Primary Extraskeletal Ewing Sarcoma Originating from Chest Wall in a Child

Serhat Avcu, Hüseyin Akdeniz, Aydın Bora, Harun Arslan, Özkan Ünal

Yüzüncü Yıl University Medical Faculty, Department of Radiology, Van, Turkey

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Correspondence:*Serhat Avcu,*
YYU Araştırma Hast. Radyoloji Anabilim Dalı, 65200 Van, Turkey
Phone: +905056480544
E-mail: serhatavcu@hotmail.com

ABSTRACT

We report the ultrasonography (US) and computed tomography (CT) findings of a rare case of extraskeletal Ewing sarcoma involving the left chest wall with a diameter of 4cm in a 5 years old girl. The lesion was heterogenously hypoechoic on sonography with a lobulated smooth contour. On CT, the lesion had central necrotic areas and was enhancing heterogeneously. With these US and CT findings surgical excision of the tumor was performed. Histopathologically the mass was evaluated as round cell malignant neoplasm (Ewing tumor/ "Askin tumor"/ primitive neuroectodermal tumor (PNET)). Chemotherapy and radiotherapy were planned to the patient.

Keywords*: Ewing sarcoma, chest wall, ultrasonography, CT*

INTRODUCTION

Ewing sarcoma (ES) is a highly malignant bone tumor composed of uniform round small cells. It was first described by James Ewing in 1921 as a malignant tumor of the shaft of the long bones in children and young adults. Later, malignant soft-tissue tumors morphologically indistinguishable from ES were reported and termed extraskeletal Ewing sarcoma (EES). Recently, a single biologic entity, Ewing sarcoma family of tumors (ESFT) has been proposed and gradually accepted. The entity includes ES, EES, and peripheral primitive neuroectodermal tumor (PNET), which shows more neural differentiation than ES (1). EES is a rare, malignant soft tissue neoplasm histologically similar to skeletal Ewing sarcoma. It occurs predominantly in adolescents and young adults between the ages of 10 and 30 years, and follows an agressive course, with a high recurrence rate. The involvement of the breast is uncommon (2). We report the ultrasonography (US) and computed tomography (CT) imaging findings in a case of EES involving the left chest wall of a 5 years old girl.

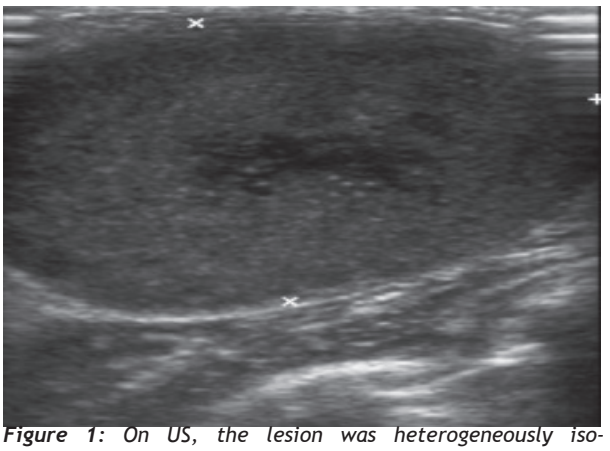

Figure 1: *On US, the lesion was heterogeneously iso-hyperechoic, with central hypoechoic necrotic areas.*

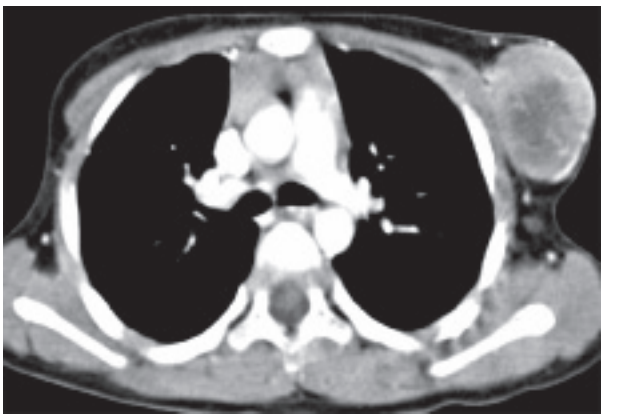

Figure 2: *On CT, the mass displayed heterogeneous enhancement, central necrotic areas, and a smooth contour.*

CASE

A 5-year-old girl with a mass on her left chest wall for 3 months was sent to the radiology department for US and CT examinations. These examinations showed a well marginated mass lesion involving the left chest wall. On US, the lesion was heterogeneously iso-hyperechoic, with central hypoechoic necrotic areas (Figure 1). On CT, the mass was homogeneously isodense with muscle and showed a maximal oblique diameter of 4cm (Figure 2). The mass had a smooth contour both on US and CT, with some centrally located necrotic areas, and displayed heterogeneous enhancement. There was no invasion of the ribs. Differential diagnosis of benign versus malignant soft-tissue tumor was suggested on the basis of these US and CT findings. Resection was performed and the histopathological examination revealed a round cell malignant neoplasm (Ewing tumor/ "Askin tumor"/ primitive neuroectodermal tumor (PNET)). Chemotherapy and radiotherapy were planned for the patient.

DISCUSSION

EES is an undifferentiated, small, round-to-oval cell tumor of uncertain origin. It differs in presentation from skeletal ES in several respects. The average age of occurrence of EES is 20 years, in contrast to 10 years for skeletal ES. EES occurs equally in both sexes, whereas skeletal ES has a 2:1 male predilection (3,4). EES commonly affects the extremities (especially the lower extremities), soft tissues of the trunk, such as paravertebral and intercostal regions, head and neck, pelvis, and peritoneum. In contrast, skeletal ES has a predilection for the long bones of the lower extremities (3). Other rare reported locations of EES are various and include the mediastinum, heart, external genitalia, and broad ligament (4,5). In our case, the tumor was located on the left chest wall. The tumor tends to spread locally, infiltrating deep fascial spaces and invading muscles or skeletal structures as well as other adjacent structures (6). In our case however, the mass did not invade the adjacent soft tissue or bony structures. EES occurs less frequently than ES of bone. The compiled data including 1,505 patients of ESFT from the United States, Europe and Japan showed that EES cases represented 8% compared to ES of bone with 87%. The remaining 5% of the cases were diagnosed as peripheral PNET (1).

The reported CT finding of EES is most commonly a heterogeneously enhancing mass (5,8,9). Occasionally a central, nonenhancing, low-density area is seen within the mass (9). In our case, the mass was homogeneously isodense compared with muscle. On MR images, the EES is generally of low to isointense signal compared with muscle on T1-weighted images, of high signal intensity on T2-weighted images, and exhibits heterogeneous enhancement (10-12). However, it is stated that the MR findings of this tumor are nonspecific (2). MR examination was not performed in our case. It is known that both local recurrences and distant metastases are common in EES (7). It is stated that the 5-year survival rate of EES is 61%, whereas that of skeletal ES is approximately 10% (13).

In conclusion, although quite rare and indistinguishable clinically and radiologically from other soft-tissue tumors, EES should be kept in mind in case of chest wall tumoral mass lesions.

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