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# RETROPERITONEAL EWING SARCOMA: A CASE REPORT

**Radio-Diagnosis**

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## ABSTRACT

Ewing sarcoma is a rare and extremely aggressive tumor. It commonly presents as a primary bone tumor but can also very rarely present as soft tissue sarcoma. These sarcomas are termed as Extraskeletal Ewing sarcoma (EES) and can occur in the paravertebral area, chest wall, head & neck and retroperitoneum. We report a case of retroperitoneal Ewing sarcoma in a 19-year-old male patient who was referred to our department for radiological workup.

## KEYWORDS

Ewing Sarcoma, Extraskeletal Ewing Sarcoma, Retroperitoneal Mass

## INTRODUCTION

Ewing sarcoma is a highly aggressive tumor commonly presenting as a primary bone tumor, but very rarely as soft tissue sarcoma [7]. They can be classified as skeletal Ewing sarcoma (SES) or Extraskeletal Ewing sarcoma (EES) with SES being the more common entity [3]. Most cases occur in the paediatric age group and young adults with varied presentation depending on the site of involvement. Different imaging modalities are used for localizing the disease and assessing for metastasis however the definitive diagnosis for Ewing Sarcoma is by core needle or true-cut biopsy [4]. Treatment of the disease consists of a combination of surgery, chemotherapy and high dose radiotherapy.

### Case Report

A 19-year-old boy presented with intermittent non-radiating abdominal pain for 6 months, weight loss of 10 kilograms over 6 months and loss of appetite. His clinical examination as well as biochemical findings were within normal limits. On abdominal ultrasound a large heterogeneous mass was seen in the retroperitoneum on the left side indenting and displacing the left kidney. Minimal vascularity was noted within the mass on color doppler study. A contrast-enhanced computed tomography (CECT) of abdomen was performed which showed a large heterogeneously enhancing lobulated retroperitoneal mass measuring 15 x 9.7 x 12.4 cm (Figure 1A). Few non-enhancing necrotic areas were seen within the mass. There was possible infiltration of the upper pole of left kidney and the left adrenal gland was not seen separately from the mass. Superiorly, the mass was seen indenting the splenic parenchyma and causing elevation of the left hemidiaphragm. Magnetic resonance imaging (MRI) of abdomen was performed subsequently and showed a T1 hypointense solid-cystic mass in the retroperitoneum. On T2 weighted images the mass showed mixed signal intensity while on diffusion weighted images, the mass showed strong diffusion restriction (bright on DWI and dark on ADC) (Figure 1B, 2A and 2B).

The mass was seen to encase the bifurcation of the left renal artery as well as the left renal vein. There was loss of fat plane between the mass and the left kidney. Multiple enhancing nodular deposits were noted in the retroperitoneum with nodular thickening of the Gerota's fascia. Multiple enlarged retroperitoneal lymph nodes with central necrosis were noted. Abnormal T1 hypointense signal was noted in D7, D8, D9, D10, L1 and L2 vertebral bodies, appearing hyperintense on T2 and STIR sequences suggestive of metastasis. Biopsy of the mass showed malignant round cell tumor. Immunohistochemistry revealed positivity for CD99, PAS, NSE, FLI-1 and a high Ki-67 index with diagnosis of Primitive neuroectodermal tumor/ Ewing sarcoma.

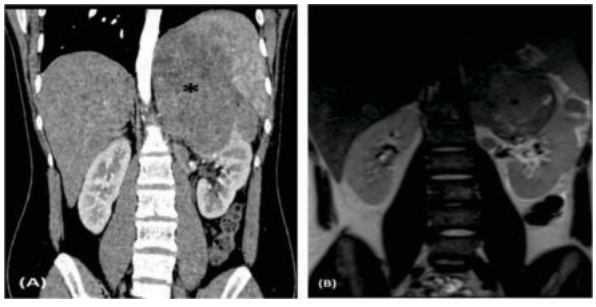


**Figure 1:** Coronal CT (A) and T2 weighted MRI (B) images shows a large lobulated retroperitoneal mass on the left side (asterisk)

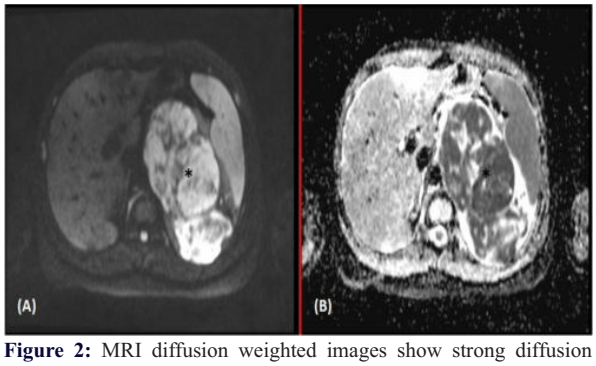


**Figure 2:** MRI diffusion weighted images show strong diffusion restriction within the retroperitoneal mass (asterisk) appearing bright on DWI (A) and dark on ADC (B)

## DISCUSSION

Ewing sarcoma is a rare malignant tumor composed of small round cells which was first described by Ewing in 1921 [1]. They can be classified as skeletal Ewing sarcoma (SES) or Extraskeletal Ewing sarcoma(EES). Ewing sarcoma has an incidence of 1 case per million per year with a lower incidence for EES [3]. EES can occur in the paravertebral region, chest, extremities, and retroperitoneum [2]. Most cases occur in the paediatric age group and young adults, presenting as local masses, localized regional pain, increased skin temperature, and restricted limb movement due to nerve invasion. Systemic symptoms such as malaise, weakness, fever, anemia, and weight loss may occur in metastatic disease. Retroperitoneal Ewing sarcomas usually present as

large masses at the time of diagnosis. They produce symptoms when they grow large enough to compress or invade contagious structures presenting with early satiety, Abdominal discomfort, distension, vomiting and other Gastrointestinal obstructive symptoms [7].

EES often spares the regional lymph nodes, but it has higher rate of distant metastasis especially to lungs and bones. Retroperitoneal Ewing Sarcoma is a rarer form of the disease and more diagnostically challenging. Different imaging modalities namely CT, MRI and positron emission tomography (PET) scan are used for localizing the disease and assessing for metastasis however the definitive diagnosis for Ewing Sarcoma is by core needle or true-cut biopsy [4]. Contrast-enhanced computed tomography (CECT) is often the preferred tool for detecting the primary retroperitoneal lesion while MRI is preferred for delineating the tumor's extent and relationship with adjacent tissues or blood supply [5]. However, while imaging studies are necessary, Tissue confirmation of the tumor is always required. Specific stains are helpful for diagnosis including those for the CD99 molecule (Xg blood group), micrometastases, vimentin, nonspecific esterase, S100 calcium-binding proteins, desmin, and cytokeratins [6].

EES is an aggressive tumor with high incidence of local recurrence and distant metastasis leading to a poor prognosis. Multimodality treatment consisting of adequate surgical resection, aggressive chemotherapy (Vincristine, Doxorubicin, Cyclophosphamide and Actinomycin - D alternating with Ifosfamide and Etoposide) and high-dose radiotherapy is recommended with better survival rate proven after combined multidrug chemotherapy [7].

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