GIANT "LIPOMA-LIKE" LIPOSARCOMA OF THE RETROPERITONEUM: A CASE REPORT

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SUMMARY – A giant retroperitoneal liposarcoma that incorporated the descending colon was presented clinically as a palpable abdominal mass in a 37-year-old male patient. Complete surgical resection with adjacent partial colectomy was performed. Total tumor mass was 10.7 kg and pathological examination revealed well differentiated ("lipoma-like") liposarcoma. Literature review shows that aggressive surgical resection remains the mainstay of treatment for retroperitoneal liposarcoma, although there are some promising results that support the use of adjuvant combination chemotherapy in advanced metastatic disease.

Key words: *Liposarcoma – diagnosis; Liposarcoma – pathology; Liposarcoma – surgery; Colon, sigmoid – pathology; Sigmoid neoplasms – pathology*

Introduction

Liposarcoma is the most common soft tissue sarcoma type in adults[1,2]. These neoplasms take their origin from primitive mesenchymal cells and are rarely encountered in fat rich areas such as subcutaneous tissue or subserosa of the intestinal tract which, on the contrary, are the two most common sites of lipomas. These tumors are most likely to arise from deep-seated, well-vascularized structures such as deep soft tissue of the extremities and retroperitoneum[1-3]. Liposarcomas are remarkable because of their frequently large size and histologic variability, and they are one of the largest among all tumors[4].

This paper reports on a case of a well differentiated "lipoma-like" liposarcoma of the retroperitoneum. We review here the natural history, pathology and prognosis of the disease, and discuss the methods of diagnosis and management of such lesions.

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Case Report

A 37-year-old man was admitted to our department complaining of stabbing pain of short onset in the left hemiabdomen. The pain aggravated on deep inhalation, cough and movement. Recent weight loss or gain was not evident. He had no history of surgery or medication, and his family history was unremarkable. The abdomen was distended owing to a huge, tender, palpable mass reaching the left groin. Bilaterally, inguinal groups of enlarged lymph nodes were also palpable.

Peripheral blood tests revealed slight elevation of CRP, ALT and alkaline phosphatase. On abdominal ultrasound and CT scan the mass appeared inhomogeneous with a mainly adipose content extending itself from the xyphoid process to the pubic symphysis, filling the abdominal cavity and the anterior part of the pelvic cavity. Internal organs were pushed to the right side of the abdomen. Preoperative ultrasound guided fine-needle transcutaneous biopsy revealed the presence of adipose tissue.

During laparotomy, a giant retroperitoneally located lipomatous tumor was found filling almost completely the abdominal cavity. The tumor was yellowish in color

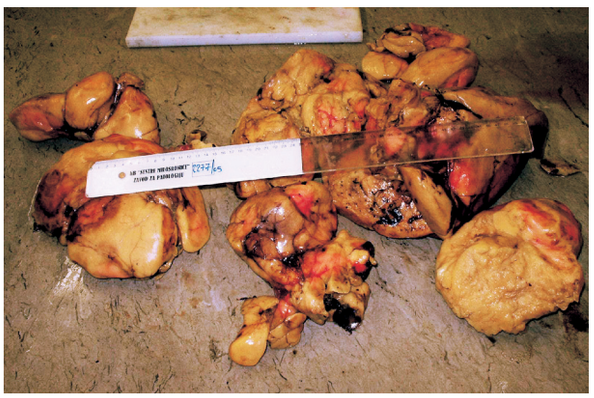

Fig. 1. Resected tumor mass consisting of lobulated yellowish mass which incorporates descending colon.

and multilobular with each lobule partially encapsulated. The descending colon was incorporated in the tumor mass and pushed anteriorly. The rest of the colon and small intestine were displaced to the lower part of the right abdomen. The tumor showed a distinct lobulated pattern. The mass was excised *in toto*, with each lobule removed separately, along with a part of the incorporated and vascularly dependent descending colon (Figs. 1 and 2). Total mass weighted 10.7 kg. Histopathologically, the tumor was well-differentiated containing mature lipocytes and scattered fibroblasts and collagen fibers (Fig. 3). Polymorphism, mitosis and lypoblasts were not found on either frozen or permanent sections. On immunohistochemistry, the lipocytes were S100 protein positive while the reactivity with desmin and SMA was negative. According to gross examination, tumor size, histology and immunohistology, the definitive histologic diagnosis of the specimen was tumor of fat tissue, lipoma-like liposarcoma.

Fig. 2. Tumor mass consisting primarily of fat tissue.

Discussion

Well-differentiated liposarcoma tends to occur in deep soft tissues of both the limbs and the retroperitoneum[1-4]. Liposarcomas are slightly more common in males than in females. The mean patient age at onset is 50 years[5,6]. Most patients with liposarcoma have no symptoms until the tumor is large and impinges on neighboring structures, causing tenderness, pain, or functional disturbances. In the retroperitoneal area, where liposarcoma is detected at a late stage, the tumor may grow to a substantial size, weighing several kilos at the time of diagnosis[4,7]. In general, liposarcoma grows silently and slowly, and the patient's estimation of the clinical duration is often unreliable. The patient eventually becomes aware of a swelling or mass and reports this finding to the physician. Diagnosis is often late, when the tumor has already reached considerable proportions, creating particular problems for its removal[8]. Most liposarcomas have well-defined and mostly lobulated margins. Well-differentiated liposarcomas may be distinguished from other types by their largely lipomatous appearance. Well-differentiated liposarcomas usually contain a predominance of mature fat cells with relatively few, widely scattered lipoblasts. A misdiagnosis of lipoma can result from inappropriate sampling. The well-differentiated type has favorable prognosis, with almost 100% 5-year survival rates. However, this tumor is poorly circumscribed and locally recurs after incomplete excision. Although it rarely metastasizes, repeated local recurrences may cause the tumor to evolve into

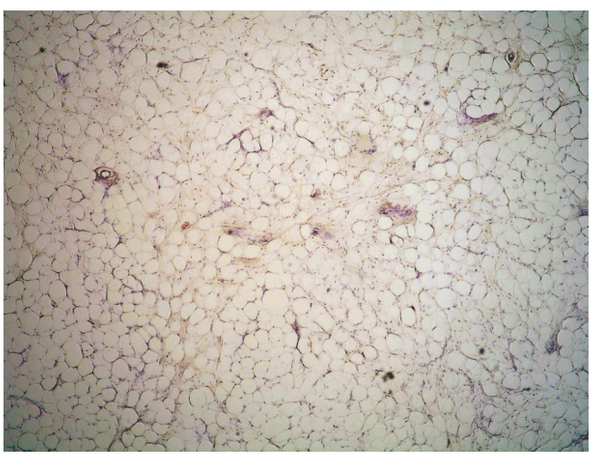

Fig. 3. H&E stained specimen showing well-differentiated liposarcoma (X40).

a higher grade of sarcoma or to dedifferentiate, in which case metastasis is possible[9]. Dedifferentiation identifies a tumor that is potentially more aggressive; yet the progression of the disease following dedifferentiation may be highly variable and probably depends on a number of factors, including the amount of dedifferentiation and type of therapy[10]. Therefore, aggressive surgical management is strongly recommended for primary or recurrent retroperitoneal liposarcomas[11]. The current trend towards aggressive surgery is backed by the improved results in terms of survival. An improved prognosis for patients with retroperitoneal tumors is linked to an early diagnosis and the development of more efficacious complementary therapies[8]. Although the role of chemotherapy for liposarcoma has not been well defined and little data support its use in an adjuvant setting, some articles report effective combination chemotherapy for advanced liposarcoma. Because of the high rate of local recurrences, on follow-up CT or MRI re-evaluation of the patient every 3-6 months during the first 2 years after operation and every 6 months for 3 years thereafter is essential[12].

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Sažetak

DIVOVSKI LIPOMU SLIČAN LIPOSARKOM RETROPERITONEUMA – PRIKAZ SLUČAJA

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Prikazan je slučaj 37-godišnjeg muškarca s palpabilnom tvorbom u abdomenu koja je odgovarala velikom retroperitonealnom liposarkomu koji je zahvatio silazni dio kolona. Učinjena je potpuna kirurška resekcija tumorske tvorbe s djelomičnom kolektomijom. Odstranjena tvorba težila je ukupno 10,7 kg, a patohistološki je odgovarala dobro diferenciranom ("lipomu sličnom") liposarkomu. Uvidom u rezultate objavljenih istraživanja smatra se da je agresivna kirurška resekcija metoda izbora u terapiji retroperitonealnog liposarkoma, dok se upotreba adjuvantne kemoterapije preporuča u liječenju proširene bolesti.

Ključne riječi: *Liposarkom – dijagnostika; Liposarkom – patologija; Liposarkom – kirurgija; Kolon, sigmoid – patologija; Neoplazme sigmoida – patologija*

ANIJA MATIČEVIĆ – Hommage à Pablo Picasso (Madeleine), 2007. ulje na platnu, 40x30 cm