CASE STUDY ARTICLE

PREVIOUS RETROPERITONEAL TERATOMA MASS CLEARED TO BE A DEDIFFERENTIATED LIPOSARCOMA – a case report

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ABSTRACT

Retroperitoneal masses are uncommon but often lead to uncertain diagnosis; the differential one can vary from benign conditions such as developmental cysts or locally aggressive lesions (desmoid tumors) to malignant conditions (retroperitoneal sarcomas, teratomas, or malignant lymph node masses). Most RPLs arise de novo but can occur in a pre-existing lipoma. There are no established causative factors but risk factors include ionizing radiation, chemotherapy, and some genetic conditions. Trauma, although suspected is not a known risk factor. The following case report aims to demonstrate an unconformity between CT scan and histological examination where a previous teratoma was cleared to be a liposarcoma.

1. INTRODUCTION

Retroperitoneal masses are uncommon but often lead to uncertain diagnosis; the differential one can vary from benign conditions such as developmental cysts or locally aggressive lesions (desmoid tumors) to malignant conditions (retroperitoneal sarcomas, teratomas, or malignant lymph node masses). Teratoma is a germ cell tumor, which can contain different types of tissue, like bone, hair, and muscle. depending on the degree of differentiation of the cells it contains, we can speak of mature or immature teratomas that usually occur on the testes in men and the ovaries in women, and the tailbone in children, retroperitoneal involvement is less common. Teratomas may be benign or malignant. the often asymptomatic evolution can lead to a late diagnosis, and the discovery may be accidental during investigations for other pathology [1]. The following case report aims to demonstrate an unconformity between CT scan and histological examination where a previous teratoma was cleared to be a liposarcoma. Retroperitoneal soft tissue sarcomas (STSs) are rare, accounting for 10-15% of all soft tissue sarcomas [2, 3] Of these STSs, retroperitoneal liposarcomas (RPLs) are the most common subtype [3]. These lesions often cause minimal or no symptoms and can reach a significant size, growing undetected in the retroperitoneal space before invading or compressing surrounding organs, eventually leading to clinical symptoms. This makes them challenging to diagnose and subsequently treat effectively.

Most RPLs arise de novo but can occur in a pre-existing lipoma. There are no established causative factors but risk factors include ionizing radiation, chemotherapy, and some genetic conditions. Trauma, although suspected is not a known risk factor [4].

2. CASE REPORT

A 60-year-old man, with a heart disease history (angioplasty for coronary artery stenosis 5 years before) consults for emergencies for abdominal pain which began in the right lower quadrant and has spread to the periumbilical area, with fever, nausea, and vomiting, without externalized gastrointestinal hemorrhage. The blood count showed a neutrophilic leukocytosis, and the CT-Scan revealed the presence of a large heterogeneous mass in the right iliac fossa, with a lot of air bubbles, it seems that this mass includes the appendix, this aspect suggests an abscess of appendicular origin. Furthermore, the Scan showed a tissue formation in the left retroperitoneum, heterogenous containing fat and a calcic

6 months later, the patient underwent surgery to remove the mass in the left retroperitoneum and to establish the continuity to ensure intestinal continuity. The drain was removed 4 days later and the patient was cleared and left the hospital 24 days later. The histological examination shows a formation weighing 481 grams measuring 115 * 80 * 55mm and the immunohistochemical study concludes an intermediate grade dedifferentiated liposarcoma (3 + 1 + 0) R0.

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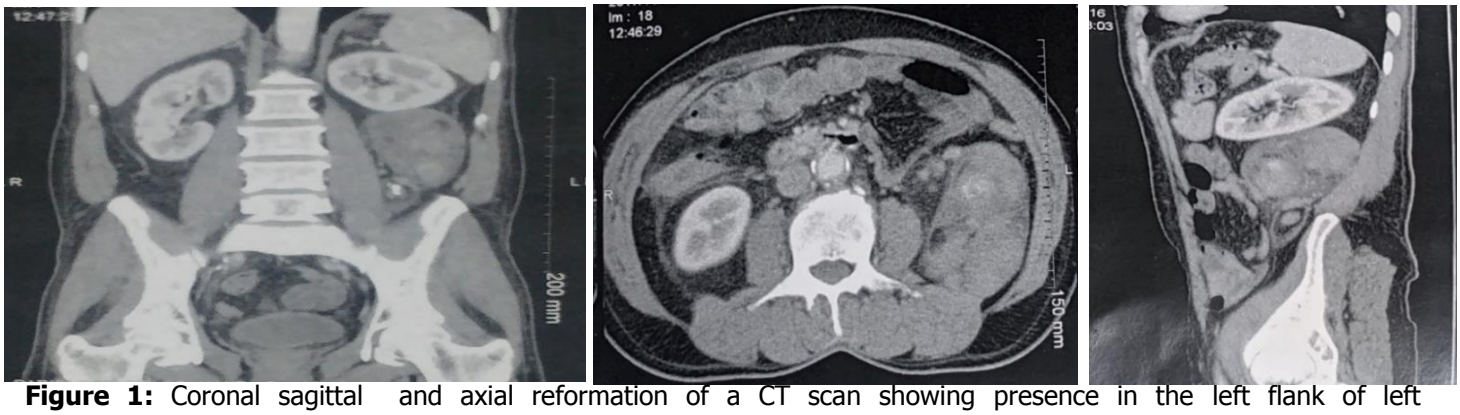

Figure 1: Coronal sagittal and axial reformation of a CT scan showing presence in the left flank of left retroperitoneal formation of 10 cm with a fatty and calcium component suggesting a teratoma.

4. DISCUSSION

The migratory property of germ cells would explain why teratomas generally occur in the extragonadal sites and along with the midline structures. Teratomas are found, with decreasing frequencies, in the ovaries, testes, anterior mediastinum, retroperitoneum, presacral and coccygeal areas, intracranial sites, neck, and abdomen [5] Less than 10% of teratomas are found in the retroperitoneum and they are also much less common in adults over the age of 30 [6].

Reviewing the radiological findings of 23 mature teratomas of the retroperitoneum, Davidson et al., (1989) [7] determined that the most characteristic radiologic finding of these neoplasms is the identification of a complex mass containing a well-circumscribed fluid volume, fat-fluid level (formed by adipose tissue or sebaceous material), and calcifications. Calcification was detected in 60 to 80% of cases of retroperitoneal teratomas and reported to be slightly more common in benign rather than malignant tumors [8]. While historically CT scan was the standard in retroperitoneal imaging, magnetic resonance imaging is very useful in providing improved soft tissue detail, including proximity to adjacent organs and material within the teratoma. It is particularly useful in assessing blood vessel invasion, or encasement [9]. Since definitive diagnosis is only achieved following histologic evaluation of the specimen, surgical resection is paramount for both diagnosis and treatment.

Discrepancies may occur between the CT scan and histological examination about the real nature of such masses; Indeed, in our case, we found an unconformity between radiological and histological examination which confirmed a liposarcoma. Liposarcoma is the most common malignant retroperitoneal soft-tissue tumor in adult patients (35%) followed by leiomyosarcoma and malignant fibrous histiocytoma [10].

It mostly affects 50-70 years old patients with no gender predilection. Unlike liposarcomas in the extremities, retroperitoneal tumors can remain unnoticed and have more time to grow and undergo dedifferentiation. Symptoms depend on their mass effect on adjacent structures. Liposarcomas are subclassified into five groups [11] well-differentiated liposarcomas (WDL), dedifferentiated liposarcoma (DL), myxoid liposarcoma (ML), round cell liposarcoma (RCL), and pleomorphic liposarcoma (PL) with different genetic, clinical, radiologic, and pathologic characteristics:

- WDL: The most common subtype. It appears as a well-defined predominantly fat-containing lesion with thin septa and minimal poorly defined soft-tissue components.
Any retroperitoneal purely fatty lesion (indistinguishable from lipomas) should be considered a WDL and undergo biopsy [12, 13].

They have a good prognosis if they can be completely resected, which is often not possible due to the large proportions the tumor can reach before being symptomatic.

- DL: Dedifferentiation has been reported to occur in 15% of the cases with an average latency period of 7-8 years [14] and has a worse prognosis due to more aggressive behavior and its ability to metastasize. 75% of these tumors occur in the retroperitoneum [12]. It is suggested by focal well-demarcated nodular non-lipomatous regions larger than 1cm [14] within a lesion consistent with WDL. Ossification or calcification often indicates dedifferentiation and is a sign of poor prognosis [15].

ML: They occur in younger patients, typically in extremities and rarely in the retroperitoneum, most of them with enough fat to suggest the diagnosis. Non-lipomatous components include the myxoid component (resembling cysts but with higher attenuation than simple fluid), focal nodules, and thick septa.

- RCL: A high-grade liposarcoma described as a subtype of ML with a minimum amount of fat and a higher amount of round stromal cells that coalesce to form sheets [12].
ML and RCL have a 5-year survival rate of 60-90%.
- PL: It is the less common subtype, affects the retroperitoneum and the extremities with equal frequency, shows little if any visible fat and has a bad prognosis with a local recurrence rate of 30-40% and a 5-year survival rate of 30%. [16]

On the computed tomography (CT), RPLs generally appear as an encapsulated mass, that contains variable amounts of fatty and soft tissue. CT is most useful in delineating the relationship of adjacent structures, assessing local invasion, and checking for the presence of metastatic disease [17].

Also, MRI may demonstrate characteristics of the mass to distinguish between benign and malignant soft-tissue masses. Factors associated with malignant lesions are larger tumor size (> 10cm), thick septa (> 2mm), and less fat content (less than 75%) [18]. Dedifferentiated RPLs often lack macroscopic fat signal intensity. MRI can also assess local tumor extent and surrounding edema, which can then be factored into treatment approaches. It is important to note that the extent of the primary tumor can be underappreciated in imaging studies.

Percutaneous biopsy is controversial as historically, there has been a fear of tumor seeding of the biopsy tract. However, a biopsy is required in cases of diagnostic uncertainty or if neoadjuvant treatment is to be considered.
Recent advancement in molecular genetics has also shed some light on the genes implicated in liposarcomas. Among these are the murine double minute 2 (MDM2) and CDK4 genes. MDM2 encodes a protein that is responsible for the degradation of p53, a known tumor suppressor gene, and CDK4 encodes an oncoprotein that promotes G1/S progression of the cell cycle. Both of these genes are characteristically amplified in well-differentiated and dedifferentiated RPLs [19] . Overexpression of these proteins are detected by immunohistochemistry or amplification may be detected by fluorescence in-situ hybridization and is useful in distinguishing well-differentiated liposarcomas (WDL) and dedifferentiated liposarcoma (DDL) from other benign and malignant soft tissue tumors [20].

The treatment of choice for non-metastasized retroperitoneal soft tissue sarcoma is complete surgical excision with negative margins. Complete en bloc excision may require adjacent organ and fat resection, but even then, surgical margins are often narrow [21]. Unfortunately, even with complete excision, prognosis remains poor, particularly for high-grade RPLs [22]. Radiation therapy and chemotherapy remain controversial [23].
Local recurrence is common and can lead to morbidity and mortality.

5. CONCLUSION

Teratoma is a benign tumor while liposarcoma and more precisely dedifferentiated liposarcoma is a malignant tumor with poor prognosis. Ossification or calcification often indicates dedifferentiation and is a sign of poor prognosis. This radiological sign can be present in both diagnoses. Treatment of RPL is multimodal and should ideally take place in a multidisciplinary center. Complete surgical excision is, however the mainstay of treatment. Careful follow-up is highly recommended to detect the first signs of recurrence and metastasis.

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