

Radiological Quiz – Musculoskeletal

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A 16-year-old girl presented with history of pain abdomen, loss of weight and a mass in the abdomen causing a bulge in the right lumbar region

since 4-months. Ultrasonography and computed tomography (CT) were performed. (Figures below) .What are the possible differential diagnosis in this case?

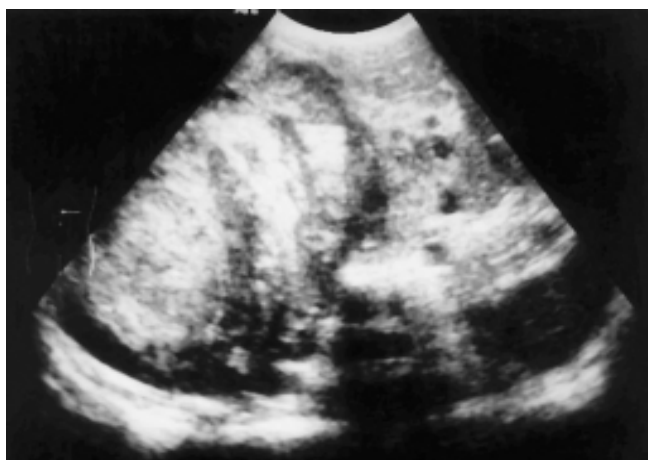


Fig. 1:-USG- transverse section mid abdomen

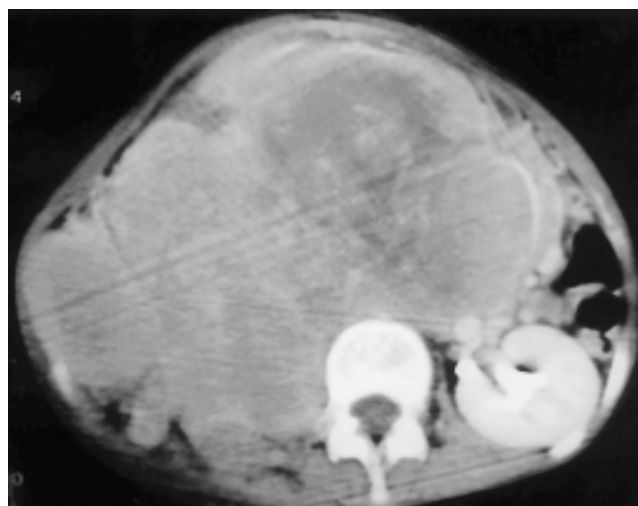


Fig.3:-Contrast enhanced CT scan abdomen

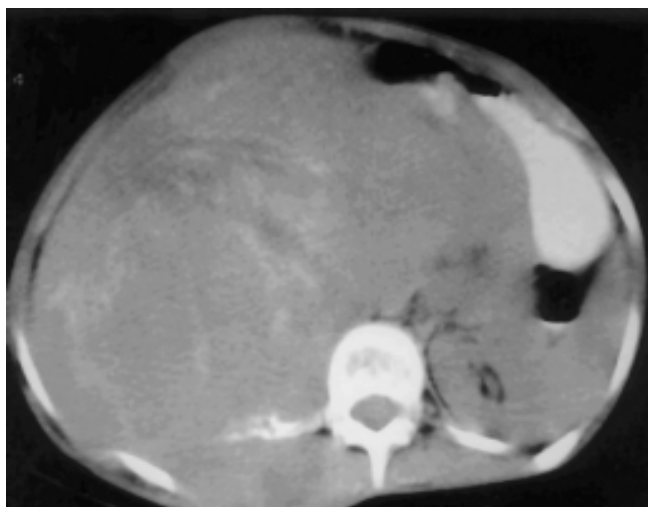


Fig .2:- Non-contrast CT scan - upper abdomen

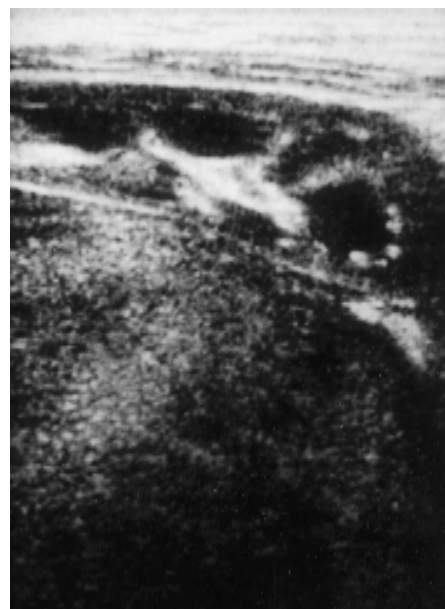


Fig.4:- USG abdomen - periumbilical region

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Radiological Diagnosis

EWING'S SARCOMA OF THE 12TH RIB PRESENTING AS RETROPERITONEAL MASS

USG abdomen revealed a solid mass in the right lumbar region. The mass was heterogeneous with multiple hyperechoic areas suggesting calcification. Pancreas was displaced anteriorly and to the left.[Fig. 1] The right kidney could not be localized in the initial ultrasound examination and hence the possibility of a retroperitoneal(renal) mass was suggested. Subsequently CT abdomen showed a large solid mass with areas of calcification and heterogeneous enhancement on the post contrast scans. Posteriorly the mass showed loss of planes with the parietal muscles; however there was no intraspinal extension of the mass. Additionally permeative lytic destruction of the right 12th rib was seen. A functioning right kidney was seen displaced anteroinferiorly stretched over the mass in the periumbilical region.[Fig.2, 3] There was no evidence of fluid or any other masses or lymph nodes in the abdomen and pelvis. The location of the right kidney was confirmed by a second ultrasound examination.[Fig.4] Chest and abdominal radiographs revealed a raised right dome of diaphragm and a moderate size right pleural effusion with lytic destruction of the right 12th rib. Hence in view of the imaging findings a diagnosis of Ewing sarcoma arising from the right 12th rib was suggested and was subsequently confirmed on cytohistopathology.

Ewing's sarcoma accounts for about 1% of all childhood cancers and is the second most common bone tumor among children and adolescents - age 3-25 years (mean age 13 years) and has a female predilection of approximately 2:1[1].

Most common sites of Ewing's sarcoma are flat bones like ribs, pelvic bones and long bones of extremities. Extra osseous Ewing's sarcoma (EES) is a rare soft tissue tumor that can occur anywhere in the soft tissues, most commonly in the extremities. Other extra-osseous sites include gut (duodenum), scalp, sino nasal region. It is usually found in older patients especially in the second and third decade of life. A case of retroperitoneal extra osseous Ewing's sarcoma (EES) with renal involvement simulating an exophytic renal mass, has been reported previously in the literature.[2]

One quarter to one third of patients with Ewing's sarcoma have metastases when they are first diagnosed. Patients with tumors closer to the trunk of the body or in the pelvic bones are more likely to have metastases than patients with tumors located on the lower leg or foot. The most common sites of spread of Ewing's sarcoma are lungs or bones; hence a chest XRay and bone scan are important in the workup of a patient with Ewing's sarcoma.

About 10% of Ewing's tumor occurs within the ribs.[3]

Ewing's sarcoma of the ribs has propensity to spread inwards towards the thoracic /abdominal cavity and may thus manifest as extra pleural or extra peritoneal masses. Ewing sarcoma involving the posterior ribs can spread into the spinal canal via the intervertebral foramina [3]. MR imaging can precisely evaluate the intraspinal and intramuscular extent of the tumor and this advantage is valuable when local therapy in the form of surgical resection of the mass is required besides chemotherapy and radiotherapy.[1]

Radiographic appearance of Ewing's sarcoma arising from the ribs is variable. The affected rib is predominantly lytic in most (82%) cases, but mixed lytic-sclerotic (9%) and even predominantly sclerotic (9%) patterns are also encountered.[4]

Ewing sarcoma is the most common malignant chest wall tumor in childhood.

Chondrosarcomas and osteosarcomas of the ribs are other common primary malignancies of ribs but are not common in this age group [5]. However, metastatic lesions to the ribs in cases of neuroblastoma and leukemia are common in pediatric patients and should be considered in the differential diagnosis.[6] Other conditions such as osteomyelitis, unusually appearing fracture, callus and direct spread of lung infection into chest wall can simulate the appearance of Ewing's sarcoma.[4]

To conclude, Ewing's sarcoma arising from the ribs frequently present as an extrapleural mass, their presentation as a retroperitoneal mass has been infrequently reported and needs to be differentiated from other retroperitoneal masses including extra-osseous Ewing's sarcoma.

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