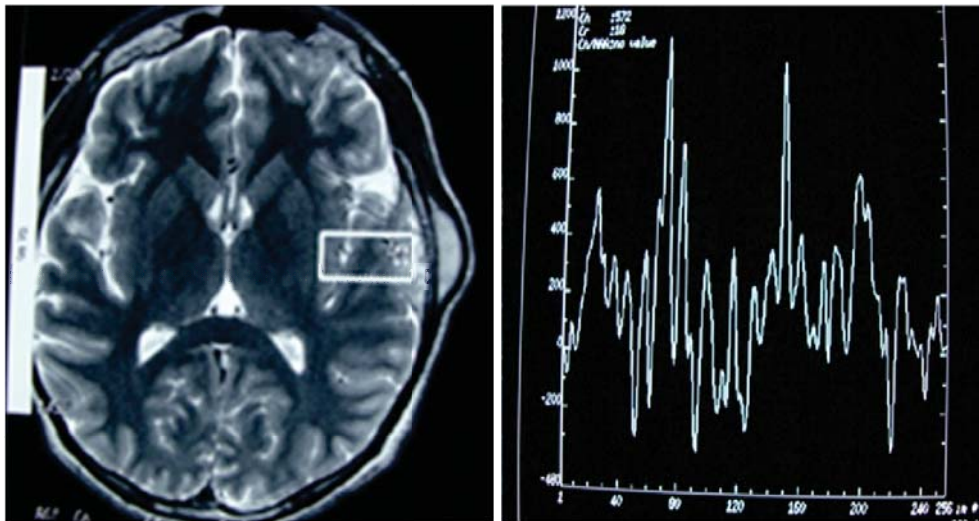


**Figure 3.** Plain skull tomography (A), with contrast (B) and filter bone window (C). At the diencephalic level an extraaxial tumor-like lesion is seen with well-defined margins and biconvex morphology, both intra- as well as extracranial, which affects the left frontal region. There is soft tissue involvement with lytic zones on the most caudal region of the lesion and intense enhancement with contrast media.

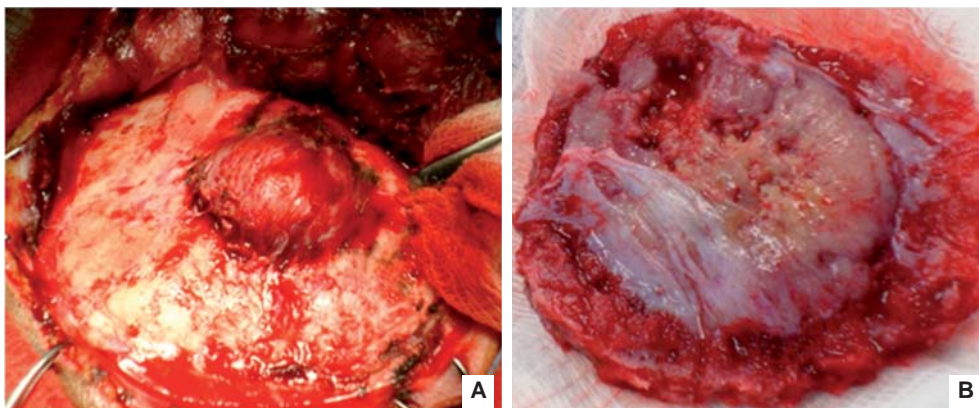


**Figure 4.** Magnetic resonance images in sequences T1 (A), T1 contrasted (B) and T2 (C). An extra-axial and extracranial lesion is seen, which is biconvex and with involvement of both the internal as well as external frontotemporal table, heterogeneous, predominantly isointense in T1 and in T2, with some hyperintensities within the intracranial lesion and with intense reinforcement and heterogeneous with contrast media of the internal lesion. No reinforcement of the epicranial external lesion is associated with a tumor of metastatic appearance.

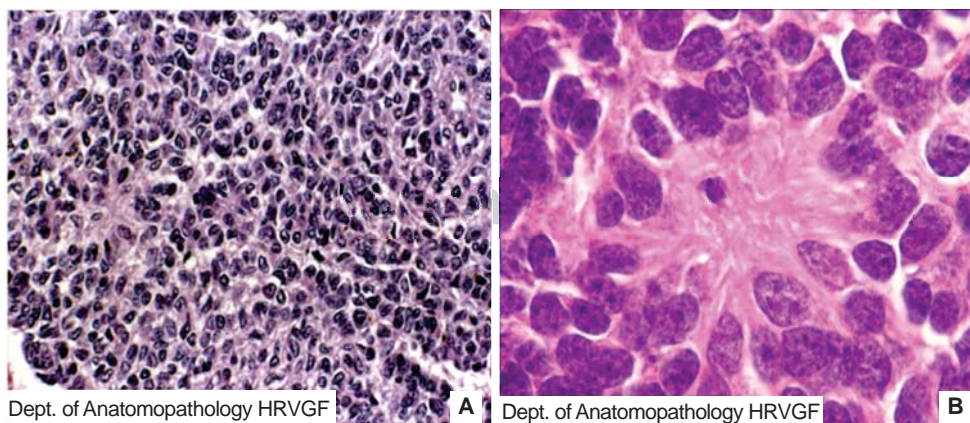




**Figure 5.** Univoxel spectroscopy with PRESS technique (*Point Resolved Spectroscopy*) and echo time of 144 msec with one ROI (*Region of Interest*) localized over the extra-axial intracranial lesion covering the cerebral parenchyma. A discrete increase in the choline peak and decrease of n-acetyl-aspartate was found as well as the presence of lipids and lactate, which supports the possibility of a metastatic lesion.



**Figure 6.** (A) Specimen of tumor tissue of ~3 x 3 cm, soft, yellowish-white, lobulated margins, irregular and infiltrating. (B) Zones of hemorrhage and adherence of the dura mater on the intracranial face are noted.



**Figure 7.** (A) Histopathological image of the temporo-parietal lesion. There is high cellularity with intense basophilia, small circular cells with homogeneous granular chromatin are observed. (B) Amplified image showing the pattern in rosettes, small and medium-sized, round and ovoid cells, coarse chromatin, pleomorphic nuclei and prominent nucleoli. It was positive for vimentin and CD99.

diagnosis upon his hospital admission, the disease was already at an advanced stage. This could have caused the malnutrition and immunosuppression and, subsequently, pneumonia, factors which constituted the direct causes of his death.

*Correspondence:* Dr. Juan Salvador Uribe Olalde  
E-mail: j.s.uribeolaldemed@hotmail.com

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