Case report 786

Akila Narasimhan, M.B.B.S. 1, M. Sundaram, F.R.C.R. 1*, S.M. Chandy, M.D. 2, M. Washburn, M.D. 1, R.R. Williams, M.D. 1

1 Department of Radiology, Christian Medical College and Hospital, Vellore, South India
2 Department of Pathology Christian Medical College and Hospital, Vellore, South India

Fig. 1. An ill-defined spiculated mass corresponding to the site of swelling is seen in the region of the left cheek. Neither the origin nor the extent of the lesion can be established from this.

Fig. 2. A Destruction of the left zygoma with adjacent spiculated bone and a large soft tissue mass is demonstrated in this computed tomogram (CT). B Contrast-enhanced CT shows the mass to be vascular extending medially into the infratemporal fossa and invading the left temporal lobe.

Clinical information

A 15-year-old male Asian-Indian child presented with left-sided swelling of the face of 4 months' duration. It had been slowly increasing in size over the preceding 2-3 months but was not associated with pain, fever, or mechanical disability. Physical examination revealed no abnormality elsewhere, and there was nothing of significance in the patient's past medical history. No family history of tuberculosis was reported. Clinical examination revealed a hemispherical swelling over the left zygoma, 20 cm x 20 cm, smooth and non-pulsatile. About three or four upper deep cervical lymph nodes were palpable. No abnormality was found in the oral cavity.

Entities considered in the clinical diagnosis were soft tissue sarcoma, lymphoma, and "cold abscess" secondary to tuberculosis. A radiograph of the facial bones (Fig. 1) showed an ill-defined sclerotic, spiculated mass. The abnormality was inadequately defined and a computerized tomogram (CT) without and with iodinated contrast medium was obtained (Fig. 2).

The CT examination revealed destruction of the left zygoma associated with a large soft tissue mass. The medical extent of the soft tissue component of this tumor extended intracranially, invading the infratemporal fossa and left temporal lobe.

The appearance of the patient's chest radiograph was within normal limits and no other skeletal or visceral lesions were apparent.

A trucut biopsy of the tumor and biopsy of a cervical lymph node were carried out. The lymph node showed reactive hyperplasia.
Diagnosis: Ewing’s sarcoma of the left zygoma

The differential diagnosis chiefly includes osteosarcoma, lymphoma, or other round cell tumor.

Discussion

Ewing’s sarcoma derives its name from James Ewing, who distinguished it from osteosarcoma and designated it a “diffuse endothelioma of bone” [3]. Almost 90% of individuals with this neoplasm are between the ages of 5 and 30 years, the highest incidence occurring in patients between the ages of 10 and 15 years [4]. Ewing’s sarcoma may develop in any bone in the body, but in at least two-thirds of cases occurs in the sacrum, pelvis, and bones of the lower extremities [4]. The mandible and maxilla account for approximately 2% of cases, while other facial bone involvement has been stated to occur in less than 1% of reported cases [6].

The plain film radiographs were of limited value in the case in determining the origin or extent of the tumor, but showed bony spiculation in the region of the clinically obvious mass (Fig. 1). The CT scan showed the partially destroyed zygoma to be the epicenter of the tumor, suggesting an osseous origin of the mass (Fig. 2A). The contrast-enhanced scan showed the soft tissue component of the tumor to be vascular. Absence of skeletal or visceral abnormality elsewhere suggested a sarcoma of bone, although its histological nature was uncertain. The trucut biopsy showed skin and connective tissue infiltrated by a tumor composed of solid clusters of round-to-oval cells with scanty-to-moderate cytoplasm and hyperchromatic round-to-oval nuclei. Mitoses and foci of necrosis were present (Fig. 3). Immunocytochemical tests using anti-leukocyte common antigen, epithelial membrane antigen, cytokeratin, vimentin, and desmin were negative. Periodic acid-Schiff stain showed focal positivity in the tumor cells and the appearances were felt to be consistent with a diagnosis of Ewing’s sarcoma infiltrating the skin and connective tissue of the temporal region of the face.

An unusual feature of the clinical presentation was the complete lack of pain, discomfort, or any mechanical disability despite the enormity of the swelling and intracranial extension.

Ewing’s sarcoma of the zygoma is exceptionally rare; its occurrence in neighboring bones of the maxilla and mandible, although still rare, is relatively more frequently reported [5]. The rarity of this tumor in these bones in comparison to the more common sites of occurrence has been attributed to the paucity of hematopoietic marrow at the locations. Unlike gnathic osteosarcomas, no sub-group of gnathic Ewing’s tumor exists, presumably because no body of opinion believes they are any different microscopically or behaviorally from Ewing’s sarcoma in more conventional sites.

Ewing’s sarcoma in this location has, as elsewhere, been subject to therapeutic procedures that have included surgical resection, radiation therapy, and chemotherapy. Our patient has been treated with a combination of chemotherapy and radiation therapy, and remains alive with disappearance of facial swelling 10 months after the diagnosis was established (Fig. 4).

In the United States of America, Ewing’s sarcoma shows a predilection for patients of European de-