Very atypical presentation of a retroperitoneal "atypical lipoma"

A well differentiated liposarcoma presenting as sciatic hernia

Ann. Ital. Chir., 2007; 78: 69-72

Alessandro Cappellani, Antonio Zanghì, Maria Di Vita, Domenico La Porta, Giovanni Alfano, Alessio Francesco D'Angelo

Dipartimento di Chirurgia, Cattedra di Chirurgia Generale, Sezione di Chirurgia Generale e Senologia (Responsabile: Prof. A.Cappellani) dell'Università di Catania.

Very atypical presentation of a retroperitoneal "atypical lipoma": A well differentiated liposarcoma presenting as sciatic hernia

Unlike hernias and neoplasms of any other body site, the sciatic hernia is uncommon and the finding of an atypical lipoma in it is probably unique. In such instance making the correct diagnosis is paramount in order to perform a procedure with a radical intent. A CT scan must be considered any time a rare form of hernia is observed and the surgical treatment of a retroperitoneal lipoma has to be radical, to prevent a recurrence. This is the strategy followed by the authors in a case of a 53 year old lady presenting with a large retroperitoneal lipomatous neoplasm within a sciatic hernia.

KEY WORDS: Atypical lipoma, Liposarcoma, Sciatic hernia

Introduction

Sciatic hernia and the so called "atypical lipoma" are very uncommon if not rare [1]. 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.

Pervenuto in Redazione Marzo 2006. Accettato per la pubblicazione Giugno 2006.
Per la corrispondenza: Prof. Antonio Zanghì, viale Andrea Doria 55, 95125 Catania (e-mail:amzanghi@unict.it).

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 foramens and the lack of specificity of the presenting symptoms.

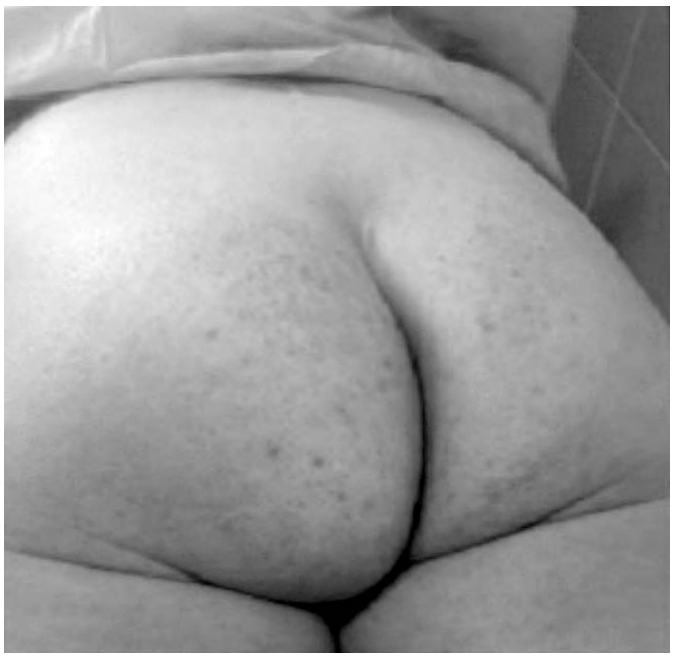
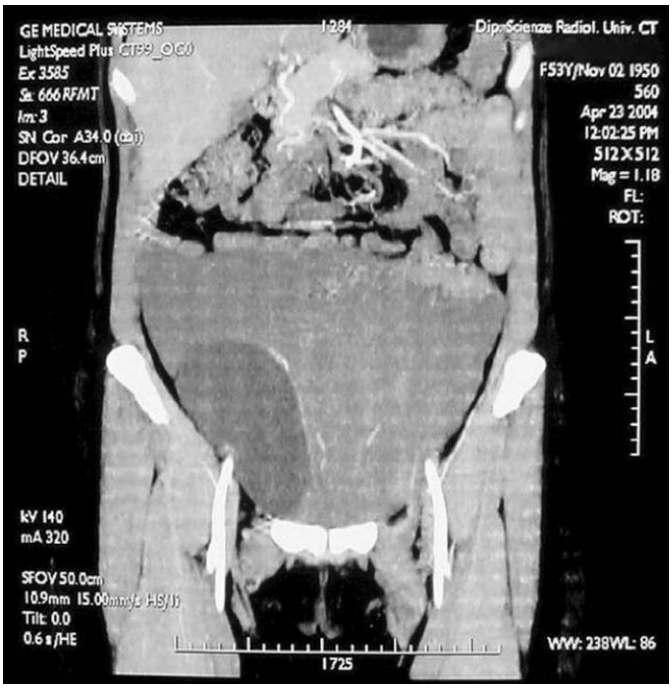


Fig. 1: 53 years old lady presenting with sciatic hernia.



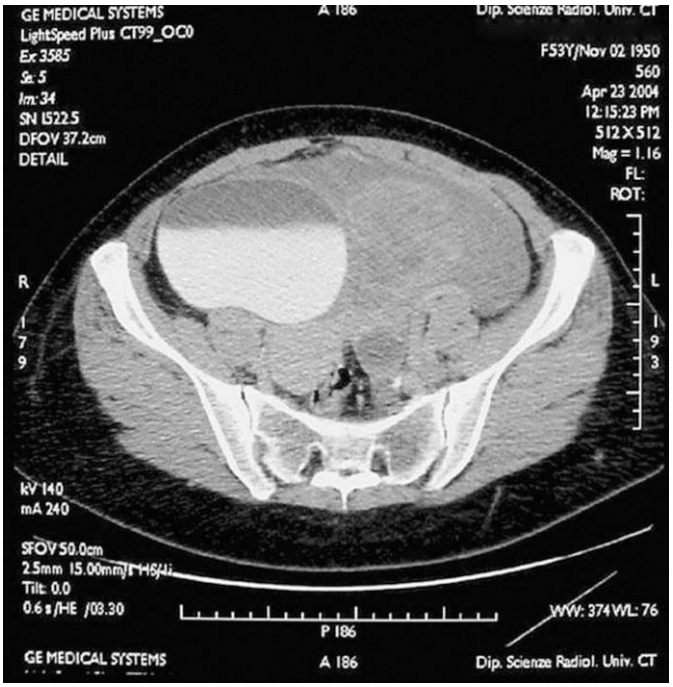


Fig. 2a, b: Preoperatory CT scan.

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 [2,3], hydroneprosis secondary to bladder and ureteral herniation [4-8], and small bowel obstruction accompanied by a gluteal abscess for strangulated bowel loops [9].
Quite often these patients complain of a long history of

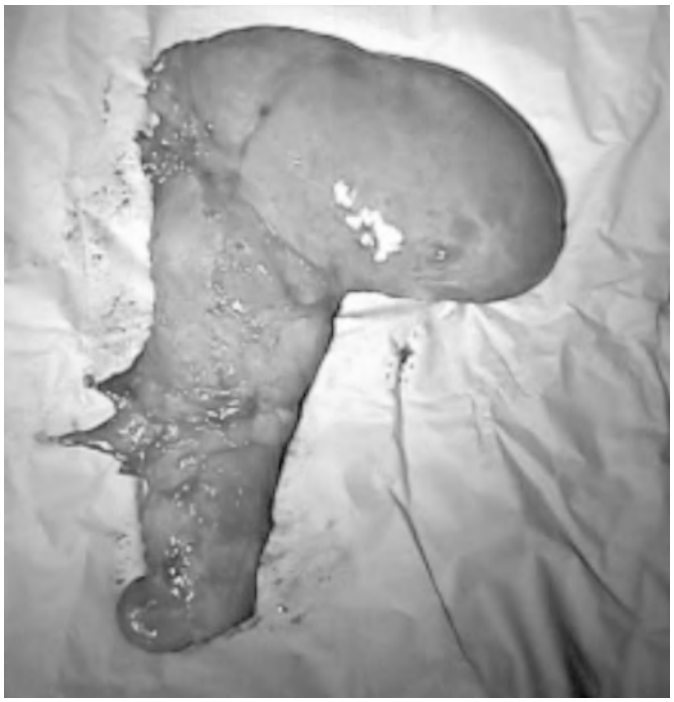


Fig. 3: The neoplasm. The arrow indicates the herniated part of lipoma.

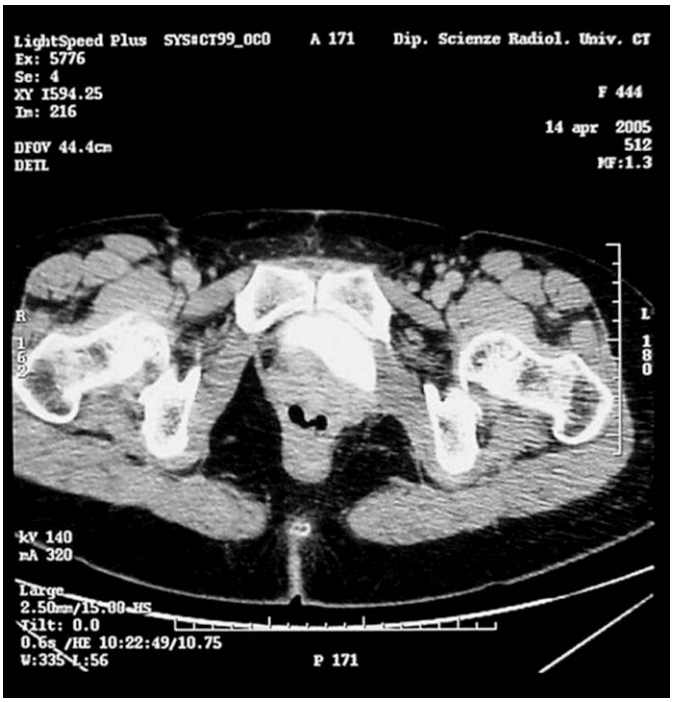


Fig. 4: 12 months follow up: CT scan.

chronic sciatic nerve pain, secondary to direct nerve compression by the hernia [10,11]. 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 [12,13].

Among all the diagnostic tools available CT scan is by far the most useful [14,15].

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 the times.

Therefore in the last twenty years be as high as 50-80%, undifferentiation can be observed 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 [16-21].

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 [22] while occasionally other neoplasms have been described [23].

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 [24,25], non completely resectable or excised in a non radical fashion.

The large experience at the Memorial Sloan Kettering Cancer [26-28] 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 69% of time.

The 5 year survival reported by different authors for retroperitoneal sarcomas range from 29% to 70% [26-32]. Nonetheless those reporting the lowest survival rates do not distinguish between patients treated with curative intent versus palliation [31,32].

Taking an intact pseudocaspule 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 [33].

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 [33].

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

1) Alexander JH, Bouillot JL, Dehni N: EMC, 1994; 40156.

2) Miklos JR, O'Reilly MJ, Saye WB: *Sciatic hernia as a cause of chronic pelvic pain in women.* Obstet Gynecol, 1998; 91(6):998-1001.

3) Carter JE: *Surgical treatment for chronic pelvic pain.* JSLS, 1998; 2(2):129-39.

4) Arat A, Haliloglu M: *Ureteral-sciatic hernia in a child demonstrated by voiding cystography.* J Urol, 1998; 160(1):157-58.

5) Ritschel S, Heimbach D, Schoeneich G: *Ureterosciatic hernia.* Scand J Urol Nephrol, 1996; 30(5):423-24.

6) Noller MW, Noller DW: *Ureteral sciatic hernia demonstrated on retrograde urography and surgically repaired with Boari flap technique.* J Urol, 2000; 164(3 Pt 1):776-77.

7) Garritano A, Vecchioli Scaldazza C, Tobaldi P: *Su di un caso di ernia dell'uretere nel forame sciatico.* Minerva Urol Nefrol, 1985; 37(3):315-16.

8) Epner SL, Lautin EM: *Case report: intermittent sciatic herniation of the ureter.* Clin Radiol, 1994; 49(11):832-33.

9) Yu PC, Ko SF, Lee TY, Ng SH, Huang CC, Wan YL: *Small bowel obstruction due to incarcerated sciatic hernia: ultrasound diagnosis.* Br J Radiol, 2002; 75(892):381-83.

10) Servant CT: *An unusual cause of sciatica. A case report.* Spine, 1998; 23(19):2134-136.

11) Kostka A, Anielski R, Kaczmarczyk D, Szymanik S: *Dwa przypadki rzadkich przepuklin brzusznych. [Two cases of rare abdominal hernias]* Przegl Lek, 1999; 56(10):684-55.

12) Guzman-Valdivia-Gomez G, Guzman-Valdivia E: *Hernia ciatica: reporte de un caso y revision de la literatura.* Gac Med Mex, 1996; 132(1):85-87.

13) Hayashi N, Suwa T, Kimura F, Okuno A, Ishizuka M, Kakizaki S, Kawakami H: *Radiographic diagnosis and surgical repair of a sciatic hernia: Report of a case.* Surg Today, 1995; 25(12):1066-68.

14) Ghahremani GG, Michael AS: *Sciatic hernia with incarcerated ileum: CT and radiographic diagnosis.* Gastrointest Radiol, 1991; 16(2):120-22.

15) Kammori M., Mafune K., Hirashima T, Kawahara M, Hashimoto M., Ogawa T., Ohta H, Hashimoto H, Kaminishi H: *Forty-three cases of obturator hernia.* Am J Surg, 2004, 187(4):549-52.

16) Dei Tos AP: *Lipomatous tumours.* Current Diagnostic Pathology, 2001: 7:178-16.

17) Evans HL, Soule EH, Winkelmann RK: *Atypical lipoma, atypical intramuscular lipoma, and well differentiated retroperitoneal liposarcoma: A reappraisal of 30 cases formerly classified as well differentiated liposarcoma.* Cancer, 1979; 43(2):574-84.

18) Kindblom LG, Angervall L, Fassina AS: *Atypical lipoma.* Acta Pathol Microbiol Immunol Scand [A], 1982; 90(1):27-36.

19) Weiss SW: *Lipomatous tumors.* Monogr Pathol. 1996; 38:207-39. Review. PMID: 8744279.

20) Weiss SW, Rao VK: Well-*differentiated liposarcoma (atypical lipoma) of deep soft tissue of the extremities, retroperitoneum, and miscellaneous sites. A follow-up study of 92 cases with analysis of the incidence of "dedifferentiation".* Am J Surg Pathol, 1992; 16(11):1051-58.

21) Mentzel T, Fletcher CD: *Lipomatous tumours of soft tissues: An update.* Virchows Arch, 1995; 427(4):353-63. Review.

22) Montgomery E., Buras R: *Incidental liposarcoma identified during hernia repair operations.* J Surg Oncol, 1999; 71(1):50-53.

23) Torretta A, La Torre V, Sorcini A, Panarese A, Tonini E, Zeri KP, Mascagni D, Arcieri S, Giacomelli L, Filippini A: *Peritoneal mesothelioma in a case of inguinal hernia. A review of the literature.* Ann Ital Chir, 2003; 74(5):583-87.

24) Van Dalen T: *Management of soft tissue sarcoma in the retroperitneal space: a population based study in the Netherlands. 2000* Utrecht University: Utrecht.

25) Pack GT, Tabah EJ: *Primary retroperitoneal tumors: A study of 120 cases.* Int Abstr Surg, 1954; 99(4):313-41.

26) Kinne DW, Chu FC, Huvos AG, Yagoda A, Fortner JG: *Treatment of primary and recurrent retroperitoneal liposarcoma. Twenty-five-year experience at Memorial Hospital.* Cancer, 1973;31(1):53-64.

27): *The continuing challenge of retroperitoneal sarcomas.* Cancer. 1981 May 1;47(9):2147-52.

28) Lewis JJ, Leung D, Woodruff JM, Brennan MF: *Retroperitoneal soft-tissue sarcoma: analysis of 500 patients treated and followed at a single institution.* Ann Surg, 1998; 228(3):355-65.

29) McGrath PC, Neifeld JP, Lawrence W Jr, DeMay RM, Kay S, Horsley JS 3d, Parker GA.: *Improved survival following complete excision of retroperitoneal sarcomas.* Ann Surg, 1984; 200(2):200-4.

30) Alvarenga JC, Ball ABS, Fisher C: *Limitations of surgery in the treatment of retroperitoneal sarcoma.* Br J Surg, 1991; 78:912-16

31) Makela J, Kiviniemi H, Laitinen S: *Prognostic factors predicting survival in the treatment of retroperitoneal sarcoma.* Eur J Surg Oncol, 2000; 26:552-25

32) Pirayesh A, Chee Y, Helliwell TR, Hershman MJ, Leinster SJ, Fordham MV, Poston GJ: *The management of retroperitoneal soft tissue sarcoma: a single institution experience with a review of the literature.* EJSO, 2001:27:491-97

33) D'Amico DF, Frego M: *I tumori retroperitoneali.* Collana monografica della Società Italiana di Chirurgia, n. 2, Roma, 1994.