

# Extraskkeletal Ewings sarcoma- A case report

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## Abstract

Extraskkeletal Ewing's sarcoma is a rare tumor and has to be kept in mind in the differential diagnosis of small cell tumors of soft tissue. We present here an unusual case involving the right arm, without bony or skin involvement and treated successfully with radiotherapy and chemotherapy

## INTRODUCTION

Primary Extraskkeletal Ewing's sarcoma of soft tissue are ones in which underlying bone involvement is not found.<sup>1</sup> They are uncommon mesenchymal tumors and have to be differentiated from other small cell malignant tumors on the basis of immuno-histochemistry and electron microscopy.

We are presenting a case of this rare tumor in a young male arising in the right arm without any predisposing factor or lesion.

## CASE REPORT

A 28 year old male presented with a rapidly progressive painful swelling in right mid arm for the past six months. It was also associated with constitutional symptoms like fever, malaise and fatigue. There was no exposure to any carcinogenic agent or radiations. Past history was not contributory.

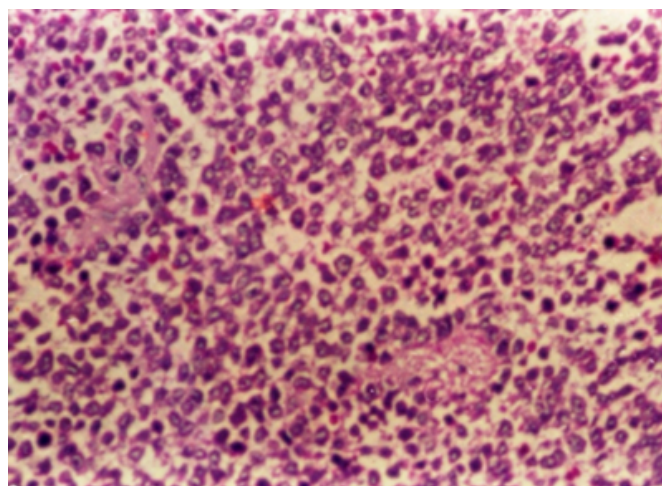
The patient was of thin built with mild pallor. No evidence of bony metastases to skull, ribs, vertebrae or lungs in the form of chest pain or haemoptysis was present. Local examination revealed a globular tender swelling involving the antero-lateral aspect of right arm, measuring 5 x 3 cms, having firm consistency with a variegated feel. It was mobile and was not attached to overlying skin or deeper structures like bone and had well defined margins. Skin was shiny with slightly raised local temperature. There was no neurovascular deficit distal to the tumor.

Haemogram with ESR revealed mild anemia and leukocytosis. Other blood tests including renal function test, liver function test, serum calcium and serum alkaline phosphatase were within normal limits. X-ray of the arm

showed soft tissue swelling without any bony involvement. Fine needle aspiration was done twice but revealed only blood. Excisional biopsy was performed and sent for histopathological examination. On microscopic examination, uniform round to oval cells having vesicular nuclei and scanty cytoplasm arranged in small sheets were seen (Fig.1), suggestive of a round cell tumor. Tumor cells were positive with Periodic Acid Schiff (PAS) stain. Immunoperoxidase staining with neuron specific enolase (NSE) showed cytoplasmic positivity within tumor cells (Fig.2), thereby confirming the diagnosis of Ewing's sarcoma.

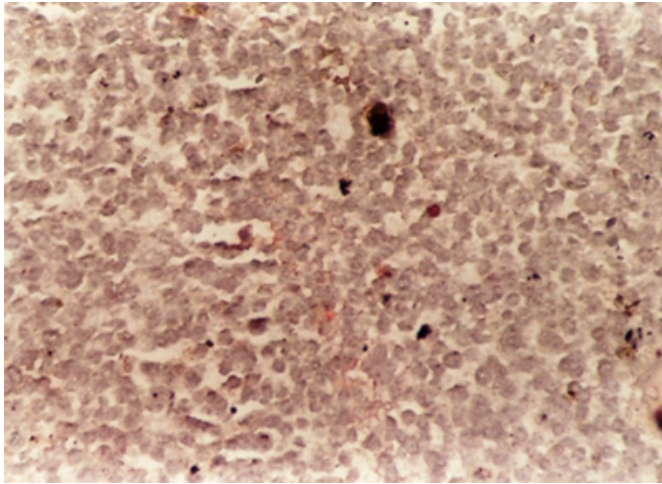
## Figure 1

Fig.1: Extraskkeletal Ewing's sarcoma: small round tumor cells with interspersed thin walled blood vessels (H & E x 250)



**Figure 2**

Fig.2: Tumor cells showing positivity with neuron specific enolase (NSE x 250).



The patient responded well to chemotherapy and local radiotherapy. Chemotherapy was given in the form of VAC regimen (Vincristine, adriamycin and cyclophosphamide), repeated every 3 weeks for 6 cycles. This was followed by external beam radiotherapy (40-50 Gy). The patient has responded well to the treatment and has not shown any recurrence after four years of follow up.

**DISCUSSION**

In 1921, Ewings<sup>1</sup> described a tumor which was referred to as diffuse endothelioma of bone.<sup>2</sup> Subsequently, it was called Ewing's tumor, and it comprises 6% of the total malignant bone tumors.<sup>3</sup> Extraskelatal Ewing's sarcoma are even rarer, only few cases have been described in literature. Similar to our case, majority occur between 10-30 years with a male preponderance. The common site being soft tissue of lower extremity and paravertebral region.<sup>4</sup> It has also been reported in epigatrium.<sup>5</sup>

The differential diagnosis includes various small cell tumors, including alveolar and embryonal rhabdomyosarcoma, peripheral neuroepithelioma, malignant lymphoma,

metastatic small cell carcinoma.<sup>6</sup> Hence, the characteristic neuron specific enolase (NSE) positivity by immuno-histochemistry confirms the diagnosis. Ultrastructurally, cells are primitive with abundant cytoplasmic glycogen, poorly developed cell junctions and no evidence of neural differentiation.<sup>7</sup>

The course of the tumor is aggressive and distant metastases are common particularly to lungs and skeleton. Our case did not show any metastasis. Lozance et al,<sup>8</sup> presented a case of rare severe hypercalcemia and acute renal failure secondary to Ewing's sarcoma.

Local treatment with radiotherapy or surgery yields the same survival rates.<sup>9</sup> We treated the patient with chemotherapy and local radiotherapy, and obtained good results.

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