Primary Ewing’s sarcoma of the orbit: case report

Abstract

A 22-year-old man presented with tenderness and swelling of the left lateral part of the orbit. Computed tomography revealed a left intraorbital mass measuring 3 cm × 3 cm involving the left lateral wall of the orbit and the greater wing of the left sphenoid bone. Magnetic resonance imaging revealed that the intraorbital mass was extra-neuroaxial. During surgery, the tumor was seen to arise from the lateral wall of the orbit and infiltrate into the left temporal muscle. Following the surgery, the patient was administered radiation therapy for the whole cranium and chemotherapy for the residual tumors. However, the tumor recurred, and the patient died about 2 years following the first surgery because the tumor had metastasized to the lung. On light microscopy, the tumor cells were closely packed with uniform, small, and round cells. Immunohistochemical studies showed that the tumor cell membrane stained positive for MIC2. Furthermore, the MIB-1 labeling index was 36.2%. On electron microscopy, small quantities of cytoplasm containing glycogen accumulations without neurosecretory granules and neurofilaments were observed. Based on these results, the tumor was diagnosed to be primary Ewing’s sarcoma. Primary orbital Ewing’s sarcoma of the skull has been considered to be extremely rare, and a review of the literature was performed.

Key words

Ewing’s sarcoma · Peripheral primitive neuroectodermal tumor · Orbit · Ultrastructure · MIC2

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Introduction

Ewing’s sarcoma (ES) is a malignant tumor that was first described as an endothelioma of the bone by James Ewing in 1921.1 It typically occurs in the primary long bones during childhood and rarely occurs in the orbit of the skull.2 Cyto genetic analysis has led to the assertion that ES and peripheral primitive neuroectodermal tumors (pPNETs) constitute a family of tumors with a common cytogenetic translocation, t(11;22)(q24;q12).3-5 To our knowledge, primary pPNET/ES family tumors (pPNET/ESFTs) and ES of the orbit have been reported in only 21 and 13 cases, respectively (Table 1).6-26 In this article, we report a rare case of primary ES of the orbit in which immunohistochemical and electron microscopic studies were performed. We then discuss the immunohistochemical and ultrastructural features of ES.

Clinical summary

A 22-year-old man presented with tenderness and swelling of the left lateral part of the orbit in December 1995. Upon his admission to our institution, a neurological examination revealed left proptosis and left ophthalmoparesis. Computed tomography (CT) revealed an intraorbital mass measuring 3 cm × 3 cm involving the left lateral wall of the orbit and the greater wing of the sphenoid bone. Magnetic resonance imaging (MRI) revealed an intraorbital mass that was extra-neuroaxial and homogeneously enhanced on T1-weighted images after the intravenous administration of gadolinium-diethylene-triaminepentaacetic acid (Gd-DTPA) (Fig. 1). The tumor was removed on April 1, 1996. The tumor was a gelatinous, soft mass that was gray in color and did not bleed. The tumor arose from the lateral wall of the orbit and infiltrated into the left temporal muscle but did not infiltrate into the left periorbit. After the surgery, his left proptosis and ophthalmoparesis disappeared; then, he was administered radiation therapy for the whole cranium
(52 Gy) and chemotherapy for the residual tumors. The primary lesion was supposed to have been in the left orbit, as no other lesions were found in the bone scan. Recurrence of the tumor was found by the MRI 8 months after the first surgery, and second removal of the tumor was performed on December 19, 1996. The resected tumor was 2.5 cm × 2.5 cm × 2.5 cm, and the tumor had infiltrated into the wide range of the frontal bone. Therefore, a second round of focal radiation therapy was administered, but this patient died 1 year following the second round of radiation therapy because the tumor had metastasized to the lung.

**Pathological findings**

The resected tumor tissues of the first surgery were fixed in 10% formalin, embedded in paraffin, and then processed for light microscopy. Tissue sections were stained with hematoxylin and eosin (H&E) and examined immunohis-tochemically. A Dako labeled streptavidin biotin (LSAB) kit was used for immunostaining. The primary antibodies used are shown in Table 2. For electron microscopy, some specimens were fixed in 2.5% glutaraldehyde cacodilate