Case Report

Giant primary synchronously bilateral mesenteric dedifferentiated liposarcoma with hyperparathyroidism, hyperthyroidism, Type-2 diabetes mellitus and hypertension

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Abstract

Liposarcomas represent the single most common type of soft tissue sarcoma, occurring most commonly in the extremities and retroperitoneum. There is no relation between liposarcomas and multiple endocrine syndromes.

We presented a 61-year old woman with giant primary synchronously bilateral mesenteric dedifferentiated liposarcoma with hyperparathyroidism, hyperthyroidism, Type-2 diabetes mellitus (T2DM) and hypertension.

The mesenteric liposarcoma was reported neither synchronously bilateral nor with endocrine disorders. We must note if the patients' presentation was a co-incidence or an undescribed syndrome, waiting to be discovered.

Introduction

Liposarcomas represent the single most common type

of soft tissue sarcoma, occurring most commonly in the extremities and retroperitoneum. Its intra-abdominal localization is rare, occurring in only 5% of cases.[1] Primary mesenteric liposarcoma is a rare malignant tumour of mesenchymal origin. These tumours are often found to be of substantial size upon first clinical presentation.

Case Report

A 61-years old female patient was admitted to our hospital with chief complaint of fatigue and abdominal distension. She had a history of hypertension and Type-2 diabetes mellitus (T2DM) which were under medical treatment.

On physical examination, large palpable masses were found in the abdomen. The white blood cell count was 9.6 x $10^3/\mu l$ with 56.1% neutrophils. Electrolytes were normal except calcium, which was 2.60 mmol/L. Other laboratory tests were normal except. Serum glucose 135 mg/dl, Blood urea nitrogen 45.2 mg/dl, ALP 395 U/L, TSH<0.01 IU/L, PTH 125 IU/L. multi-nodular goitre and a parathyroid adenoma at inferior part of right lobe was reported at ultrasonographic examination of the neck. Also, abdominal computerized tomography was reported as bilateral giant mesenteric liposarcomas (Figure). After being euthyroid with medication,

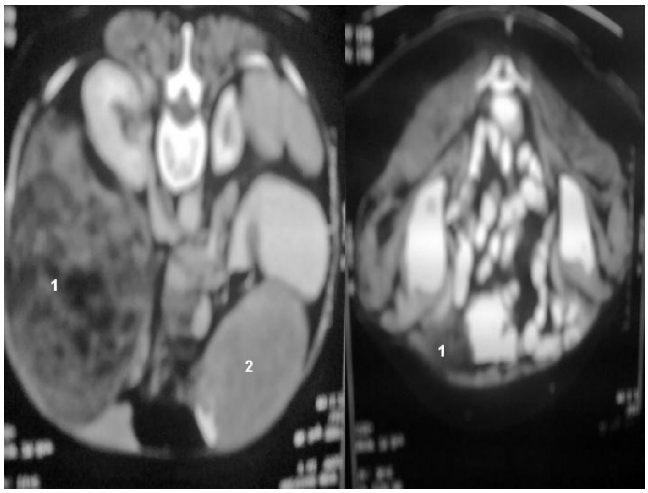

Figure: Giant primary synchronously bilateral mesenteric liposarcomas can be seen at the intravenous contrast enhanced computerized tomography. Right is larger than the left.

an abdominal surgery was planned.

On laparotomy there were 2 masses of diameters, 25 and 16 cms respectively and weighing approximately 14.5 kilograms together. Histopathology showed features of atypical lipomatous tumour/well differentiated liposarcoma (ALT/WDLS) with mixed histological pattern varying from area to area, for example lipoma like sclerosing and myxoid areas. Immunolabeling for S-100 was positive in the well-differentiated component, as would be expected of a liposarcoma. CD117 (C-KIT) and CD34 were negative in the dedifferentiated component, thus ruling out a gastrointestinal stromal tumour (GIST). Based on the histology and immunoprofile, a diagnosis of dedifferentiated liposarcoma was rendered. The resection margins of the specimen were clear.

The patient was adapted to a chemotherapy programme. Also, 3 months after the abdominal operation her parathyroidectomy and thyroidectomy were performed. The patient is living a healthy life 12 months after the surgery.

Discussion

Primary mesenteric liposarcoma is a rare malignant tumour of mesenchymal origin. Liposarcoma is divided into five subtypes according to the World Health Organization; well differentiated, dedifferentiated, myxoid, pleomorphic, and mixed type. ALI/WDLS comprises 40% to 45% of all liposarcomas, whereas myxoid and pleomorphic liposarcomas represent 35% to 40% and 5%, respectively.[2]

Both, well-differentiated and dedifferentiated liposarcomas have an equal sex predilection with the highest incidence in the 6th-7th decade of life. Also, our patient was 61-years old. Dedifferentiated liposarcomas are defined histologically by a transition from well-differentiated liposarcoma to a non-lipogenic sarcoma with variable histological grade.[2] ALT/WDLS does not metastasize, but dedifferentiated liposarcoma has a 15% to 20% metastatic rate.[3,4] Myxoid and pleomorphic liposarcoma have metastatic rates of 33% and 30% to 50%, respectively.[2] Histological patterns of metastasis also vary. Although pleomorphic and dedifferentiated liposarcomas may metastasize to unusual sites such as liver, brain, and bone. Involvement of the lungs is most common.[3,4]

Approximately, 40% of dedifferentiated liposarcomas will recur locally, 17% will metastasize and 28% of the patients will ultimately die as a result of the tumour.[2] In this patient, there were two synchronic dedifferentiated liposarcomas with no metastasis. Evans reported that the median survival of patients with the well differentiated type, the myxoid type, the dedifferentiated type and pleomorphic type was 119, 113, 59 and 24 months, respectively.[4]

The location of the lesion in our case raises the possibility of a gastrointestinal stromal tumour (GIST). GISTs usually resemble smooth muscle tumour with a variety of histological patterns, which does not consist of a well-differentiated liposarcoma component, necessary for the diagnosis of a dedifferentiated liposarcoma. GISTs typically stain with CD117 and CD34. Immunohistochemically, dedifferentiated liposarcoma is usually negative for CD117 and CD34 in the dedifferentiated component and positive for

S100 protein in the well-differentiated component. Dedifferentiated liposarcoma needs to be distinguished from other high-grade sarcomas such as malignant fibrous histiocytoma because these high-grade sarcomas have a much worse prognosis.[5]

To best of our knowledge, literature search for multiple endocrine syndromes presenting as liposarcoma was not present. However, Type 2 Diabetes Mellitus and liposarcoma may be co-incidental. Also, hypertension may occur due to hyperparathyroidism. Nevertheless, hypertension has been reported with renal liposarcoma.[6] Up to 65% of patients with primary hyperparathyroidism have associated thyroid disorders.[7] Parathyroid-dependent hypercalcaemia is reported to occur in no more than 1% of patients with thyrotoxicosis, and probably even less frequently because only a few cases have been reported to date.[7]

Mesenteric liposarcoma was reported neither synchronously bilateral nor with endocrine disorders. We must note if the patient's presentation was a co-incidence or an undescribed syndrome.

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