Cancer Imaging (2008) **8,** 156–158
DOI: 10.1102/1470-7330.2008.0023

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CASE REPORT

Extraskeletal Ewing sarcoma of the abdominal wall

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Date accepted for publication 14 July 2008

Abstract

Ewing sarcoma is most commonly a bone tumour which has usually extended into the soft tissues at the time of diagnosis. Exceptionally, this tumour can have an extraskeletal origin. Clinical or imaging findings are non-specific and diagnosis is based on histology. We report a case of an extraskeletal Ewing sarcoma developed in the soft tissues of the abdominal wall in a 35-year-old woman who presented a painful abdominal wall tumefaction. Ultrasongraphy and computed tomography showed a large, well-defined soft tissue mass developed in the left anterolateral muscle group of the abdominal wall. Surgical biopsy was performed and an extraskeletal Ewing sarcoma was identified histologically.

Keywords: *Ewing sarcoma; soft tissue neoplasms; imaging; ultrasonography; computed tomography.*

Introduction

Ewing sarcoma is a bone neoplasm that usually occurs in children and young adults and which can extend into the soft tissues. Exceptionally, this tumour can have an extraskeletal origin. We report a case of Ewing's tumour involving the left anterolateral muscle group of the abdominal wall without associated skeletal location and we describe ultrasonography and computed tomography (CT) imaging features of this disorder.

Case report

A 35-year-old woman presented with a 2-month history of a painful swelling of the left abdominal wall. Physical examination revealed an irregular mass approximately 6 cm in diameter on the left side of the abdominal wall.

Ultrasonographic evaluation revealed a large hypoechoic and inhomogeneous well-defined soft tissue mass, developed in the left anterolateral muscles of the abdominal wall measuring about 6.5×4 cm (Fig. 1a). The study in Doppler mode showed mixed, arterial and venous flow signals within part of the tumour (Fig. 1b). The ultrasonography of the abdomino-pelvic cavity showed no anomaly.

An abdominal helical CT evaluation was performed using a helical single detector CT (Siemens Somatom Plus 4). Scanning covered from the diaphragm to the pubic bone before and after intravenous contrast with arterial and portal venous phases. The patient received intravenous contrast material (Scanlux) administered at a rate of 3 ml/s and a volume of 120 ml with a power injector. Collimation was 3 mm and pitch was 1.5. The unenhanced study showed a large mass involving the left anterolateral muscle group of the abdominal wall. This mass was isodense to the adjacent muscles with hypodense areas and no evidence of calcification or fatty areas (Fig. 2a). After bolus contrast administration, the mass was well circumscribed and lobulated with heterogeneous but intense enhancement (Fig. 2b).

A whole body bone scintigraphy study showed normal skeletal uptake of tracer and no evidence of bone involvement. According to the imaging findings, the diagnosis of rhabdomyosarcoma was suspected. The patient underwent an incisional biopsy and the tumour was determined to have the microscopic characteristics of a Ewing sarcoma involving the soft tissues.

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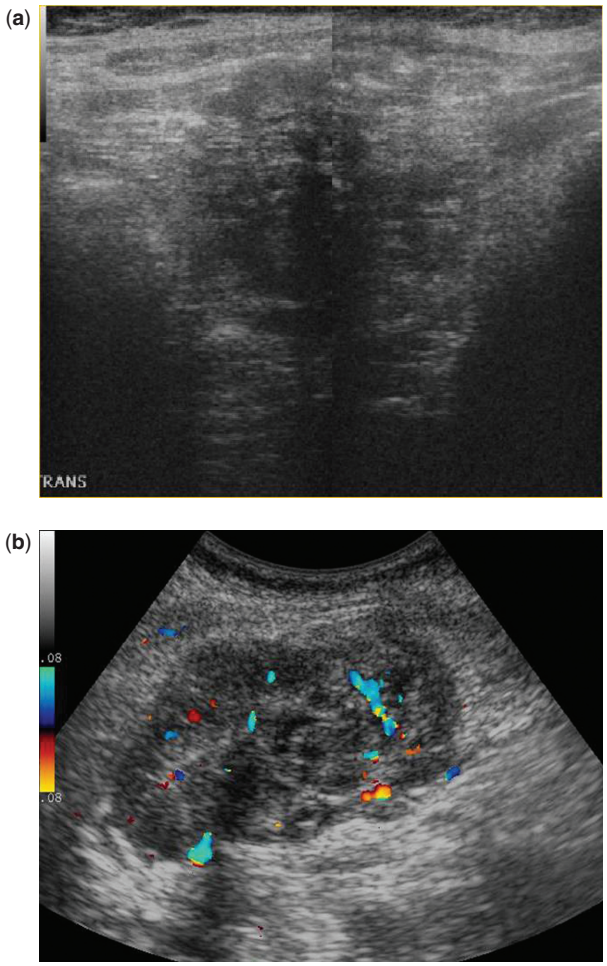

Figure 1 **Ultrasonographic evaluation. (a) Transverse gray-scale sonogram showing a large hypoechoic and inhomogeneous well-defined soft tissue mass developed in the left anterolateral abdominal wall. (b) Doppler ultrasound scan shows flow signals within part of the tumour.**

Neoadjuvant chemotherapy was carried out including vincristin, cyclophosphamid and adriamycin for three cycles but the patient showed a poor response and surgery was required.

Discussion

Ewing sarcoma commonly arises from bone and it can involve soft tissues at the time of the diagnosis. Rarely, Ewing sarcoma may have an extraskeletal origin. Soft tissue Ewing sarcoma is a rapidly growing, round-cell, malignant tumour which can reach 10 cm by the time of the diagnosis[1]. Young adolescents and adults between the ages of 10 and 30 years are predominantly affected with a slight predominance in males[2,3]. However, some cases have been reported in patients between 14 months and 77 years of age[4]. Commonly affected extraskeletal sites are the paravertebral spaces, lower extremities, head and neck, and pelvis[2,3,5–9].

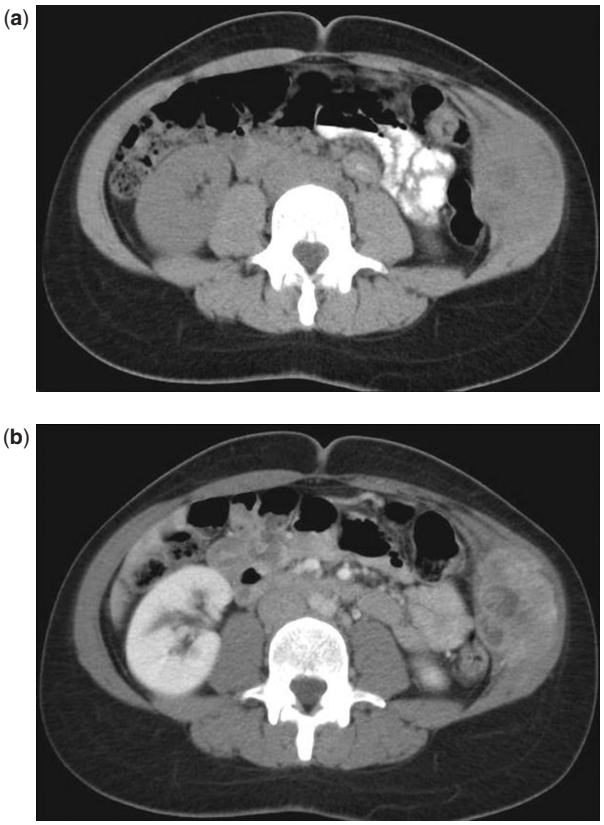

Figure 2 **CT evaluation. (a) Axial CT slice showing a mass involving muscles of the left anterolateral abdominal wall. (b) Enhanced slice, after bolus contrast administration, showing a well-defined mass with intense and inhomogenous enhancement.**

Other rare reported locations of extraskeletal Ewing sarcoma are various and include the retroperitoneum, orbit, skin, and chest wall[10–13]. To the best of our knowledge, only one previous paper reported a Ewing sarcoma involving the abdominal wall[14]. The most frequent presenting symptom is a rapidly growing mass with local pain.

The imaging features soft tissue Ewing sarcoma are non-specific[3,8,9,11]. It often presents as a well-limited mass which should not be confused with a benign lesion. Ultrasonography often shows a hypoechoic and heterogeneous mass with intra-tumour flow signals in a Doppler study. CT shows a large, sharply delineated mass which is relatively hypodense or isodense compared to the adjacent muscle. It can contain areas of lower attenuation than the adjacent muscle. Post-contrast medium enhancement is intense and heterogenous. Hypodense foci are frequent in the large mass and are due to intratumour necrosis[3,5,9]. On magnetic resonance imaging (MRI), this tumour is often of low to intermediate signal intensity on T1-weighted images; of high signal intensity on T2-weighted images, and exhibits heterogeneous contrast enhancement[1,9,15]. Sometimes, MRI shows a homogeneous, moderate enhancement on

contrast-enhanced T1-weighted images[9]. Spontaneous tumour haemorrhage and adjacent bone destruction have been reported in some cases[3]. The secondary osseous involvement is rare even when the mass is located near a bone segment. It can result in cortical erosion and/or a periosteal reaction[2]. MRI is able to provide evidence of non-involvement of the marrow cavity.

Radiological differential diagnosis includes rhabdomyosarcoma, malignant fibrous histiocytoma and dedifferentiated liposarcoma. Diagnosis is confirmed by CT-guided core-needle biopsy or pathological analysis of the operative specimen. Extraskeletal Ewing sarcoma is confirmed by characteristic features on histologic analysis, histochemistry, immunohistochemistry and electron microscopy. Differential diagnoses include other small, blue round cell tumours (SBRCTs) and other members of the Ewing family of tumours such as the primitive neuroectodermal tumour (PNET)[12].

The mainstay treatment should include multi-agent chemotherapy and aggressive surgical treatment. Tumours that are not appropriate for surgical resection or have positive margins are treated with radiation[1,5,14].

The prognosis for extraskeletal Ewing sarcoma appears more favourable than that of Ewing sarcoma in bone[12]. Two previously published series devoted to the study of extraskeletal Ewing sarcoma stated that it is a curable disease and has the best prognosis in young patients (age <16 years) treated with complete resection with wide surgical margins in conjunction with chemotherapy[2]. Ahmad *et al.* also demonstrated that tumour size did not have a significant effect on overall 5-year survival or disease-free survival[2]. There was no differences in overall or disease-free survival between the patients with metastatic disease and those without[2]. Lung, liver, brain and bone are the most common metastatic sites[3].

Imaging is useful for evaluation of the rate of resectability and the tumour response to treatment[5]. Tumours that are not appropriate for surgical resection or have positive margins are treated with radiotherapy.

In summary, extraskeletal Ewing sarcoma arising from soft tissues is extremely rare. Although its radiological features are non-specific, it should be kept in mind in the differential diagnosis of soft tissue tumours in young people.

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