

Retroperitoneal Cystic Lymphangioma in a Patient with Previous Surgery for Seminoma. A Case Report

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ABSTRACT

Retroperitoneal cystic lymphangioma is a rare disease, more frequent in children. The indication for resection is due to the presence of symptoms or compression of the neighboring organs. Hereby, it is described the case of an adult patient with previous surgery for seminoma. Complete resection of the tumor is the treatment of choice. The differential diagnosis in this particular situation is discussed.

Keywords: cystic lymphangioma, retroperitoneum, seminoma, diagnosis

INTRODUCTION

Cystic lymphangioma is a rare malformation of the lymphatic system and usually is located to the craniofacial, neck and chest regions (1). Seminoma represents 40% of the testicular neoplasms and its incidence is increasing (2). For stage I seminoma, radical orchiectomy followed by either chemotherapy or

radiotherapy is associated with a very low recurrence rate (0.2% at 3 years) (3). Hereby we describe the case of a patient with previous surgery for seminoma who was diagnosed with a cystic retroperitoneal lymphangioma. The differential diagnosis in this particular case is discussed. To the best of our knowledge, this is the first description of such an association of the diseases in the English literature. □

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CASE REPORT

A 31-year-old man presented with persistent right abdominal pain, without other clinical symptoms or signs. His medical history included left radical (inguinal) orchiectomy in another surgical unit; pathology revealed testicular seminoma pT1, pN0, cM0, with beta human chorionic gonadotropin (beta HCG) serum level 0.8 mIU/ml (normal range, <2.6 mIU/ml), alpha-fetoprotein (AFP) serum level 5.5 ng/ml (normal range, <6 ng/ml) and lactic dehydrogenase (LDH) serum level 222 U/l (normal range, 135-225 U/l) – stage IA, according to American Joint Committee on Cancer. Assessment of the disease extension was made using only chest ray and abdominal and pelvis ultrasonography. The patient underwent carboplatin-based chemotherapy complicated by aseptic femoral necrosis due to dexametazone administration. Two years after orchiectomy, beta HCG serum level was 0.47 mIU/ml, AFP serum level was 5.87 ng/ml and LDH serum level was 150 IU/l. Other laboratory tests did not revealed any abnormalities. Serum ELISA tests for hydatid disease was negative. Magnetic resonance imaging examination of the abdomen and pelvis showed a large (144/78/153 mm) retroperitoneal mass (Figure 1). No other abnormalities were observed. The patient was referred to surgery and complete tumor resection was performed along with the retroperitoneal lymph nodes; no other abdominal organs were resected. Macroscopic examination of the operative specimen showed a well-defined whitish-tan multilocular cystic mass (Figure 2). The histology revealed cystic lesions with single layer of flattened endothelium, confirming the diagnosis of cystic lymphangioma. □

DISCUSSION

Retroperitoneal cystic lymphangioma is a rare congenital abnormality of the lymphatic vessels, with a benign behavior. Its discovery in adults is very uncommon and appears to be less aggressive than in children (4). The main clinical symptoms are abdominal pain along with palpable abdominal mass (1).

Ultrasonography and computed tomography are the most commonly used imaging techniques for preoperative assessment of cystic lymphangioma (1,5). A typical radiological appearance of a retroperitoneal cystic lymphangioma is a tumor with thin, well-defined

walls, containing a homogenous fluid, with or without septa (5). Magnetic resonance imaging has a better resolution for soft tissue, cystic lymphangiomas exhibiting hypointensity in T1 images and increased intensity in T2 images (6). Retroperitoneal cystic lymphangioma are usually unilocular and large tumors (around 10 cm or more) (1,6). Preoperative diagnosis of a retroperitoneal cystic lymphangioma is challenging; an accurate preoperative diagnosis can be established in around two third of the patients (1,4).

The indication for surgery is due to the presence of symptoms or compression of the surrounding organs. In up to 10% of the cases cystic lymphangioma may complicate with infection or hemorrhage requiring surgery in an emergency setting. Up to now, the malignant potential of such an abnormality has not been demonstrated (1).

Complete resection of the retroperitoneal cystic lymphangioma is the standard surgical approach (6) and can be achieved in more than 90% of the patients (1,4). The local recurrence rate is less than 10% (1), and it is due mostly to an initial incomplete resection of the cyst (6).

The final diagnosis of cystic lymphangioma is revealed at pathology: presence of dilated lymphatic vessels, lined with flattened endothelial cells, abundant lymphoid tissue, smooth muscle present in the cyst wall and absence of atypia (1,4,7).

Seminoma is diagnosed and treated in stage I in almost 75% of the patients. Staging and risk assessment of a patient with seminoma require, besides data from the pathology report, the se-



FIGURE 1. Magnetic resonance imaging of the abdomen: axial T1 (A) and T2 (B) examination showing a large (144/78/153 mm), well-circumscribed, apparently unilocular, retroperitoneal mass (hypodense in T1 and hyperdense in T2), with thin walls; the tumor is situated anterior to the aorta (Ao), inferior vena cava (white arrow); right psoas major muscle (PM) and right kidney (RK)

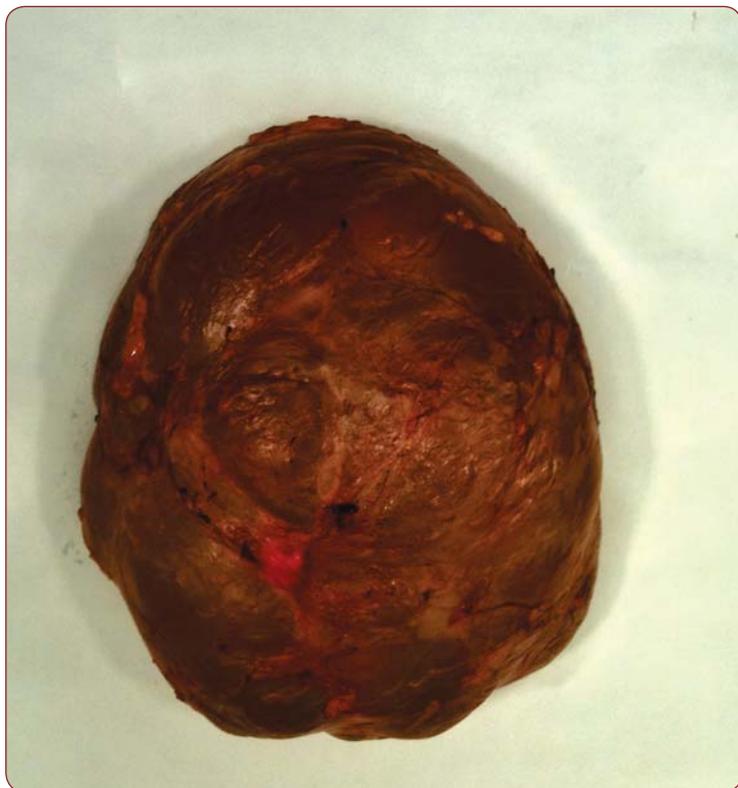


FIGURE 2. Macroscopic aspects of the resected operative specimen

rum level of beta HCG, AFP and LDH. Radical orchiectomy performed through an inguinal incision followed by surveillance is the most co-

mon approach for stage I seminoma. Five years recurrence rate after surgery in patients without associated risk factors is around 12%, retroperitoneal and high inguinal lymph nodes being the most frequent sites of relapse (more than 97% of the cases) (2). However, recent data suggests the benefits of either radiotherapy or carboplatin chemotherapy as adjuvant treatment for stage I seminoma, with an overall cancer-specific survival of 99.8% (3).

In a patient with previous surgery for seminoma the differential diagnosis of a retroperitoneal mass should include retroperitoneal lymph node metastases (2), lymphoma (8) and hydatid disease, the latter especially in endemic areas (1). Up to 1.1% of the patients with previous surgery for seminoma will develop a second nontesticular neoplasm (9). However, lymphoma represents less than 4% of the secondary neoplasia that can develop after radical surgery for seminoma and it is usually seen in patients over the age of 30 years (8). □

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