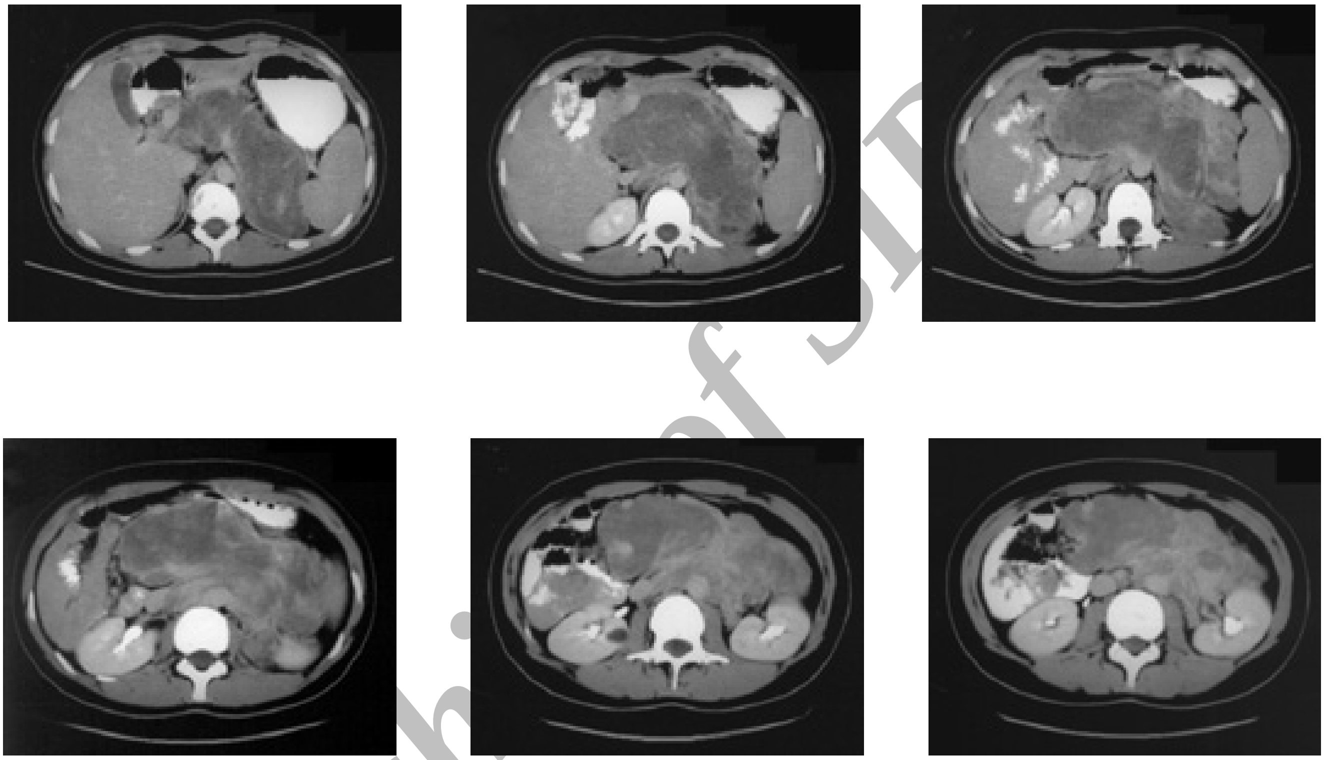
PHOTO QUIZ

What is your diagnosis?



Figs 1-6.

A 15-year-old girl was presented with epigastric pain, mild anorexia and vomiting of 2 weeks duration. There was no change of bowel habit, weight loss or icterus. The patient had no history of peptic ulcer disease, abdominal trauma, or inflammatory bowel disease.

Abdominal sonography was not of value because of the bowel gas, and the upper GI endoscopic study was normal. CT-scan imaging (Figures 1-6) and biopsy confirmed the diagnosis.

What is your diagnosis?

Diagnosis: Pararenal Liposarcoma

Prepared by: H. Saberi MD[1], M. Shaabani MD[1], H. Abootalebi MD[2], S. Markarian BSc.[2]
1. Assosiated Professor, Department of Radiology, Medical Imaging Center, Imam Khomeini Hospital, Tehran University of Medical Sciences.
2. Research Unit, Medical Imaging Center, Imam Khomeini hospital, Tehran University of Medical Sciences.

Figures 1-6 show large retroperitoneal mass, mostly low attenuated (fatty) that contains multiple strands is seen. It originated from the suprarenal soft tissue and via the anterior pararenal space; it extends to the right side. Pancreas and mesenteric vessels have also displaced anteriorly.

Primary retroperitoneal neoplasms constitute only 0.1–0.2% of all malignancies. Retroperitoneal primary tumors are mesodermal, neurogenic or lymphatic in origin, the majority of which are lymphoma, liposarcoma, leiomyosarcoma and malignant fibrous histiocytoma. Despite the overlapping radiographic features of retroperitoneal neoplasms, there are certain CT characteristics, pattern of spread and different demographics and tumor prevalence that can help suggest the tumor type.[1] CT is the modality to diagnose and assess the size and extent of such tumors; and the involvement of other organs and vasculature should be assessed when resection is in mind.[1] Liposarcoma is the most common primary retroperitoneal malignancy.[2]

Liposarcoma is a malignant tumor arising in adipocytes in deep soft tissue, such as that inside the thigh or in the retroperitoneum. Presenting typically as large bulky tumors, they tend to have multiple smaller satellites extending beyond the main confines of the tumor. Patients usually note a mass deeply seated in their soft tissue. Symptoms of pain or functional disturbances usually do not occur till the tumor is very large.[3] The presenting picture may be as signs of weight loss and emaciation and abdominal pain. When the tumors happen to compress the kidney or ureter, renal failure can ensue. Most frequent among middle-aged and older adults (age 40 and above), liposarcomas are the most common of all soft-tissue sarcomas; the annual incidence of 2.5 cases per million population. The prognosis is determined by the site of origin, the type of cancer cell, the tumor size, the depth, and proximity to lymph nodes. Metastases are common.[3] Low-grade liposarcomas have a high recurrence rate but rarely metastasize, while high-grade liposarcomas frequently metastasize to the liver, lung, and bone.[4] The 5-year survival rate for a high-grade liposarcoma is less than 50%.[3]

The radiographic appearance reflects the degree of cellular differentiation of the tumor. Lipogenic (well-differentiated) liposarcomas contain mature fat cells, whereas myxoid liposarcomas are comprised of mucin and connective tissue but relatively little fat. Pleomorphic liposarcomas are poorly differentiated and contain primarily connective elements.[2]

The CT appearance is quite varied, reflecting the degree of cellular maturity of the liposarcoma. Lipogenic sarcoma contains large amounts of fat but usually can be distinguished from benign lipomas by areas of higher density, which may represent myxoid liposarcoma. Both myxoid and pleomorphic liposarcomas contain heterogeneous areas with both mature fat and higher-density tissue. If no recognizable fat is present, a liposarcoma is indistinguishable from other soft tissue tumors.[2]

Fatty tumors can also be detected with MRI. However, lipomas may be difficult to distinguish from old hematomas, and CT remains the examination of choice for the detection and characterization of fatty tumors.[2]

CT or MRI can suggent a diagnosis of liposarcoma when fat is detected within a retroperitoneal mass. Fat in liposarcomas can often be detected because it demonstrates characteristically low attenuation on CT scans (measuring -10 HU or less). Fat can also be identified by its high T1- weighted and high T2-weighted signal intensity on spin-echo MRI images and by signal loss in fat-saturated, T1- weighted, spin-echo MRI images.[5]

Definitive differentiation of rertoperitoneal liposarcomas, (particularly of the lipogenic type) from benign lipomas is not possible with CT or MRI, although some CT or MRI findings suggest that a fatty mass is more likely to be malignant than benign. These findings include identification of soft tissue septa more than 2mm thick within a fatty retroperitoneal mss, septal irregularity or bulging, and obvious enhancement.[5]

Differentiation of liposarcoma from lipoma may be difficult even for the pathologist. Percutaneous biopsy and evaluation of the resected tumor may not resolve the issue. For this reason, many surgeons and oncologists rely on clinical findings to determine whether a fatty tumor should be considered benign or malignant. Since malignant fatty tumors of the retro peritoneum–in contrast to those in the extremities are much more common than benign fatty tumors, a fatty retroperitoneal tumor should generally be considered a liposarcoma and should be resected.[5]

It may be difficult to differentiate between an extrarenal angiomyolipoma and a liposarcoma of the retroperitoneum, especially when the tumor is large. In the early stage, liposarcomas are most often confined outside the perirenal fascia, while angiomyliopomas grow in the perirenal fat. A fat line between the mass and the kidney, compression of the renal surface and preservation of the renal cortex are suggestive for a liposarcoma, while discontinuity of the renal cortex and a spur sign are suggestive for angiomyolipoma.

References

1. Neville A, Herts BR. CT characteristic of primary retroperitoneal neoplasms. Crit Rev Comput Tomogr 2004; 45: 247-270
2. Cohan RH, Dunnick NR. The retroperitoneum in: Haaga JR, Alfidi RJ. Computed tomography of the whole body. 2nd ed, Vol. 2. Missouri: Mosby; 1988: 1091
3. Liposarcoma in: http://en.wikipedia.org/wiki/Liposarcoma 2006
4. Jorgensen M, Prokop M. Primary peritoneal tumors in: Prokop M, Galanski M. Spiral multislice computed tomography of the body. Stuttgart: Thieme; 2003: 621- 622
5. Ashok KG, Richard HC. The peritoneum in: Haaga JR. CT and MR imaging of the whole body. 4th ed. Vol. 2. Missouri: Mosby; 2003: 1695