

Ewing's Sarcoma Localized in the Mandible: A Case Report

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ABSTRACT

Ewing's sarcoma is one of the most aggressive primary bone tumours. Ewing's sarcoma arising from the bones of the head and neck region is extremely rare; only 4–9% of all Ewing's sarcoma originate in this region. We report a case of Ewing's sarcoma localized in the mandible because of its unusual presentation.

Keywords: Child, Ewing's sarcoma, mandible

Sarcoma de Ewing Localizado en la Mandíbula: Reporte de un Caso

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RESUMEN

El sarcoma de Ewing es uno de los tumores primarios óseos más agresivos. El sarcoma de Ewing de los huesos de la región de cabeza y cuello es extremadamente raro: sólo 4–9% de todos los sarcomas de Ewing se originan en esta región. Reportamos un caso de sarcoma de Ewing localizado en la mandíbula, debido a su inusual presentación.

Palabras claves: Niño, sarcoma de Ewing, mandíbula

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INTRODUCTION

Ewing's sarcoma (ES) is one of the most aggressive bone tumours known and accounts for 4–7% of primary bone malignancies (1). Ewing's sarcoma usually occurs in the first or second decades of life and males are more often affected than females [male:female = 3:2] (2). Primary ES arising from the mandible is extremely rare (3). Only 4–9% of all ES originate in the head and neck region, usually involving the mandible (4). Most ES lesions in the mandible have been located in the posterior regions (1). Most authors claim a better prognosis for ES of the head and neck region as compared to that arising in other anatomic locations (4, 5).

In this article, we report a case of ES localized in the mandible because of its unusual presentation.

CASE REPORT

A 10-year old girl was admitted with the complaint of painless swelling on the left side of her face. The swelling started one month ago, and then it gradually enlarged. On examination, a diffuse mass, non-tender and hard with a size of 10 x 10 cm

diameter was noted over the left mandible. Other physical examination was unremarkable. The results of haematological and biochemical investigations were within normal ranges. Examination of the bone marrow was normal. On skull roentgenogram, the sixth lower tooth on the left side was displaced medially (Fig. 1).

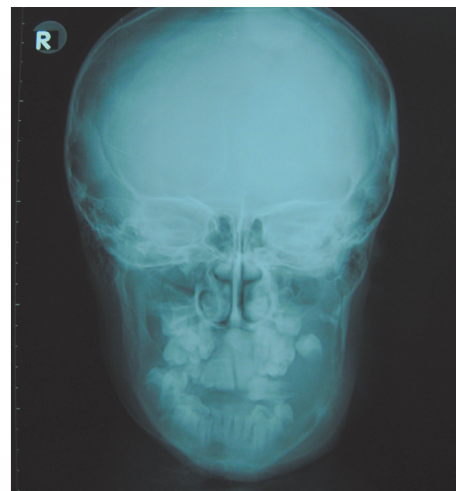


Fig. 1: On skull roentgenogram, the left 6th lower tooth is displaced medially.

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Computed tomography (CT) examination of the neck demonstrated a 7 x 4.5 cm mass on the left of the mandible causing bone expansion and destruction and reaching to soft tissue planes (Fig. 2).

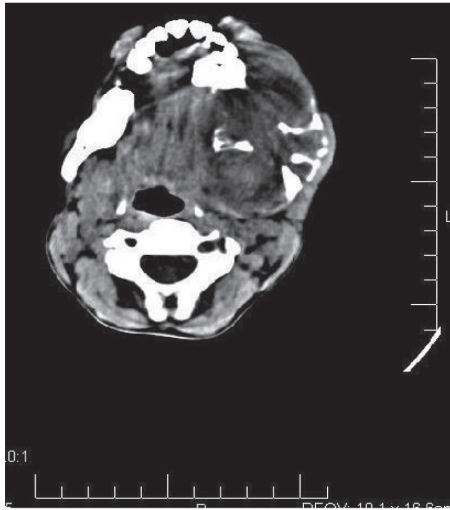


Fig. 2: Computed tomography examination of the neck demonstrates a 7 x 4.5 cm mass on the left of the mandible causing bone expansion and destruction and reaching to soft tissue planes.

On post-contrast T1-weighted magnetic resonance imaging (MRI) of the left temporomandibular joint, a 7 x 4.5 cm mass with a lobulated contour was observed on the left mandible corpus causing bone expansion and destruction and reaching to the neighbouring soft-tissue planes. The mass was displaying peripheral contrast enhancement, fluid levels and having regions of bleeding anteriorly (Fig. 3).



Fig. 3: Post-contrast T1-weighted magnetic resonance imaging in coronal plane demonstrates a mass with peripheral contrast enhancement, fluid levels, and regions of bleeding anteriorly.

Computed tomography of the abdomen and thorax and MRI of the brain were normal. Histopathological examination of the mass showed small, round cells with hyperchromatic nuclei, which was consistent with malignant small round cell tumour. Demonstration of intracytoplasmic glycogen was done by periodic acid-Schiff (PAS) stains. A positive expression for CD99 was also noted on tissue samples consistent with

ES. The patient was planned for chemotherapy regime (EURO-Ewing 99-Pulmon VIDE Protocol).

DISCUSSION

Ewing's sarcoma is the second most frequent primary bone tumour in children and adolescents (6). Primary malignant tumours of the jaws are rare and the diagnosis and treatment of ES can be especially challenging (2). Approximately 90% of reported cases occurring in the mandible have been primary lesions and 10% have been metastases (1).

Clinical symptoms such as swelling, pain and sensory disturbances are rather nonspecific and can sometimes be misleading. Ewing's sarcoma usually occurs in the first or second decades of life. The median age for a patient with the ES family of tumours is 15 years and more than 50% of patients are adolescents (7). Our patient was also in the first decade of life.

Although ES can occur in any part of the skeleton, it shows a particular predisposition for the long bones of the extremities and pelvis, which account for 58% and 20%, respectively. Only 3% arise in the skull and 7% in the ribs (8). Ewing's sarcoma arising from the bones of the head and neck region is extremely rare (9). Siegal *et al* (4) reported 29 cases of ES of the head and neck region, which represented 4% of all Ewing's sarcoma cases reported by the Intergroup Ewing's Sarcoma Study (IESS) group.

Histologically, ES is composed of small round cells with oval or round nuclei and scarce clear cytoplasm, arranged in sheets. This tumour has been classified as one of the "small, round, blue cell tumours of childhood", and differential diagnosis should consider small cell osteosarcoma, malignant lymphoma, neuroectodermal tumours and metastatic neuroblastoma (9). Our case was confirmed with demonstration of intracytoplasmic glycogen by PAS stains and immunohistochemical staining positivity for CD99.

The typical presentation of ES in the mandible is one of insidious onset of pain and swelling (9).

The treatment of ES involves a multimodal approach. Multiagent chemotherapy is important because it can shrink the tumour rapidly and usually is given before local control is attempted. Ewing's sarcoma is considered a radiosensitive tumour and local control may be achieved with radiation or surgery (10). In our case, chemotherapy (EURO-Ewing 99-Pulmon VIDE Protocol) was initiated.

In conclusion, despite ES of the mandible being rare, it is important to consider this diagnosis in a child who presents with prolonged swelling of the face.

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