Very atypical presentation of a retroperitoneal “atypical lipoma”
A well differentiated liposarcoma presenting as sciatic hernia

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Introduction

Sciatic hernia and the so called “atypical lipoma” are very uncommon if not rare. A combination of both like in our case is probably unique in the literature. The potential for recurrence, undifferentiation and mortality shown by the retroperitoneally located atypical lipomas justifies the term of well differentiated liposarcomas used to define them. We present, a retroperitoneal “atypical lipoma” of 3.5 kg, presenting with subtle symptoms as sciatic hernia. A 53 year old woman with recent onset of dysuria and constipation was referred to us and found to have a sciatic hernia caused by a gigantic retroperitoneal mass protruding through the obturator sciatic foramina. Pathology was consistent with an “atypical lipoma” which is synonymous of well differentiated liposarcoma. The care of a patient like the one we present is greatly affected by the preoperative work-up, the knowledge of the issues around this topic and the surgical technique. History and physical must be thoroughly taken otherwise this pathologic condition can remain unrevealed. CT scan or MR represents a main guidance toward the most appropriate surgical treatment.

Case report

A 53 year old, otherwise healthy woman was referred to us with a 2 year history of worsening dysuria and constipation. An extensive work-up including colonoscopy, pelvic ultrasound and cistoscopy had been pursued by other consultants and it was not contributory. Physical exam was remarkable only for a left gluteal, soft mass, expanding under Valsalva maneuver. A CT scan showed an impressing low density retroperitoneal mass with a 25 cm transversal diameter extending for at least 15 cm above the iliac crest reaching more caudal the pelvis and occupying the space between the rectum, the uterus and the bladder. The mass was then emerging through the perineum medial to the left internal obturator muscle and mainly through the sciatic foramen. Densitometry was suggestive for a mixoid type of neoplasm.
The patient was taken to the operating room and laparotomy was performed. Intraoperative findings were consistent with a very soft, pink-yellow, multilobulate mass, extending in the left retroperitoneum from the transverse mesocolon down into the pelvis, among the pelvic organs and viscera. The most caudal portion of it entered the left obturator canal, the inguinal canal and the sciatic foramina of the same side. There was not evidence of metastatic disease.

Full excision was thoroughly entertained, en-bloc with the parietal peritoneum, paying attention not to open the capsule. The rectum, bladder and uterus were not infiltrated and they were carefully freed-up.

The specimen was passed over but a small portion of it was sent for frozen section which revealed a mesenchimal neoplasm with fibro-mixoid features. In light of the pathology report a primary repair of hernia site was performed and no mesh was used.

The patient had an uneventful postoperative course and was discharged home on the fifth postoperative day. Final pathology showed a 3.5 kg, “atypical lipoma”. Fourteen months after surgery the patient is symptoms free and a follow-up CT scan does not show any recurrence.

Discussion

Sciatic hernias are very rare and they can be of very difficult diagnosis. That is due to both the gluteal muscles covering any mass protruding through the obturator foramina and the lack of specificity of the presenting symptoms.

More often the diagnosis is made either because the hernia reaches a significant volume or incidentally by imaging studies performed during the work-up of non-specific and different symptoms. A wide range of presenting symptoms has been reported by different authors depending upon the herniated viscus: chronic pelvic pain for adnexal involvement, hydronephrosis secondary to bladder and ureteral herniation, and small bowel obstruction accompanied by a gluteal abscess for strangulated bowel loops. Quite often these patients complain of a long history of...
chronic sciatic nerve pain, secondary to direct nerve compression by the hernia. This pain usually goes underestimated by the patients like in the case we observed. A thorough history and physical may sometimes uncover useful findings to make diagnosis but most of the time imaging studies (CT scan, barium enema, and small bowel contrast study) allow to achieve the goal. Based on the findings of these studies a surgical approach can be tailored to the case and dangerous therapeutic errors can be prevented.

Among all the diagnostic tools available CT scan is by far the most useful. The term “atypical lipoma” can be misleading if its potential for recurrence, undifferentiating and mortality is not considered. This potentiality is particularly seen in deeply located lesions of greater volume, like the one we observed. Under the above circumstances the recurrence rate can be as high as 50-80% in 20% of cases and the mortality 30% of it has been stressed that those lesions showing the above mentioned features should be considered “well differentiated liposarcoma”, emphasizing their potentially malignant behaviour, whereas the term “atypical lipoma” should be reserved for the superficially located, more easily managed ones.

The clinical presentation of retroperitoneal tumors is usually non-specific and very rarely they show themselves as hernias. In a large series of 1736 of inguinal hernia repairs only two liposarcomas were found while occasionally other neoplasms have been described.

Recurrence rate and disease free survival of liposarcomas are based on the extent of surgical excision at the time of first operation. In fact a radical operation at that time, respectful of capsular integrity, prevents recurrence and need for reoperation unless the primary tumor was poorly differentiated, non completely resectable or excised in a non radical fashion.

The large experience at the Memorial Sloan Kettering Cancer showed an improvement in the 5 survival, reported as 2% in 1951 versus 78% in 114 more recent cases (1982-1987) treated with curative intent.

The 5 year survival reported by different authors for retroperitoneal sarcomas range from 29% to 70%. Nonetheless those reporting the lowest survival rates do not distinguish between patients treated with curative intent versus palliation.

Taking an intact pseudocapsule en-bloc with organs or structure involved by the tumor at the time of the first operation is an important step toward a curative treatment. There is not clear consensus regarding the use of adjuvant treatments for this category of tumors, although some improvement in local recurrence control has been reported.

In conclusion we feel that the finding of a rare type of hernia like in the case we described must prompt a thorough preoperative work-up either by CT scan or MR. Our experience as well as the others suggest that the hernia content is not always predictable and the appropriate surgical treatment can be provided only if adequate information can guide the preoperative planning.
References


A. Cappellani et al