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Case Report:

A rare case of synchronous occurrence of retroperitoneal liposarcoma and gastrointestinal stromal tumor

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ABSTRACT

Background: Liposarcoma and Gastrointestinal Stromal tumors are both mesenchymal in origin. However, their coexistence is quite rare as they develop from different cell layers. Liposarcoma arises from adipose tissue and is one of the most common soft tissue sarcomas of adults. GISTs are one of the most common mesenchymal neoplasms of the gastrointestinal tract. They arise mainly from the stomach and the small bowel.

Keywords: Liposarcoma, gastrointestinal stromal tumor, mesenchymal, sarcoma

INTRODUCTION

Liposarcoma (LPS) is one of the most common soft tissue sarcomas of adults [1]. World Health Organization (WHO) has categorized LPS into 4 subtypes namely atypical lipomatous tumor /well-differentiated liposarcoma (ALT/WDLS), dedifferentiated liposarcoma, myxoid liposarcoma, and pleomorphic liposarcoma. WDLS accounts for 40-45% cases of all the liposarcomas [2]. They commonly affect deep-seated soft tissues of the thigh followed by retroperitoneum [3]. These lesions are painless and enlarge slowly [4]. Complete surgical resection is considered to be the most effective treatment [4]. The prognosis depends on the gross resection of the tumor, margin of resection, and the need for contiguous organ resection [5]. Tumors occurring in the deep anatomical location like retroperitoneum tend to recur and can even dedifferentiate and metastasize [2]. Grossly, liposarcomas are well-circumscribed and lobulated mass [3]. Microscopy shows lipoblasts with nucleus pushed to the periphery and multiple cytoplasmic vacuolations. Atypical cells with hyperchromatic nucleus along with areas of necrosis and myxoid change can also be seen [2].

Gastrointestinal stromal tumor (GIST) is the most common type of mesenchymal neoplasm of the gastrointestinal tract which is believed to develop from interstitial cells of Cajal [6]. More than 50% of these tumors arise from the stomach, followed by duodenum, colon, and rectum [7,8]. Patients usually present with vague abdominal symptoms. However one-third of the cases are found incidentally during radiological investigations, endoscopy, or surgery performed for other tumors, which holds true in our case also [9]. Complete surgical resection is the mainstay of treatment [10]. The prognosis correlates with the tumor size, location, and mitotic index [8].

Although we have seen frequent cases of synchronous occurrence of GISTs with other tumors, however GISTs with simultaneous liposarcoma are very rare and only 2 such cases have been reported in the past. They are both mesenchymal in origin however, they develop from different cell layers which makes their coexistence quite rare [11].

Here, we report a case of synchronous occurrence of retroperitoneal liposarcoma and GIST in a 66-year old male who presented with burning sensation over the upper abdomen for 3 months.

CASE HISTORY

A 66-year-old male presented with burning sensation over the upper abdomen of 3 months duration. It was insidious in onset, gradually progressive, increased on bending forwards, and decreased on lying down. There was no history of vomiting, fever, abdominal pain, bladder and bowel disturbances and weight loss. On palpation, an ill-defined mass was felt in the epigastric, umbilical, and left lumbar region measuring approximately 20cm x 15cm. The mass was firm in consistency, fixed, and did not move with respiration. However, the size was reduced on head lifting and leg lifting. The USG showed a heterogeneous mass lesion in the epigastric, umbilical, and left lumbar region with increased vascularity and areas of calcification within the lesion. Left iliac fossa also showed two heterogenous lesions. The CECT (Contrast enhanced computed tomography) of the abdomen and pelvis showed a large multilobulated heterogeneously enhancing solid-cystic retroperitoneal mass lesion with fatty attenuation measuring 15cm x 10.9cm x 1.7cm. (Fig 1) The lesion was encasing the perirenal fat of the left kidney and was causing the displacement of adjacent structures. The diagnosis of primary retroperitoneal neoplasm with the possibility of liposarcoma was made. Exploratory laparotomy with tumor excision and left nephrectomy was done. However, the surgeons also found a small lesion in the stomach measuring approximately 1.5cmx1.5cmx1cm. An excisional biopsy from the stomach lesion was done. On gross examination, the soft tissue mass showed lobulations with areas of hemorrhage and dilated blood vessels and measured 26cmx24cmx10cm and weighed 2.8 kg. The left kidney was completely displaced by the mass and parenchyma appeared normal. The cut surface of the mass showed multiple, lobulated gelatinous areas admixed with pale yellow homogenous areas. (Fig 2) Areas of myxoid change with calcifications were seen. The excisional biopsy specimen from the stomach appeared pale white to pale yellow and measured 1.5x1.5x1cm. Cut surface showed a well-circumscribed lesion which was myxoid in appearance and contained few grey white foci. Histologically, soft tissue mass showed sheets of mature adipocytes of varying sizes admixed with fibro-collagenous stroma showing spindle cells and bizarre tumor giant cells. Few lipoblasts showed multi-vacuolations. (Fig 3) Focal areas of hemorrhage and necrosis admixed with neutrophilic aggregates were seen. The lower pole of the kidney showed inflammatory cell infiltrates and foci of autolysis. There was no tumor infiltration into the kidney. Diagnosis of Well-differentiated Liposarcoma – Grade I was made. Pathological staging was pT4N0. Sections from the excisional stomach biopsy showed spindle cells with elongated nucleus arranged in fascicles and surrounded by muscle tissue. (Fig 4) Focal areas of calcification and hyalinization were also seen. Some areas showed a palisaded pattern with vacuolated cells. Mitosis was < 5/50 HPF. Diagnosis of Gastrointestinal Stromal tumor – low grade was made. He has received six cycles of chemotherapy and one cycle of radical radiotherapy on subsequent follow up and is doing well.

DISCUSSION

GISTs account for 2.2% of all malignant gastric tumors [8]. They are one of the most common mesenchymal neoplasms of the gastrointestinal tract, predominantly arising from stomach followed by the small intestine, colorectum, and esophagus [7,8]. Males and females are equally affected with a peak incidence in the 6th decade of life. However, some rare cases have also been depicted in a younger population [7]. These tumors arise from the interstitial cells of Cajal, also known as pacemaker cells of the gastrointestinal muscularis propria [9]. 90% of GISTs have oncogenic, gain-of-function mutations in the receptor tyrosine kinase KIT whereas the remainder have mutations of PDGFRA [11]. Immunohistochemically, most GISTs are positive for KIT (CD117) and CD34. Some are focally positive for smooth muscle actin, desmin, and very few for S100-protein [8]. Patients usually present with symptoms due to mass effects, alimentary tract hemorrhage, and mucosal ulcerations, however, one-third of the cases are found incidentally [9]. As in our case, based on the tumor detected during exploratory laparotomy, the gastric GIST could be classified as an incidental tumor. Grossly, size may vary from minimal mural nodules to > 30cm solitary, well-circumscribed, fleshy mass [7]. Histologically they can be either composed of thin elongated cells - spindle cell type, or epithelial appearing cells - epithelioid type. Some can even show nuclear atypia and mitotic activity [8]. The prognosis correlates with the tumor size, location, and mitotic index with gastric GISTs being less aggressive [6]. Tumors less than 5cm and mitotic counts less than 5 per 50 HPF are considered low grade whereas tumors over 5 cm and mitotic rate > 5/50 HPF are usually high grade [9]. Considering the size and the mitotic index, the GIST in our case would be graded as low grade. For all the mesenchymal tumors, surgical excision is the mainstay of treatment followed by chemotherapy/radiotherapy for a better survival rate [12].

Liposarcoma (LPS) is one of the most common soft tissue sarcomas of adults [1]. Well Differentiated Liposarcoma accounting for 40-45% cases of all the liposarcomas, is the most common subtype [2]. Usually seen in the 5th -6th decade of life, there is no sex predilection. They occur most commonly in the deep soft tissues of the thigh followed by retroperitoneum [2]. These tumors are usually missed pertaining to the fact that the retroperitoneal area provides a potential space for their growth and is only found when their size exceeds>20cm and the patient presents with some abdominal symptoms [3,4].

A radiological investigation like USG is used to screen retroperitoneal sarcomas as it can accurately determine the size, location, and relationship of the tumor with the surrounding blood vessels [4]. CT and MRI show inhomogeneous appearance with irregular enhancement [5]. In our patient, the USG showed a heterogeneous mass lesion in the epigastric, umbilical, and left lumbar region measuring 158mm x 86mm with increased vascularity and areas of calcification within the lesion. Left iliac fossa also showed two heterogeneous lesions measuring 52mmx40mm and 39mmx38mm respectively. The CECT of the abdomen and pelvis showed a large multilobulated heterogeneously enhancing solid-cystic retroperitoneal mass lesion with fatty attenuation. The lesion was extending across the midline and was causing displacement of adjacent structures. It was also encasing the perirenal fat of the left kidney.

In treating patients with retroperitoneal neoplasm, complete surgical resection is the mainstay failing which they tend to recur [4]. In this case, exploratory laparotomy with tumor excision and left nephrectomy was done.

Grossly, well differentiated liposarcomas are well circumscribed and unencapsulated. Histologically they show lipoblast, which appears as a mononuclear or multinucleated cell with multiple cytoplasmic vacuoles containing lipid. The nucleus is often pushed aside by the vacuole. But sometimes, it may remain centrally located and exhibit small sharp indentations by multiple small lipid vacuoles. On high power, scattered tumor cells with large hyperchromatic nuclei can be easily appreciated [2,3]. According to French Federation Nationale des Centres de Lutte Contre le Cancer (FNCLCC), soft tissue sarcomas can be graded depending upon the degree of differentiation, necrosis, and mitotic activity. Well differentiated liposarcoma is graded as grade 1 [2]. Retroperitoneal liposarcoma tends to recur and even dedifferentiate and metastasize and therefore in those cases, the mortality rate can be as high as 80% [1]. The important prognostic factors for survival are the completeness of the surgical excision with negative margins and the grading of the tumor. Post-operative additional radiotherapy can increase the survival rate [2]. In our case, the patient has received six cycles of chemotherapy and one cycle of radical radiotherapy on subsequent follow up and is doing well.

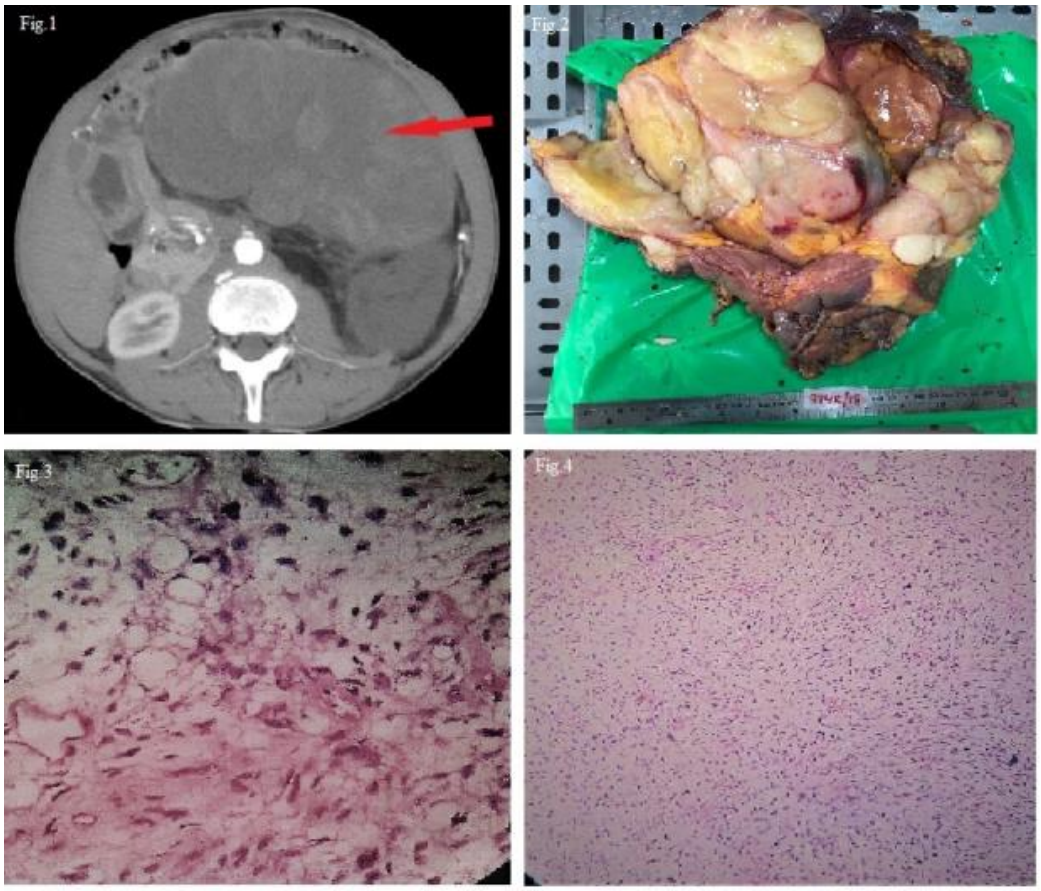

Fig 1: CECT shows a large multilobulated heterogeneously enhancing solid cystic retroperitoneal mass lesion with fatty attenuation, Fig 2: Grossly, the soft tissue mass showed multiple, lobulated pale yellow homogenous areas. Lower pole shows left nephrectomy specimen, Fig 3: Microscopic image of liposarcoma showing vacuolated lipoblasts and occasional spindle cells. (H/E, 40x), Fig 4: Microscopic image of GIST showing spindle cells in fascicles. (H/E, 10x)

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