Resection of a giant retroperitoneal lipoma herniating through the inguinal canal

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SUMMARY

Retroperitoneal lipomas are extremely rare with few cases reported so far in the literature. They can reach different sizes and present with a variety of symptoms. The differential diagnosis is mainly with well-differentiated liposarcoma (WDLPS). We present a 34-year-old woman with a retroperitoneal lipoma herniating through the inguinal canal into the proximal thigh. The patient underwent complete oncological resection using a Karakousis's abdominoinguinal incision. Retroperitoneal lipomas are a very rare condition and sometimes require resections technically challenging. MDM2 amplification is critical for its differential diagnosis with WDLPS.

BACKGROUND

Lipomas are the most frequent soft tissue tumour in adults. They frequently arise from the subdermal tissue of trunk and extremities and are very rare in the retroperitoneum.[1] They are well-circumscribed, lobulated lesions and demarcated from surrounding tissues by a thin fibrous capsule.[2] On the other hand, primary retroperitoneal tumours represent less than 1% of all body tumours and majority of them are malignant. We present a case of a retroperitoneal lipoma herniating through the inguinal foramen.

CASE PRESENTATION

A 34-year-old woman was referred to our surgical unit because of a retroperitoneal mass found on abdominal CT scan performed due to distension and mild unspecific abdominal pain. Her medical history accounted for insulin-resistance, open right inguinal hernioplasty at age 4 and pilonidal cyst resection. Physical examination showed a large lump in the proximal right thigh, but with normal neurological and functional tests. CT-scan showed a retroperitoneal hypodense soft-tissue mass measuring 218×67×53 mm. The tumour was in contact with the right psoas muscle and medially displaced the ipsilateral iliac right vessels. Moreover, it herniated through the inguinal canal into the proximal thigh (figure 1).

An incisional biopsy was performed before referral to our department. Histopathology study of the sample revealed a lipomatous tumour but in that moment MDM2 amplification on the fluorescent in situ hybridisation (FISH) could not be done. After discussing the case in a multidisciplinary tumour board, the resection was made with negative margins (figure 2A,B).

The patient underwent en bloc resection of the tumour using a Karakousis's abdominoinguinal approach. Margins of the tumour lodge were marked with clips. She was discharged on postoperative day 2 without pain and normal extremity functions. Final histopathological examination confirmed a 14×13×7 cm lipoma (636 g) without MDM2 amplification on FISH (figure 2C). After 20 months of follow-up, the patient remains without symptoms and no evidence of local recurrence.

DIFFERENTIAL DIAGNOSIS

Due to the impossibility to establish a definitive diagnosis on an imaging basis only, pathological assessment remains the gold standard of diagnosis. Despite the lack of complete histopathological diagnosis at initial evaluation the final diagnosis was made with the complete surgical specimen using standard techniques and FISH (MDM2 amplification analysis).

TREATMENT

Complete surgical resection is the cornerstone of the treatment of soft tissue tumours.

OUTCOME AND FOLLOW-UP

After 20 months of follow-up, the patient remains without symptoms and no evidence of local recurrence.

DISCUSSION

Benign retroperitoneal lipomas in adults are exceedingly rare with less than 20 cases reported in the literature since 1980 and the first one with these anatomical characteristics to our awareness. The retroperitoneum has a wide potential space and tumours tend to grow to large sizes before becoming symptomatic. Hence, clinical presentation of retroperitoneal lipomas and soft-tissue sarcoma (STS) in general varies in different reports. The differential diagnosis is mainly with well-differentiated liposarcoma (WDLPS) which is—by far—the most frequent histotype in this anatomical site (along with dedifferentiated liposarcoma).[1 3] Of note, WDLPS has high tendency to recur locally and does not metastasise.

CT-scan and MRI are the modalities of choice to assess a retroperitoneal mass. Radiological findings are very similar to those of a simple lipoma making not possible to exclude a WDLPS only by imaging assessment in most cases.[4] Smooth margins with lobular contours and a fatty component with low contrast enhancement are usually present in both.

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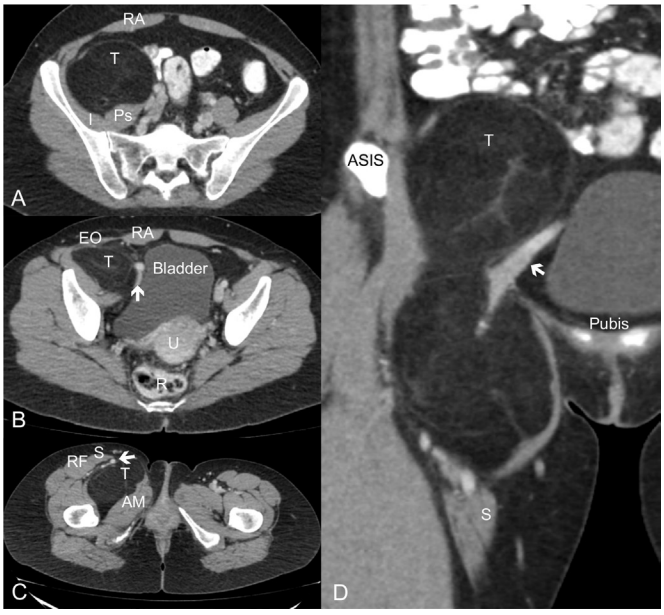

Figure 1 CT scan. (A) Axial view showing a right retroperitoneal soft-tissue tumour in contact with the iliopsoas muscle and medially displacing ipsilateral iliac vessels. (B,C) Axial view showing herniation through inguinal canal into the thigh and displacing forwards the femoral vessels. (D) Coronal view of the tumour with a 'dumbbell appearance'. AM, adductor magnus; Arrows, iliofemoral vessels; ASIS, anterosuperior iliac spine; EO, external oblique; R, rectum, RA, rectus abdominis; RF, rectus femoris; S, sartorium muscle; T, tumour; U, uterus.

Due to this, pathological assessment is the mainstay of diagnosis. Any lipomatous tumour in retroperitoneum should be suspected as malignant unless genetically proven otherwise.[5] Only some WDLPS and DDLPS (de-differentiated liposarcoma) with unequivocal radiological findings can be certainly diagnosed without sample analysis.[1] Therefore, core-needle biopsy, especially decided after a multidisciplinary tumour board evaluation, is the standard diagnostic procedure even though biopsies often remain inconclusive.[4]

In the case presented, we could not reliably rule out the diagnosis of a WDLPS, and, moreover, the tumour had a prominent pelvic extension herniating through the inguinal notch. It had been described that some pelvic soft tissue tumours can extend through the foramina of the pelvis. Furthermore, WDLPS is the histological subtype more propense to do it through the sciatic notch—referred as 'dumbbell tumours'.[6] In our case, because of the foramen involved, we choose to make an abdominoinguinal incision. This approach, along with others, is commonly used for tumours herniating through the obturator foramen.[7 8] Finally, we believe a critical preoperative and intraoperative judgement of tumour's anatomic and histological characteristics are of utmost importance in these scenarios in order to make the best decision. In the case presented, the tumour was encapsulated not showing an infiltrating growth and sole tumour resection was performed. So, surgical planning is critical in retroperitoneal and pelvic STS surgery, especially if malignancy cannot be ruled out. Complete surgical resection is the mainstay treatment[1 9] and the best chance of resection with curative intent is the primary presentation.[10] Since data on retroperitoneal lipomas are scarce, a close and periodic follow-up is advised.

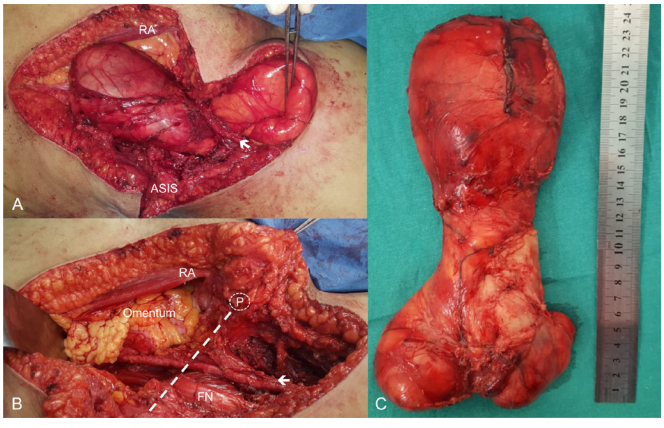

Figure 2 (A,B) Dumbbell lipomatous tumour herniating through the inguinal canal. (C) Gross macroscopic specimen. Arrows, iliofemoral vessels; Dashed line, inguinal ligament; FN, femoral nerve; P, pubic tubercle; RA, rectus abdominis.

Learning points

- Retroperitoneal lipomas in adults are very rare and differential diagnosis is mainly with well-differentiated liposarcoma.
- MDM2 amplification is critical for its final diagnosis.
- Surgical planning is critical in retroperitoneal and pelvic STS surgery; all cases should be discussed in a specialised Multidisciplinary Tumor Board Meeting.

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