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Atypical Diffuse Lipomatosis with Widespread Abdominal Involvement Showing Increased FDG Uptake on PET/CT Scan

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Abstract

Lipomas are discrete and encapsulated benign tumors which usually involve head, neck, shoulder trunk, lower and upper extremities. However, lipomatosis is a more diffuse fatty tissue growth. A 34 year-old male was presented with a painless abdominal mass and diffuse lipomatosis that involving the peritoneum, retro-peritoneum and abdominal wall. The disease was noticed during the first year of his life as masses in the axillary fossa. Later the disease presented as multi-focal diffuse lipomatosis synchronously involving the intra-peritoneum and retro-peritoneum which is a rare pattern of involvement as in as few cases previously reported in the literature. Radiological evaluation revealed extension into the abdominal organs and structures. Interestingly, all lesions showed high FDG uptake on PET/CT scan, although lipomas do not take up FDG contrary to liposarcomas. The patient was operated many times and the pathological evaluations of all excised samples from the intra and retroperitoneum were consistent with mature lipid tissue. The FDG hypermetabolism on PET/CT scan was attributed to possible aggressive nature of the disease even if it was histopathologically benign. Non-invasive management instead of extensive surgical approach was considered for the patient with asymptomatic disease because of the possible high rates of morbidity and mortality which would be caused by extensive excision.

Keywords:* *Diffuse Lipomatosis; Lipoma; PET/CT scan; FDG uptake; Abdominal involvement

1. Introduction

Lipomas are benign tumors of mature fat cell origin and represent the most common mesenchymal lesion in adults. Diffuse lipomatosis characterised by diffuse non capsulated overgrowth of mature adipose is extremely rare. It usually affects the

extremities, abdomen or trunk. This condition shows a wide phenotypic and distinct histopathologic feature. Even though the condition is clearly defined as a clinical entity, the pathogenesis and biological behaviour remain uncertain [1]. Lipomatosis is typically either neoplastic or hamartomatous in nature. It is often one component in a constellation of developmental or structural abnormalities. Diagnosis may be difficult since many of these disease entities have overlapping features [2]. Though it is histologically benign and resembles simple lipomas, the invasive nature of this lesion makes it potentially life-threatening. Infiltrating lipomatosis of the face, head, neck, extremities, trunk, abdominal cavity and pelvis have been previously reported in the literature. However, involvement of the abdominal cavity including retroperitoneal and intra-peritoneal spaces by diffuse infiltrating lipomatosis has been reported in only a small number of cases [3-5].

2. Case

A 34-year-old man presented with slowly progressive abdominal distension in 2-year duration. There was no associated history of constitutional symptoms, altered bowel habits or alcohol consumption. Blood test revealed a total cholesterol and serum triglycerides in normal range. The other laboratory parameters were also normal. On physical examination, moderate abdominal distension, palpable abdominal mass and signs of operations were found. The computed tomography (CT) scan of the abdomen showed diffuse fatty infiltration of the abdomen including both retroperitoneal space and peritoneal cavity, with encased mesenteric stump by the diffuse lipomatoid masses (FIG. 1). The mass caused displacement, separation, conglomeration and mild compression of the bowel loops. The CT results were typical for diffuse infiltrating abdominal lipomatosis. The biopsy taken from involved site of body revealed a fat tissue without nuclear atypia. The whole-body F-18 Flourodeoxyglucose (FDG) PET-CT scan revealed heterogeneous and peripherally increased FDG uptake in the intraperitoneal and retroperitoneal lipomatous lesions that had also been showed on the previous CT (FIG. 2 and 3). The patient underwent surgical resection for the mass but it couldn't be totally resected because of widespread abdominal infiltration however cholecystectomy and biopsy from the mass were performed. Histopathological examination of the biopsy confirmed the diagnosis of benign lipomatosis.

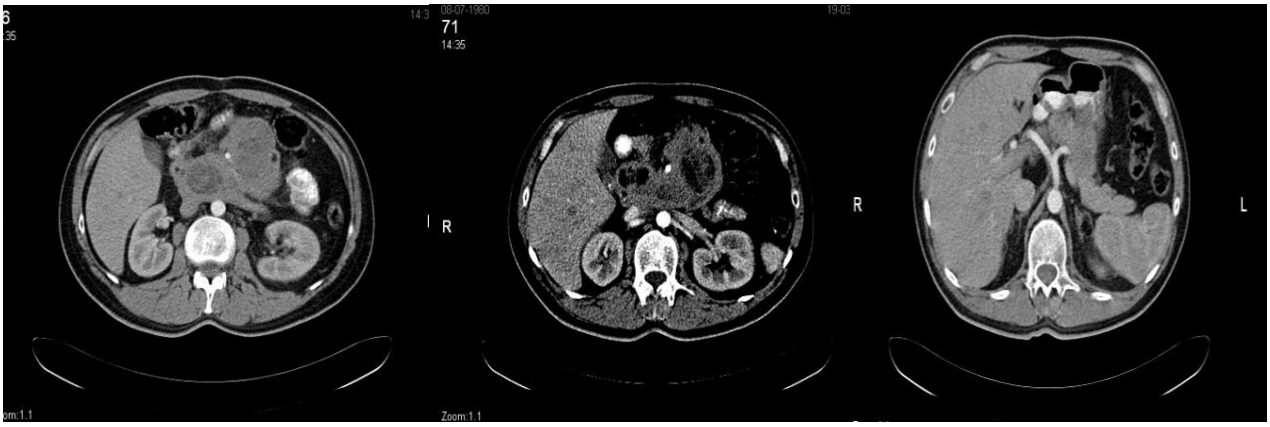

FIG. 1. **Computed tomography images show a large lipomatous mass that infiltrating abdominal structure and mildly compression of the bowel loops.**

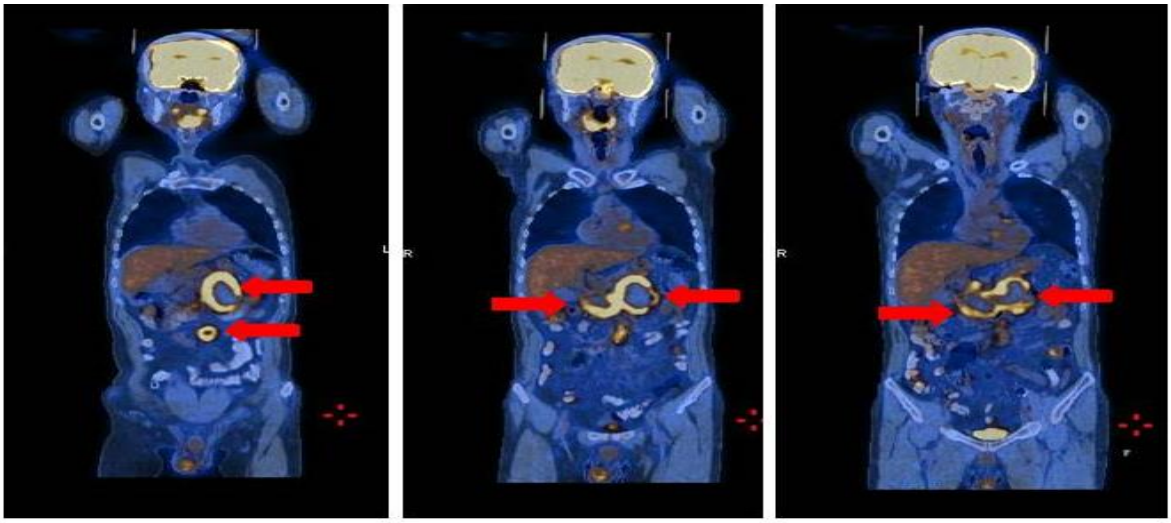

FIG. 2. **Coronal images of FDG PET-CT scan showing heterogeneous and peripherally increased FDG uptake in intraperitoneal and retroperitoneal lesions (arrows).**

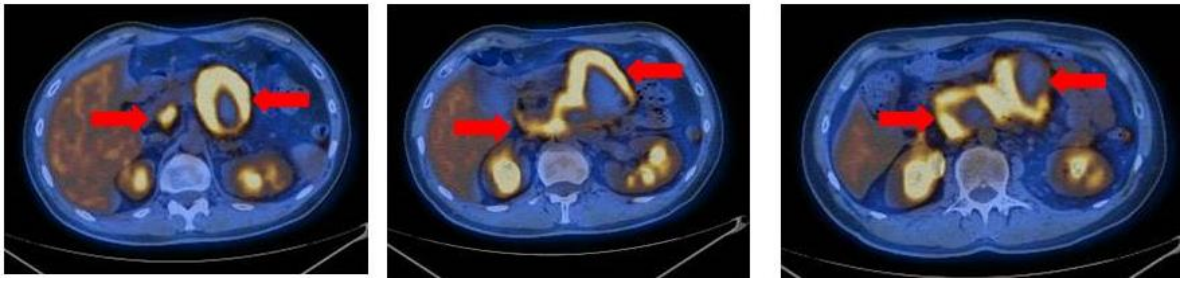

FIG. 3. **Transaxial images of FDG PET-CT scan displaying increased glucose metabolism in intraperitoneal and retroperitoneal lipomatous lesions with heterogeneous and peripheral uptake pattern.**

3. Discussion

Lipomatous tumors are the most common mesenchymal tumors of the abdominal cavity. The most common location of involvement is the skeletal muscles of both upper and lower extremities [1]. They occur usually in the retroperitoneal space but may also arise within the peritoneal cavity. Lipomatous tumors appear in a variety of benign and malignant forms and their classification is often of prognostic and therapeutic significance. Simple benign lipomas are encapsulated tumors with a composition similar to that of normal adipose tissue. Diffuse infiltrating lipomatosis is histologically benign and resembles a simple lipoma except for its invasive nature. Malignant lipomatous tumors that are usually in homogeneous, poorly marginated and invasive are defined as liposarcomas [6]. Diffuse infiltrating lipomatosis can involve a multitude of anatomic sites. Multiple symmetrical lipomatosis (benign symmetric lipomatosis) is a rare entity affecting mostly men between 25 and 60 years of age. This disease which is also called Madelung's disease was first described by Brodie in 1846 and is perhaps one of the more well-known causes of lipomatosis of the neck. It is characterized by collection of large amount of non-

encapsulated lipomas mainly located symmetrically in the subcutaneous tissues of the cervical, supraclavicular, deltoid, thoracic, abdominal and pelvic regions. The lesion most likely originates in the subcutaneous adipose tissue, subsequently penetrating between the muscular fascia or in the spaces between organs [7]. Benign symmetrical abdominal lipomatosis is a variant of multiple symmetrical lipomatosis characterized by massive enlargement of abdomen (pseudoascites) due to intraperitoneal and retroperitoneal fat accumulations. Although an association with alcohol abuse, metabolic abnormalities, polyneuropathy and certain malignancies has been described, the etiology and pathogenesis is largely unknown [8]. Computed tomography is an accurate and clinically useful method to assess mesenchymal tumors of the abdominal cavity. The CT appearances of abdominal lipomatous tumors correlate closely with their gross and microscopic anatomy [6]. Lipomatosis secondary to extended corticosteroid use or Cushing's syndrome is also commonly reported. These lesions can manifest in the mediastinum, mimicking cardiomegaly on imaging with widening of the mediastinal silhouette [9]. This case is also of interest with regard to high FDG uptake in benign lipomatous lesions in the intraperitoneal and retroperitoneal spaces. Normally, lipomas do not show increased FDG uptake, contrary to liposarcomas. Moreover, Suzuki et al [10] concluded in their study that FDG PET-CT imaging can be used successfully in differentiating lipoma from liposarcoma depending on the degree of FDG uptake of the lesions [10]. To the best of our knowledge, there are only a few reports in the literature with high FDG uptake in benign lipomatous lesions. Burdick et al reported a case of hibernoma with increased FDG uptake which mimics a malignant lipomatous lesion [11]. They concluded that hibernoma mostly resemble brown adipose tissue which may be a possible cause of increased glycolytic metabolism presented as high FDG uptake. In our case, increased FDG uptake of benign lipomatous lesions may be attributed to the fact that such intraperitoneal and retroperitoneal lesions had an invasive and aggressive nature even though they were histopathologically benign and they were probably more metabolically active compared to benign lipoma lesions. This possible increased metabolism of invasive lipomatous lesions in our case might be the causative factor for increased FDG uptake on PET/CT scan. The only treatment option for diffuse lipomatosis is limited to surgical excision. Due to diffuse infiltration, complete excision is often impossible. Prognosis and treatment approaches vary from case to case. For example; an infiltrating type of lipomatosis near a major vessel or the spinal cord will have a worse prognosis and surgical excision is more difficult. In our case, imaging revealed a relatively large lesion extending into the abdominal location. Despite the extensive nature of the lipomatosis, the patient was asymptomatic without evidence of bowel obstruction or compression of vital structures, and histopathology ruled out malignancy. Our decision was to follow the lesion conservatively. Long-term follow-up with imaging examination at least once in three months is important because of active proliferative behavior. Likewise there were signs of progression these have been observed in 3 months after surgery in our case.

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