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Case Report

Perirenal GIST: Imaging Features of Unusual Perirenal Tumor

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Abstract

Gastrointestinal stromal tumors (GIST) are most common mesenchymal tumor of gastrointestinal tract. In addition, they rarely occur at other locations, mostly in omentum and mesentery. Primary retroperitoneal GISTs are very rare. To date only 59 cases have been described, its perirenal location being very rare. We describe a case of 64 year old man presenting with abdominal lump of 6 months duration and abdominal discomfort. Computed tomography (CT) scans of abdomen revealed large right perirenal mass compressing the right kidney. Histopathological examination showed uniformly proliferating spindle cells with immunoreactivity for C-KIT confirming it to be a GIST. En bloc resection of the perirenal mass along with right kidney was done and proved to be curative. The present case was rare perirenal GIST with typical immunopathological features.

Keywords: *Gastrointestinal Stromal Tumors (GIST); Computed Tomography (CT)*

Introduction

Gastrointestinal stromal tumors (GIST) are rare tumors accounting for about 1 - 3% of all gastrointestinal malignancies. They are most common mesenchymal tumors of the gastrointestinal (GI) tract [1].

GISTs are thought to arise from the Interstitial Cells of Cajal, which regulate the gastrointestinal motility. The characteristic histological feature of GIST is kit (CD117) positivity on immunohistochemistry [2].

In the gastrointestinal tract, 40% - 70% originate from the stomach, 20% - 40% from small intestine, 5% - 15% from the colon and rectum and 5% from the esophagus [2,3].

Rarely, tumors with histological characteristics of GIST may occur outside the gastrointestinal tract like omentum, mesentery and retroperitoneum, when they are termed as extra-gastrointestinal GIST (EGIST). Retroperitoneal GISTs are extremely rare; to date only 59 cases have been reported. Perirenal location is one of the rare sites of this tumor [4].

Since these tumors commonly present with vague complaints and the location of mass is often obscure on clinical findings alone, cross sectional imaging like computed tomography (CT) scan and magnetic resonance imaging (MRI) helps to delineate the extent, location and relationship to adjacent structures.

Surgical removal with 'en bloc' resection of tumor along with contiguous tissues is the standard treatment of non-metastatic EGIST. The role of tyrosine kinase receptor inhibitor imatinib mesylate is not clear [5].

Due to the rarity of the tumor, it is imperative to know the imaging features and possible mimics to be able to keep this as a possible differential in the report.

We present the case of 64-year old man with perirenal GIST describing the CT appearance and imaging differentials.

Case Report

A 64 -year old man was admitted in our institute with the complaints of abdominal distension and dragging sensation in right side of abdomen for 4 months. There were no other complaints. Prior to admission, he underwent ultrasonography in the outpatient department, which revealed a large well defined heteroechoic mass in right lumbar region with a part of kidney visualized medially. The patient underwent a Computed Tomographic scan of the abdomen before and after administration of intravenous contrast for further evaluation. A large well-defined hypoattenuating mass about 18 cm x 14 cm x 11 cm was noted in right perirenal location compressing the right kidney and displacing it anteromedially. The mass had well defined fat planes with adjacent bowel loops. The mass showed heterogeneous mild post contrast enhancement with preserved corticomedullary differentiation of compressed right kidney. Right adrenal was not visualized separately from the mass. The bowel loops appeared displaced towards left side of the abdomen by the large right perirenal mass. The right renal vessels were well opacified by contrast with no evidence of thrombus within. No other focal lesion or enlarged lymph nodes were seen on the scan. As part of the workup, standard blood tests, electrocardiogram (ECG) and chest radiograph were done which showed no abnormality. In view of the dimensions of the mass, laparotomy was performed and 'en bloc' resection of the mass was done along with right nephrectomy, as right kidney was inseparable from the mass. Laparotomy was preferred over laparoscopy due to the large size of the lesion. No postoperative complaints were recorded and patient was discharged after six days. Postoperative pathological examination revealed a solid tumor, which appeared yellow-brown on cutting the tissue. On microscopic examination, the surgical margins were negative for tumor cells. The tumor consisted of spindle cells with mitotic activity of < 5 per high power field (low mitotic activity). Immunohistochemistry showed positivity for kit (CD 117), focal positivity for S-100 protein and NSE (neuron specific enolase), negativity for CD34, smooth and skeletal muscle actin. Morphological and immunohistochemical features suggested a diagnosis of gastrointestinal stromal tumor. Based on the dimensions and mitotic activity, a low risk profile was indicated according to the risk assessment proposed by Yamamoto., *et al.* [6] and intermediate risk form according to Fletcher., *et al* [7]. As complete resection of the tumor was achieved, adjuvant therapy with imatinib mesylate was not considered necessary.

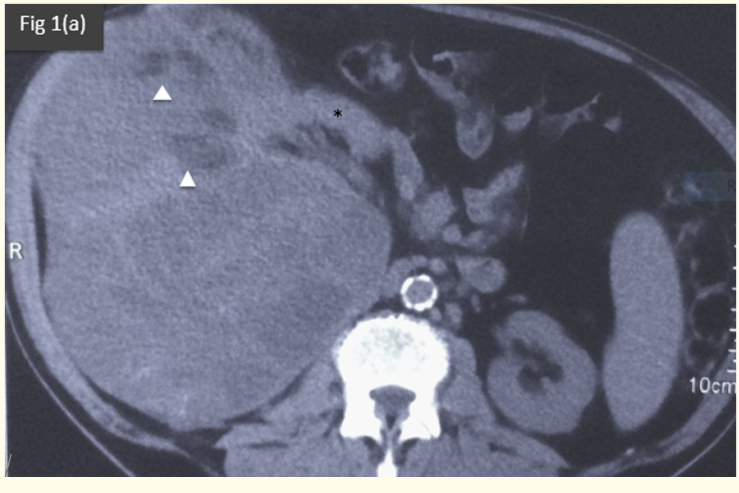

Figure 1a: *Non contrast computed tomography scan of abdomen showing a large perirenal mass with hypodense areas (white arrowheads) suggestive of necrosis, surrounding and compressing the right kidney (*).*

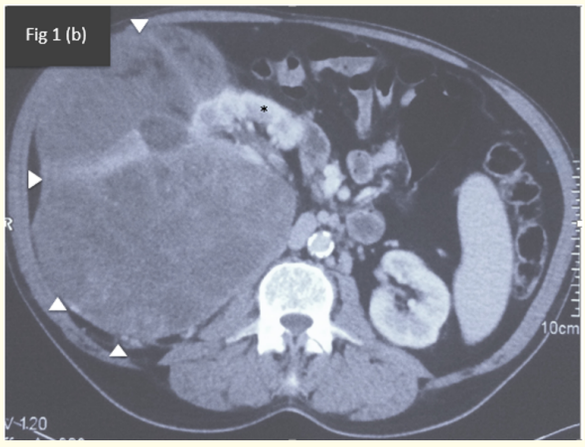

Figure 1b: *Contrast enhanced computed tomography scan showing heterogeneous mild post contrast enhancement of the well-defined perirenal mass (white arrowheads) surrounding and compressing the right kidney (*).*

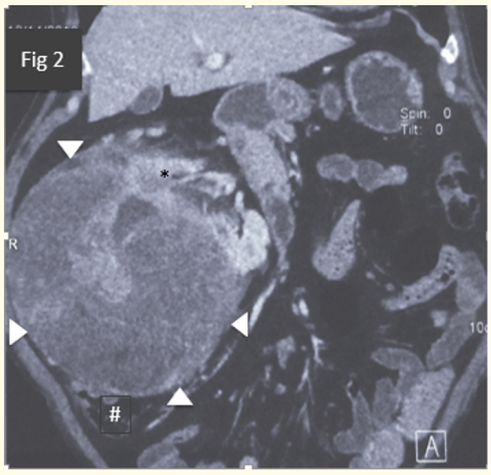

Figure 2: *Coronal multiplanar reformat (MPR) showing the well-defined perirenal mass (white arrowhead) and its relationship to right kidney (*). Fat planes appear preserved with adjacent bowel loops (#).*

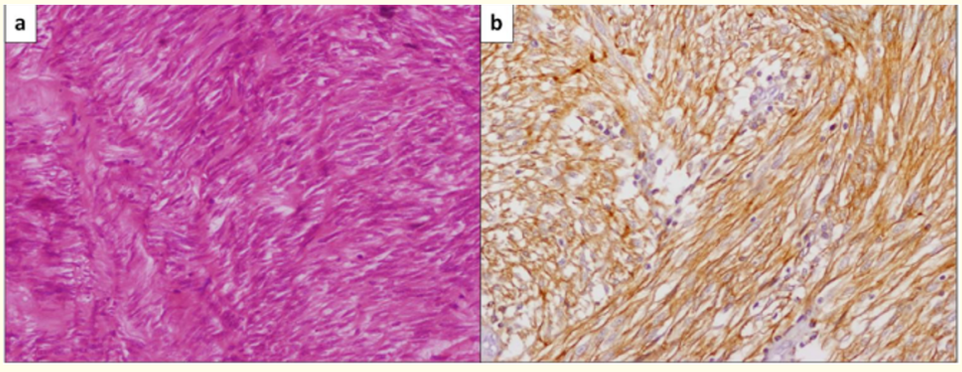

Figure 3: *Photomicrographs showing histomorphology and immunohistochemistry features of c-kit positive spindle cell GIST A: Interlacing fascicles of tumor spindle cells (HEx200), B: c-kit showing membranous (4+) positivity (x200).*

Discussion

GIST comprises the most common mesenchymal neoplasm of the GI tract after epithelial and lymphoid neoplasms. They can occur in any part of the tubular GI tract but are most common in stomach and proximal small intestine [1]. Occasionally, they occur outside the GI tract when they are called EGIST. Such phenotypically identical lesions can occur in omentum, mesentery and retroperitoneum. GIST is believed to originate from the pacemaker cells of the intestine or interstitial cells of Cajal. Like their cell of origin, they stain strongly positive for c-kit or CD117, which is used to distinguish them from other non-GIST spindle cell tumors like leiomyomas and schwannomas [2].

The pathogenesis of GIST has been extensively researched in recent decade. However, the origin of EGIST remains obscure. Due to CD34 positivity of both GIST and EGIST, both are believed to have common precursor cells [8-10]. CD34 is a transmembrane glycoprotein expressed in hematopoietic progenitor cells, dendritic cells of skin appendages and nerve fibres as well as vascular endothelial cells and hence found in their neoplastic proliferation [8,10,11].

GISTs express a tyrosine kinase membrane receptor, kit. Due to exon mutations, activation of kit occurs resulting in unchecked cell growth and inhibition of apoptosis.[1] In addition, mutations of platelet derived growth factor receptor alpha (PDGFRA) have also been found relevant to tumorigenesis of some GIST [6]. Kit expressing tumors similar to GIST have been reported in omentum and mesentery by Miettinen., *et al* [9]. Yamamoto., *et al.* have recently described kit expression and mutation and sporadic PDGFRA mutations in retroperitoneal GIST [6].

The diagnosis of GIST is often suspected histopathologically when one of the following three patterns is seen: spindle cell type (70%), epithelioid type (20%) or mixed type. For confirmation, kit immunostaining is done. The clinical behavior of GIST can be predicted by the tumor size and mitotic activity. Small (< 2 cm) tumors with low mitotic activity (< 5 mitoses per high power field) have excellent prognosis [12].

The CT and MR appearances of GIST have been described in radiological literature. GISTs vary from well-defined small homogeneous mass to large heterogeneous ill-defined mass with necrosis and calcification being seen in large masses. Central fluid attenuation is seen in 67% of cases [13]. Crescent- shaped necrosis has been described as a sign of large GIST, also known as Torricelli-Bernoulli sign [1]. The CT features predictive of poor outcome are presence of hepatic metastases, wall invasion and lesions larger than 11.1 cm as reported by Tatesihi., *et al* [14]. The signal intensity appearance pattern on MRI depends on the presence of necrosis or hemorrhage [15]. The solid portion of tumor is low signal intensity on T1-weighted images, high signal intensity on T2-weighted images and shows post contrast enhancement. Hemorrhage within the tumor varies in signal intensity from low to high on T1 and T2 weighted images depending on the age of the hemorrhage.

Retroperitoneal GIST especially perirenal location is very uncommon and its imaging has not been well described in the literature. In our case, the perirenal GIST appeared as a large well defined heterogeneously enhancing mass with areas of necrosis but no calcification. These findings are similar to large GISTs of the GI tract. Distinction from other retroperitoneal sarcomas like leiomyosarcoma, liposarcoma, malignant fibrous histiocytoma and fibrosarcoma based on clinical and imaging findings alone is difficult [12]. However, a differential diagnosis of GIST should be considered when retroperitoneal tumor with such imaging findings is seen.

Conclusion

Primary retroperitoneal GISTs are rare tumors. The radiologist should be aware of the imaging characteristics and diagnosis should be considered even though the imaging features alone may not distinguish them from other retroperitoneal sarcomas.

Bibliography

1. Sandrasegaran K., *et al*. "Gastrointestinal stromal tumors: CT and MR I findings". *European Radiology* 15.7 (2005): 1407-1414.

2. Miettinen M., *et al*. "Pathology and diagnostic criteria of gastrointestinal stromal tumors (GISTs): a review". *European Journal of Cancer* 38.5 (2002): S39-S51.

3. Miettinen M., *et al*. "Gastrointestinal stromal tumors". *Annales Chirurgiae et Gynaecologiae* 87.4 (1998): 278-281.

4. Casella C., *et al*. "Primary Extra-gastrointestinal Stromal Tumor of Retroperitoneum". *Clinical Medicine Insights: Oncology* 6 (2012): 189-197.

5. Logrono R., *et al*. "Recent advances in cell biology, diagnosis and therapy of gastrointestinal stromal tumor (GIST)". *Cancer Biology and Therapy* 3.3 (2004): 251-258.

6. Yamamoto H., *et al*. "C-kit and PDGFRA mutations in extragastrointestinal stromal tumor (gastrointestinal stromal tumor of the soft tissue)". *American Journal of Surgical Pathology* 28.4 (2004): 479-488.

7. Fletcher CDM., *et al*. "Diagnosis of gastrointestinal stromal tumors: a consensus approach". *Human Pathology* 33.5 (2002): 459-465.

8. Miettienen M., *et al*. "Gastrointestinal stromal tumors/smooth muscle tumors (GISTs) primary in the omentum and mesentery: clinicopathologic and immunohistochemical study of 26 cases". *American Journal of Surgical Pathology* 23.9 (1999): 1109-1118.

9. Reith JD., *et al*. "Extragastrointestinal (soft tissue) stromal tumors: an analysis of 48 cases with emphasis on histologic predictors of outcome". *Modern Pathology* 13.5 (2000): 577-585.

10. Takizawa I., *et al*. "Primary gastrointestinal stromal tumor in the retroperitoneum". *International Journal of Urology* 13.9 (2006): 1245-1248.

11. Suster S., *et al*. "Solitary fibrous tumors of soft tissue. A clinicopathologic and immunohistochemical study of 12 cases". *American Journal of Surgical Pathology* 19.11 (1995): 1257-1266.

12. Takao H., *et al*. "Gastrointestinal stromal tumor of the retroperitoneum: CT and MR findings". *European Radiology* 14.10 (2004): 1926-1929.

13. Ghanem N., *et al*. "Computed tomography in gastrointestinal stromal tumors". *European Radiology* 13.7 (2003): 1669-1678.

14. Tateishi U., *et al*. "Gastrointestinal stromal tumor. Correlation of computed tomography findings with tumor grade and mortality". *Journal of Computer Assisted Tomography* 27.5 (2003): 792-798.

15. Levy AD., *et al*. "Gastrointestinal stromal tumors: radiologic features with pathologic correlation". *Radiographics* 23.2 (2003): 283-304.

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