Retroperitoneal schwannoma presenting as an ovarian tumor

F. Tomao[1], L. Mariani[3], E. Miele[2], G.P. Spinelli[2], S. Tomao[1, 3].

[1]Department of Gynecology, Perinatology and Puericulture Science, University of Rome "La Sapienza"
[2]Department of Experimental Medicine and Pathology, University of Rome "La Sapienza", [3]National Cancer Institute of Rome (Italy)

Summary

A mass in the left annexal zone was discovered in a 56-year-old woman at the Department of General Surgery and was diagnosed as ovarian cancer. After the operation the mass appeared histologically to be retroperitoneal leyiomiosarcoma and because of residual disease, confirmed by computed tomography (CT) and nuclear magnetic resonance (NMR), complementary radiotherapy was carried out. Restaging supported the persistence of the tumor and so a second laparotomy was performed with complete tumor resection; the pathologic diagnosis was retroperitoneal benign schwannoma. The importance of careful preoperative imaging, such as echography, CT, NMR, arteriography and urography should be stressed for a correct clinical and surgical approach. Moreover, considering that in some selected clinical cases these tumors could be confused with others deriving from contiguous organs and structures, a different surgical approach may be needed together with dedicated and expert surgeons.

Key words: Schwannoma; Ovarian cancer; Sarcoma.

Introduction

Primary retroperitoneal tumors are relatively rare; the histopathological types vary and preoperative diagnoses are very difficult, suggesting the necessity to have a multidisciplinary approach in the diagnostic phase and in the surgical attempt. We present a case of retroperitoneal schwannoma presenting as an ovarian neoplasm with the common staging methods of investigation.

Case Report

A 56-year-old woman, with no significant oncologic anamnesis, was referred to the Department of General Surgery; the main characteristics were menopause at 36 years, two pregnancies and two spontaneous deliveries, ex-smoker, moderate consumer of alcohol and hypothyroidism.

She came to our attention due to a positive pelvic sovrapubic echography that showed tumefaction in the left annexal zone, 96 mm x 70 mm x 71 mm in size, with irregular margins showing solid and liquid material inside with numerous concamerations.

One day after a subsequent sovrapubic pelvic echography confirmed the presence of numerous transonic lesions in the left ovary. These lesions were contiguous and of variable sizes from 2.5 to 4.7 cm, which, moreover caused an increment in overall sizes (about 7 cm). Ultrasonographic diagnosis was completed with addominal and pelvic nuclear magnetic resonance (NMR) imaging that confirmed the presence of a lesion in the left annexal zone. The lesion appeared with numerous concamerations (8.5 x 7 x 8 cm) and with a dishomogeneus paranchymatous spot in the superior left profile. Tumor markers (CEA, CA125, TPA, CA19-9, β-hCG, and αfetoprotein) were normal. The patient was therefore sent to surgery for cytoreduction and removal of the mass. During the surgery the lesion appeared to be located in the retroperitoneal space; it was characterized by a round lesion with hard consistency, fixed to the posterior peritoneum, and without any adnexal connection.

The mass, because of the large size and tenacious adherences that invaded the left ilac vessels and homolateral ureter, was removed leaving tumoral residual infiltration.

Extemporaneous histological exam showed a well differentiated leiomyosarcoma characterized by proliferation of the fusal cells organized in intertwisting bundles, divided by a fibrosis matrix, with acidophilic cytoplasm and long leptochromatic, pleomorphic nucleous and rare mitoses.

Not considering radical surgery, an angio-spiral volumetric CT of the thoracic, abdominal and pelvic regions was carried out. This evaluation showed a residual parenchymatous mass in the left iliac fossa (5 cm) with a wildly necrotic component, inglobing the common iliac vein and artery, and internal and external iliac vein of the same side. Furthermore there were psoas muscle infiltrations and adherences of the ureter which were anteriorly and laterally dislocated. There were also adherences to the anterior sacral bone cortix without erosive aspects. Tumefaction appeared divided by cleavage planes from the left annexal zone, uterus, bladder and sigmoid ansa.

The patient received radiotherapy treatment adjuvated by radiosensibilization with cisplatin given with a bi-daily fractionation of 120 cGy and continuous infusion of 5-fluorouracil. The radiotherapy was executed with a two-field wedge technique until a partial dose of 5080 cGy and with a four-field wedge technique as a boost for the lesion using CT-guided centring.

After consulting a radiotherapist, angio-CT was executed two months later, in which a volumetric increment of the parenchymatous mass in the left iliac fossa was evidenced (5-6 cm) with a larger necrotic component with respect to the original lesion.

The tumefaction was not adherent to the psoas muscle, but it did infiltrate the ureter.

The patient was hospitalized in the surgical division for retroperitoneal mass asportation and cholecystectomy for cronic cholecystitis.

The anatomopathological response showed a lesion of 8 x 5 x 5 cm with a smooth surface, clear outline, grayish-white color and with a cystic mass about of 3 x 2 cm. This mass consisted

Revised manuscript accepted for publication October 20, 2005

of fusal cells with edema, ialinosi which tended to form whirl like formations.

There were also some cellular polymorphisms and rare mitoses. The histopathological diagnosis was cellular schwanoma with periureteral and tissue outlines without neoplastic infiltration.

Discussion

Schwannoma can be present in many locations in which there are schwann cells. The most important involve head and neck nerve endings in 40-50% of cases, peripherical nerves of the upper limbs in 19% and lower limbs in 13%. There are also other localizations, for example, the pelvis in 14%. Von Recklinghausen syndrome is frequently associated with retroperitoneal localization, especially for malignant histotypes, but its frequency is 3% of cases and represents about 4% of all retroperitoneal cancers.

Retroperitoneal schwannomas can be asymptomatic, until the mass size produces compression to the contiguous organs. The clinical features are a palpable mass and continuous pain. Other symptoms such as fever, weight loss and aspecific gastrointestinal and urogenital illness deriving from compression can also be present.

Instrumental examinations such as echography, CT, NMR, arteriography and urography are useful for the preoperative diagnosis.

In several situations these lesions are confused with others; deriving from contiguous organs and structures. Many articles have reported numerous cases of schwannomas mimicking lesions in the hepatic caudal lobe, psoas muscle abscess or a pancreatic tumor, etc.

However, to be sure about the preoperative diagnosis, a categorical pathological opinion should be mandatory but this is not always possible from a technical point of view. In fact very frequently a percutaneous preoperative biopsy is not diagnostic for two reasons: the first is that the histological sample may be not satisfactory from a qualitative and quantitative point of view, while the second reason is that a presumable benign lesion does not exclude the presence of degenerative areas in non biopsied regions of the mass.

One of the first descriptions of a retroperitoneal tumor was by Morgagni, who in 1761 described a retroperitoneal lipoma [8]. In 1946 Donnelly [9] reported a study with 95 cases of retroperitoneal tumors, 32 of which were sarcomas. Then some years later, in 1954, Johnson *et al.* reported on a total of 56 cases of retroperitoneal tumors, 24 sarcomas [10].

The most important factors that influence patient prognosis are anatomical barriers which make radical resection difficult, high histological grading and local recurrence. In fact from 1951 to 1971 five-year-survival was about 40% in patients who received complete surgical resection of a mass.

Some authors instead affirm that after apparently complete resection, the probabilty of a recurrence within five years is approximately 85%. Karakousis *et al.* [12] and Kilkenny *et al.* [13] reported cases of complete resection, respectively, of 95% and 78% with a 5-year-survival of 66% and 56%. Other opinions suggest that a preoperative biopsy is not useful, even if CT-guided, and advise making a diagnosis on the basis of radiological examinations to obtain a sure diagnosis and to address surgical treatment. Furthermore some authors have observed that survival was directly influenced by the morphological properties of tumors. A study conducted by van Dallen showed a 5-year-survival of 39%, with a median follow-up of seven years and with 42% of local recurrence and 22% of metastasis.

In this case the benign characteristics of the mass gave us the opportunity to cure the patient with no apparent risk of relapse. However some considerations were mandatory and oblige a discussion about the best diagnostic procedure in similar cases and also the opportunity to involve more specialists in the clinical decision in a multidisciplinary approach. Mostly it is mandatory to have a team of surgeons, radiologist and pathologist with proven experience in the diagnosis and therapy of retroperitoneal tumors and all of them must consider the possibility of an error in the diagnosis of the lesion, and consequently in the consequent therapeutic approach. In our case the pathological error that occurred after the first surgical approach, comported unnecessary radiotherapy, with ethical, legal and clinical consequences. It is evident that in case of connective tumors arising from complex sites of the body it is necessary to discuss the pathological characteristics of the tumor because of the high risk of a misleading diagnosis. In this case the definitive surgical cure was successfully carried out during the second surgery, performed by surgeons with strong expertise in the treatment of retroperitoneal tumors, considering also the technical difficulties derived from the precedent exposure of the patient to surgery and radiotherapy.

References

[1] Ziparo V., Bocchetti T., Mercantini P., Di Giacomo G., Caleno M.P., Lucandri G.: "Lo schwannoma a localizzazione retroperitoneale". *Giorn. Chir.,* 1996, *17,* 399.

[2] Napoletano C., Mutter D., Steib I.P., Christmann D., Marescaux J.: "Schwannoma benigno retroperitoneale con lisi vertebrale". *Ann. Ital. Chir.,* 1998, *69,* 6.

[3] Pirayesh A., Chee Y., Helliwell T.R., Hershman M.J., Leinster S.J., Fordham M.V., Poston G.J.: "The managment of retroperitoneal soft tissue sarcoma". *EJSO,* 2001, *27,* 491.

[4] Hurley L., Smith J.J., Larsen C.R., Silverman M.L.: "Multiple retroperitoneal schwannoma: a case report and review of the literature". J. *Urol.,* 1994, *151,* 413.

[5] Das Gupta T.K., Brasfield R.D., Strong E.W., Hajdu S.I.: "Benign solitary schwannoma (neurilemamas)". *Cancer,* 1969, *24,* 355.

[6] Steers W.D., Hodge G.B., Johnson D.E., Chaitin B.A., Charnsangavej C.: "Benign retroperitoneal neurilemoma without von Recklinghausen's disease: a rare occurrence". *J. Urol.,* 1985, *133,* 846.

[7] Weber O., Zerbib M., Peyret C., Desligneres S., Debrè B.: "Schwannome bénin rétropéritonéal". *J. Urologie,* 1993, *99,* 189.

[8] Morgagni: "Large retroperitoneal lipoma". *S. Clin. North. Am.,* 1934, *14,* 601.

[9] Donnelly B.A.: "Primary retroperitoneal tumours: a report of 95 cases and review of the literature". *Surg. Gynecol. Obstet.,* 1946, *83,* 705.

[10] Johnson A.H., Searls H.H., Grimes O.F.: "Primary retroperitoneal tumors". *Am. J. Surg.,* 1954, *88,* 155.
[11] Cody H.S., Turnbull A.D., Fortner J.G., Haiadu S.I.: "The continuing Challenge of retroperitoneal sarcomas". *Cancer,* 1981, *47,* 2147.
[12] Karakousis C.P., Kontzoglou K., Driscoll D.L.: "Resectability of retroperitoneal sarcomas, a metter of surgical technique?". *Eur. J. Surg. Oncol.,* 1995, *21,* 617.
[13] Kilkenny J.W., Bland K.I., Copeland E.M.: "Retroperitoneal sarcoma: the University of Florida experience". *J. Am. Coll. Surg.,* 1996, *182,* 329.
[14] Voros D., Theodorou D., Ventouri K. *et al.*: "Retroperitoneal tumors: do the satellite tumors mean something?". *J. Surg. Oncol.,* 1998, *68,* 30.
[15] van Dalen T.: "Management of soft tissue sarcoma in the retroperitoneal space: a population based study in the Netherlands". Utrecht University, 2000.
[16] Schindler O.S., Dixon J.H., Case P.: "Retroperitoneal giant schwannomas: report on two cases and review of the literature". *J. Orthopaed. Surg.,* 2002, *10,* 77.
[17] Patocskai E.J., Tabatabain M., Thomas M.J.: "Cellular schwannoma: a rare presacral tumor". *Can. J. Surg., 45,* 141.
[18] Murat D., Gokhan Y., Mufit C.Y., Semith G., Salih D., Sadettin C., Saffet D.: "Retroperitoneal benign schwannoma: Report of three cases and analysis of clinico-radiologic finding". *Tohoku J. Exp. Med.,* 2003, *200,* 93.
[19] Kishi Y., Kajiwara S., Seta S., Kawauchi N., Suzuki T., Sasaki K.: "Retroperitoneal schwannoma misdiagnosed as a psoas abscess". *Surg. Today,* 2002, *32,* 849.
[20] Pittash D., Klose S., Schmith J., Roessner A., Ridwelski K., Lippert H., Lehenrt H.: "Retroperitoneal schwannoma presenting as an adrenal tumor". *Esp. Clin. Endocr. Diabetes,* 2000, *108,* 318.
[21] Daneshmand S., Youssefzadeh D., Chamie K., Boswell W., Wu N., Stein J.P., Boyd S., Skinner D.G.: "Benign retroperitoneal schwannoma: a cese series and review of the literature". *Urology,* 2003, *62,* 993.
[22] Calo P.G., Congiu A., Ferreli C., Nicolosi A., Tarquini A.: "I tumori retroperitoneali primitivi. Nostra esperienza". *Min. Chir.,* 1994, 49, 43.
[23] Cavallaio V., Bonaccorso R., Selvaggio C., Mazzone S., Fisichella G., Guardalà V.F. *et al.*: "Inquadramento nosografico dei tumori retroperitoneali". *Acta Chir. It.,* 1991, *47,* 820.

Address reprint requests to:
S. TOMAO, M.D.
Via A. Baldovinetti, 83
00142 Rome (Italy)