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Radiological Features of Extra Osseous Ewing Sarcoma: A Case Report

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Abstract **Case Report**

Ewing's sarcoma (ES) primarily affects bones and commonly presents in adolescents and young adults. This paper reports radiological features of a rare case of extra osseous ewing sarcoma of the leg region in a 11 years old boy. He was treated by a multidisciplinary approach including surgery, chemotherapy and radiotherapy. He was followed up for two years and remained well.

Keywords: Ewing's sarcoma, Extra-osseous, radiological features.

INTRODUCTION

Ewing sarcoma is the second most common childhood primary bone cancer, though a substantial proportion of these tumors arise from extraskeletal sites [1].

X Ray and magnetic resonance imaging (MRI) features of Ewing's sarcoma are non-specific, and a radiological differential diagnosis should be considered.

In the management of patients with tumours, imaging techniques are useful for biopsy guidance, evaluating the possibility of resection, and tumour response to treatment[7].

CASE REPORT

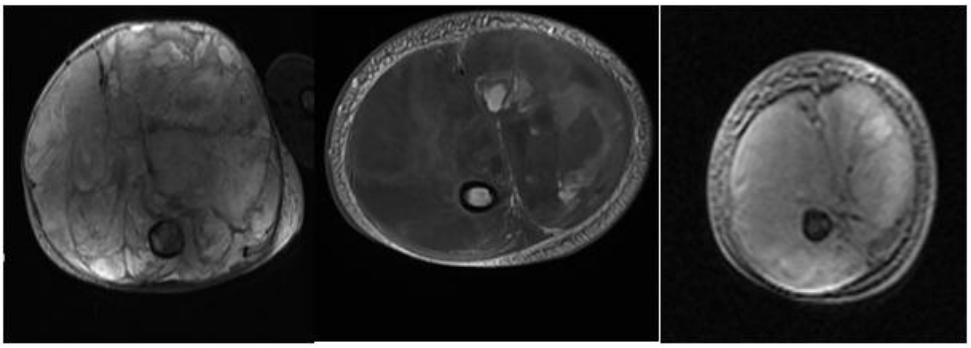
A 11 year old boy presented with history of a progressive swelling over right leg region for the last three years. There was no history of exposure to any carcinogenic agent or radiation. Past history was not significant.

The general examination of the child was normal. Local examination revealed a swelling over the right leg region, measuring 40x36x38 cm, It was mobile with well defined margins and not attached to deeper structures. Overlying skin was mobile, shiny with multiple visible vessels and a small ulcer noted in the center of the swelling. There was no neurovascular deficit distal to the tumour.

X-ray of the leg region showed soft tissue swelling without any bony involvement, MRI showed a circumferential tissue lesion process of the right leg, inhomogeneously iso- to hyper-intense on T1, Hyperintense on T2 STIR and diffusion, with heterogeneous but prominent enhancement, this process circumscribes the femoral diaphysis with cortical lysis in places.

Ultrasound guided biopsy and histopathology confirmed the diagnosis of extraskeletal ewing sarcoma in child. As part of the extension assessment, abdominal and thoracic CT reveled distant metastas in the lung and liver.

The patient was treated by multidisciplinary approach including surgery, chemotherapy and radiotherapy. He was followed up for two years and remained well.



Circumferential tissue lesion process of the right leg, inhomogeneously iso- to hyper-intense on T1, Hyperintense on T2 STIR, circumscribes the femoral diaphysis with cortical lysis in places

DISCUSSION

The Ewing's sarcoma family of tumours is an aggressive form of childhood cancer [2,3]. Extraskeletal Ewing sarcoma is rare in comparison with Ewing sarcoma of bone [4]. The prevalence of extraskeletal Ewing sarcoma is generally accepted to be between 15% and 20% of that of Ewing sarcoma of bone [5,6]. PNET has also been rarely reported in a previous site of irradiation [7]. It's affects the 10 - 20 years age group (range 4 - 25 years), involving the extremities (32%), paravertebral (15%), head and neck region (11%) and the chest in 11% of cases **[**2**,** 3].

About 25% of sufferers will present with metastatic disease, the most common sites for metastases being the lung (50%), bone (25%) and bone marrow (20%)[8]. Patients with extremity lesions usually present with a painless mass, but tumours arising elsewhere may be painful, Unusually the patient may present with a painful and rapidly enlarging mass due to spontaneous haemor- rhage. There may be no findings on conventional radio- graphs. When present, they are usually of a non-specific mass which can vary widely in size [9].

CT demonstrates a nonspecific soft-tissue mass, most commonly of similar attenuation to that of muscle (87% of cases) [10,11]. Low attenuation may also be seen, likely corresponding to areas of hemorrhage or necrosis [10,11]. Lesion margins are often poorly defined at CT (60% of cases), which is likely a reflection of the more limited contrast resolution of CT in comparison with that of MR imaging [11]. Calcification is seen in 25%–30% of cases [11]. Osseous involvement of the bone surface with cortical erosion or periosteal reaction is seen in 40% of cases [11]. However, the medullary cavity retains its normal fatty marrow attenuation, a finding reflecting lack of involvement [10,11].

MR imaging features are also nonspecific in evaluation of extraskeletal Ewing sarcoma. MR imaging demonstrates a soft-tissue mass with heterogeneous signal intensity (91%) similar to that of skeletal muscle on T1-weighted images and intermediate to high signal intensity on T2-weighted images in 100% of cases [11]. Areas of hemorrhage appear as high signal intensity on all pulse sequences and are not uncommon; fluid levels may also be evident (Fig 8). Focal areas of necrosis with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images are also frequent [11]. As in other soft-tissue masses, MR imaging is also useful for tumor staging and to evaluate the extent of involvement of surrounding structures.

The following criteria are proposed for the diagnosis of extraskeletal Ewing sarcoma: *(a)* no osseous involvement at MR imaging; *(b)* no increased uptake in bone or periosteum adjacent to the tumor on static images from bone scintigraphy; *(c)* a lesion composed histologically of small round blue tumor cells with no differentiating features at light microscopy, immunohistochemical analysis, or electron microscopy; and *(d)* demonstration of cytoplasmic glycogen [4, 6]. Meister and Gokel [9] reported three cases of extraskeletal Ewing sarcoma with the lesion located adjacent to the periosteum and with the presence of extrinsic bone erosion or periosteal reaction, but with no evidence of the tumor involving the marrow space.

Imaging modalities such as X ray, US, MRI and PET-CT help in diagnosis, however, the imaging findings are nonspecific. The diagnosis is usually confirmed by histopathological and IHC examination[12].

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). Molecular and cytogenetic analysis should be considered as the standard practice in the diagnostic evaluation of ES [13, 14].

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. The results of surgery alone for extra-osseous ES are poor in most of the cases, while patients receiving multimodal chemotherapy and radiotherapy have a much better

prognosis. With the combination of local surgical treatment and systemic chemotherapy, long-term survival has improved from 10% to 50%-60% or greater. The prognosis for extra-osseous ES appears more favourable than that of ES in bone[14, 15].

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