

# Extraskelatal Ewing's sarcoma

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### ABSTRACT

نستعرض في هذا التقرير حالتين نادرتين من الأورام العظمية الساركومية الخبيثة خارج الجهاز الحركي. الحالة الأولى لرجل في متوسط العمر ظهرت في النسيج الخلفي للصدر، أما الحالة الثانية فهي لشاب صغير في العمر ظهرت في الكلية اليسرى. أظهرت الأشعة المقطعية (CT) هذه الأورام كتكتلة كبيرة من النسيج المتغاير الخواص. تم تشخيص الحالة الأولى على أنها أورام ليفمفاوية، والحالة الثانية شخّصت على أنها ورم كلوي خبيث. وقد أثبتت نتائج فحص الأنسجة المريضة أنها أورام عظمية ساركومية خبيثة خارج الجهاز الحركي.

We report 2 rare cases of extraskelatal Ewing's sarcoma, one is arising primarily from the posterior mediastinum in a middle-aged man (patient 1), and the other one is arising from the left kidney in a young male patient (patient 2). The CT in the first case showed a large mass of heterogeneous texture, with areas of cystic changes in the right side of the posterior mediastinum, no underlying bony changes or intra-spinal extension, and this mass was diagnosed as lymphoma. The second case showed almost complete replacement of the left kidney by a mass with extension through the renal vein and inferior vena cava, and it was diagnosed as renal cell carcinoma. The histological analysis of these lesions revealed extraskelatal Ewing's sarcoma

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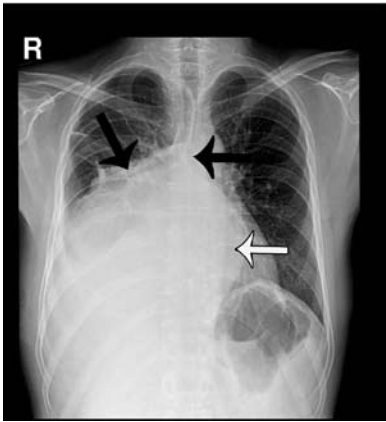
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Ewing sarcoma is the most frequent malignant bone tumor in the first 10 years of life. Although the most common place for Ewing sarcoma is the bone, it may also arise from soft tissues. Ewing sarcoma arising from soft tissue is referred to as an extraskelatal Ewing's sarcoma (EES).<sup>1</sup> The EES occurs predominantly in

adolescents and young adults between the age of 10 and 30 years, and follows an aggressive course, with a high recurrence rate. We describe the computed tomography (CT) imaging findings of 2 cases of EES located in the posterior mediastinum and left kidney. We present these cases for early awareness of ESS, and to include it in the differential diagnosis of renal and posterior mediastinal masses.

**Case Reports. Patient 1.** A 46-year-old male presented with shortness of breath for 5 months. Plain x-ray of the chest was carried out and showed a large opacity overlying the lower part of the right mediastinal shadow (Figure 1) with extension to the left paraspinal region and marked splaying of the carina, it was posterior in location in the lateral view (Figure 2). The CT showed a huge mass located in the right side of the posterior mediastinum (Figure 3) extending from the level of the carina (D5) downward to the lower chest (D12), markedly compressing the carina, right main pulmonary artery, and left atrium (Figure 4) surrounding and encasing the lower part of the esophagus down to the esophagogastric junction, associated with passive collapse of the adjacent part of the right lung with mild right side pleural effusion. It exhibited heterogeneous texture with central irregular area of cystic degeneration, no underlying bone changes or intraspinal extension. It was reported as lymphoma. Thoracotomy and open biopsy was carried out and the pathological examination showed sheets of anaplastic small round blood cells with irregular nuclear membrane and occasional nucleoli with scant cytoplasm, immunohistochemically, the tumor cells showed strong diffuse positive staining with CD-99 and focal positive staining for vimentin, synaptophysin, CD56, and Bc12 features consistent with EES, and negative for all lymphoma markers including CD34, CD45, smooth muscle actin (SMA), and desmin. The patient received chemotherapy with remarkable regression in size (Figure 5) and mass effect (Figure 6) of the mass noted in the follow up study after 4 months.

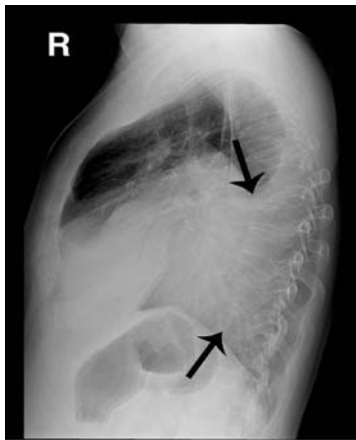
**Patient 2.** A 25-year-old male patient presented 7 months ago with easy fatigability, left flank pain, and hematuria. He had no major medical history.



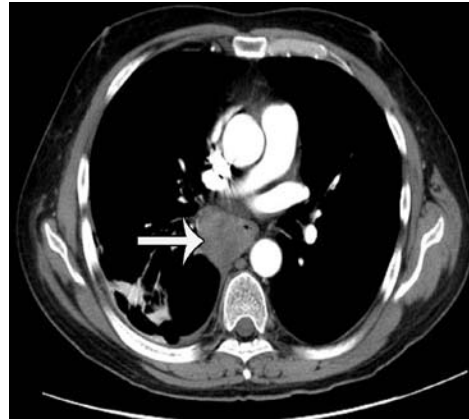
**Figure 1** - Plain x-ray of the chest posteroanterior showed a large opacity overlying the lower part of the right mediastinal shadow with extension to the left paraspinal region (white arrow) and markedly splaying of the carina (black arrow).



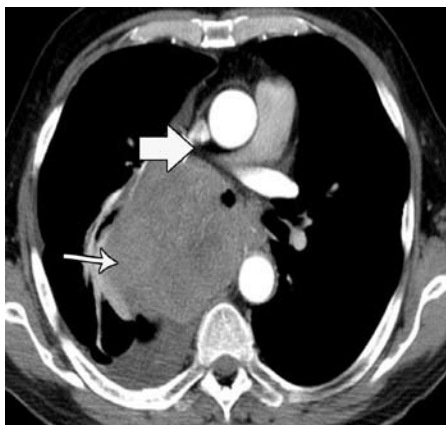
**Figure 4** - The mass is markedly compressing the carina (thin white arrow) and left atrium (thin black arrow) associated with passive collapse (thick white arrow) of the adjacent part of the right lung. It exhibits heterogeneous texture with central irregular hypodense areas of necrosis, (thick black arrow) no underlying bone changes or intraspinal extension.



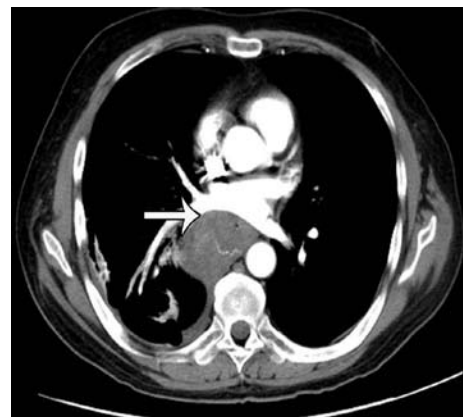
**Figure 2** - The mass is posterior in location in the lateral view (black arrows).



**Figure 5** - The CT scan after chemotherapy showed remarkable regression in size (white arrow).

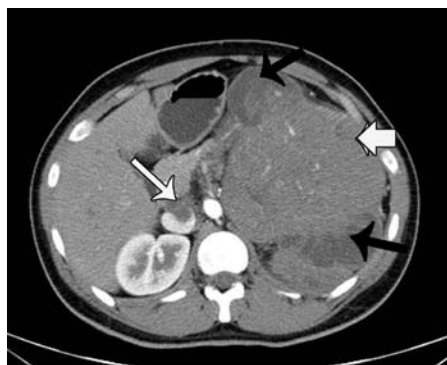


**Figure 3** - Post contrast axial CT scan of the chest mediastinal window showed a huge mass located in the right side of the posterior mediastinum (thin white arrow) markedly compressing the right main pulmonary artery (thick white arrow).



**Figure 5** - The CT scan after chemotherapy showed regression of mass effect on the left atrium (white arrow).

Physical examination revealed left flank swelling, and CT showed that the left kidney was almost replaced by a huge mass (Figure 7) sparing only its lower pole (Figure 8). It is of mixed solid and cystic components, no areas of hemorrhage, and it displaces the pancreas and spleen upward and forward with no infiltration. Also it displaces the bowel to the right side of the abdomen, the tumor extended to the left renal vein and adjacent infrahepatic portion of the inferior vena cava (IVC), infiltrating the left psoas muscle (Figure 8), associated with increased density of the surrounding fat planes with thickening of the Grotz's fascia, no regional enlarged lymph nodes, and the ipsilateral left adrenal gland could not be appreciated. The case was diagnosed as renal cell carcinoma, and the patient underwent radical nephrectomy with resection of the IVC thrombus. It was proved histopathologically to be EES of the left kidney. At 5-month follow-up, there was no clinical and radiological evidence of recurrent disease.



**Figure 7** - Post contrast axial CT scan of the abdomen showed a huge mass replacing the left kidney (thick white arrow). It is mainly solid with hypodense areas of necrosis (black arrows), no areas of hemorrhage, the tumor extends to the adjacent infrahepatic portion of the IVC (thin white arrow).



**Figure 8** - The mass sparing the lower pole of the left kidney (thick white arrow), small hypodense areas of necrosis are noted (black arrows), the mass infiltrates the left psoas muscle (thin white arrow).

**Discussion.** The EES is simply Ewing sarcoma arising in soft tissues, which are now regarded as a member of the family of small, round cell neoplasms of bone and soft tissue, including primitive neuroectodermal tumor and neuroblastoma.<sup>1</sup> The mean age is 19.1 years (range 4-47) at presentation, with 50% of patients being between 10-20 years old. There is a predominance in occurrence in male patients, and the male-female ratio is 1.5:1.<sup>2</sup> The EES is an uncommon disease that predominantly involves the soft tissues of the extremities (especially the lower extremities) trunk, paravertebral, intercostal regions, head and neck, pelvis, and peritoneum.<sup>3</sup> Other rare locations of EES have been reported.<sup>4</sup> Involvement of the left kidney is an unusual and rare presentation. Clinical and imaging findings are non-specific, and the diagnosis is based on histology.<sup>5</sup> The reported CT findings of EES are most commonly a mass of heterogeneous density and enhancement. Occasionally a central, non enhancing, low density area is seen within the mass, representing an area of necrosis.<sup>6</sup> The CT enabled us to detect the size of the mass, the integrity of the adjacent bones, and to determine the presence or absence of metastases, and was useful for evaluation of the tumor response to treatment.<sup>7</sup>

The EES shares histopathologic and immunohistochemical findings with Ewing's sarcoma and thus may be confused with other small round blue cell tumors, including embryonal rhabdomyosarcoma and lymphoma. Therefore, confirmation of the diagnosis of EES should rely on positive staining for CD99 on immunohistochemical analysis.<sup>8</sup> The treatment of EES is aggressive as most patients with Ewing sarcoma die within 2 or 3 years from diagnosis. The most effective treatment is surgical, with a combination of chemotherapy and high dose radiation therapy.<sup>9</sup>

In conclusion, the EES is a rare group of malignant tumors carrying a poor prognosis, but the literature shows the prognosis of EES is difficult to determine as most of these are case reports with only limited follow up. Early awareness and aggressive management with both surgery and chemotherapy may allow long survival term. Although EES is quite rare, it should be included in the differential diagnosis of renal and posterior mediastinal masses.

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