Extraosseous Ewing’s Sarcoma: A Case Report with MR Manifestations

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CASE REPORT

Extraosseous Ewing’s Sarcoma

Extraosseous Ewing’s sarcoma (EES) is a rare soft tissue tumor that is histologically indistinguishable from the osseous form. We present the clinical course and magnetic resonance (MR) findings of a 30 year-old man with an EES in the right thigh. The soft tissue mass was well-defined with tissue characteristics of heterogenous intermediate signal intensity on T1- and high signal intensity on T2-weighted images that became enhanced after gadolinium contrast agent administration. Central tumoral necrosis and hemorrhage were observed. The lesion was subsequently proved to be an EES after resection. Unfortunately, the patient died because of multiple metastasis ten months after discovery of the tumor.

Key words : Ewing’s sarcoma, MR, soft tissue tumor

CASE REPORT

A 30 year-old man had a palpable mass at the posteromedial aspect of right thigh and numbness of the right foot for 1 month. On physical examination, a mass about 6 cm was palpated. It was not movable and not tender. Plain radiography of the right femur on AP and lateral views showed normal bony structure. MR imaging was performed on a 0.5 Tesla superconductive system (GE-MAX, Milwaukee, Wisconsin, USA). An 8 × 8 × 5 cm³ lobulated mass attached to the adductor magnus muscle was noted. The heterogenous mass had intermediate signal intensity on T1 weighted images (TR/TE: 500/20 msec) (Fig. 1a), and high signal intensity on T2 weighted images (TR/TE: 1600/90 msec) (Fig. 1b) without evidence of fat suppression on STIR images (TR/T1/TE: 1500/100/20msec) (Fig. 1c). The mass was heterogously enhanced after contrast enhancement (Fig. 1d). In the center of the mass, several well-defined areas of variable sizes with low T1 and high T2 signal intensities were identified. There was minimal perineoplastical infiltration noted around the soft tissue mass but it was distinctively separated from the femur.

The patient underwent surgical excision under the impression of sarcoma. The excised soft tissue mass measured 8 × 8 × 4 cm³ and was attached to the adductor magnus muscle, invading the sciatic nerve. Grossly, this tumor was soft and elastic and dull grayish-pink with multiple central necrotic foci. Microscopically, neoplastic proliferation of ovoid or spindle cells with vesicular nuclei and scanty neoplasm was noted in a pseudorosette pattern with stromal invasion (Fig. 2).

Immunohistochemical stain was positive for vimentin, neuron-specific enolase (NSE), S-100, neurofilament protein (NFP) and chromogranin.

Neoadjuvant chemotherapy was administered with a regimen of CYVADDIC (Cyclophosphamide and Endoxan, 500mg/m² on day 1; Vincristin, 1.4mg/m² on day 1; Adriamycin and Epirubicin, 50mg/m² on day 1; Dacarbazine and DTIC, 400mg/m² on days 1-3) for four courses at one-month intervals. Two courses of post-operative radiotherapy (6200 cGy 45 times and 3300 cGy 11 times) were also given. Half a year after surgery, a follow up bone scan revealed multiple metastasis in the right tibia and fibula. Spinal metastasis of the L4 vertebra was noted on MR images 9 months after surgery. The patient then received posterior instrumentation and posterolateral fusion with a bone graft and 3 level laminectomy. The patient died one month after lumbar spinal surgery. The duration from presentation to death was ten months.
DISCUSSION

EES is a rare soft tissue tumor. The sarcoma is composed of small undifferentiated, round to oval cells. It is histologically and ultrastructurally indistinguishable from the osseous form. The diagnosis of EES is made on the basis of histological findings in the absence of bony involvement at the time of presentation. Genetic predisposition to EES is similar to the osseous form with the same translocations involving band q12 of chromosome 22 [2]. There is also a slight predominance in men, with a male to female ratio of 1.5:1 [3]. Patients’ ages range from 20 months to 30 years old, with a median age of 20 years.

EES can be located in almost all parts of the body with a predilection for the paravertebral region in the chest and the soft tissues of the extremities [4]. EES is usually manifested as a solitary soft tissue mass between 5-10 cm in diameter and may be superficially or deeply located. EES is usually painless, but in occasional cases, symptoms of local heat and tenderness are reported, as in the case of Tay et al and also in our patient [5]. Numbness and pain at the dorsal and plantar side of the right foot in our patient was caused by sciatic nerve involvement. Shiang et al reported an EES in the left anterolateral chest wall with numbness over the left arm caused by intercostal nerve invasion [6].

On conventional radiography, EES is usually an extraosseous soft tissue mass without any calcification matrix. However, reactive changes such as periosteal proliferation, cortical thickening, bony erosion or sclerosis may occur [2]. When these reactive changes are present, histological examination is required to differentiate EES from the osseous form.

The role of MR imaging in EES is to delineate the extent of tumor and to confirm the soft tissue mass is totally extraosseous. Tissue characterization of EES is non-specific, that is, there is intermediate signal intensity on T1 weighted images and high signal intensity on T2 images [3].

The immunohistochemical diagnosis of EES varies greatly. Positive staining of vimentin is seen in 50-66% of cases. Less frequently, NSE stains positive. NFP, S-100 and chromogranin are usually negative [7]. In this case, a positive stain for vimentin, NSE, NFP, S-100 and chromogranin was observed. Therefore, the immunohistochemical diagnosis is pertinent. Although the diagnosis of EES is made primarily based on histology, some clinical and radiological features may help in the differential diagnosis from other soft tissue tumors such as malignant fibrous histiocytoma and liposarcoma. Specific tissue characterization such as calcification in malignant fibrous histiocytoma and fat in well differentiated liposarcoma is helpful in the diagnosis. Moreover, malignant fibrous histiocytoma usually presents after the age of 45 and liposarcoma is found after the age of 30 [4]. The mean age of EES patients is 20. Fatty or calcified contents are unusual in EES.

The treatment of EES includes combined surgical resection, chemotherapy and post-operative radiotherapy [5]. Local recurrence and metastasis are common. The most common sites of metastasis are the lung and bone. In most cases, metastasis is found within a few
months after surgical removal of the primary tumor. The prognosis is grave. The two-year disease-free survival rate is less than 50% [5]. Although our patient had combined therapy consisting of surgery, neoadjuvant chemotherapy and radiotherapy, metastasis was noted 6 months after surgery. In this case, the patient survived for only 10 months after surgery.

Although the MR appearance of EES is non-specific, MR imaging is used in defining the extent of tumor and the involvement of adjacent structures. EES should be included in the differential diagnosis of non-calcified soft tissue tumors in specific anatomic sites and age groups.

REFERENCES
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骨髂尤文氏肿瘤是一种罕见的软组织肿瘤，显微下其组织构造，骨髂尤文氏肿瘤与一般骨髂尤文氏肿瘤是难以区分的。本文报告一名二十六岁男性在右大腿的骨髂尤文氏肿瘤，其临床过程及磁振造影表现。此肿瘤经外科切除后病理证实为骨髂尤文氏肿瘤。此病患进行术前化学治疗并术后放射治疗，但在术后十个月因多处转移死亡。

关键词：尤文氏肿瘤，磁振造影，软组织肿瘤