CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 6 (2015) 233–236

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Giant retroperitoneal ancient schwannoma: Is preoperative biopsy always mandatory?

Mohammad Saad Al Skaini[a], Hesham Haroon[a], Ali Sardar[a], Mohammad Bazeed[b], Abbas Al Zain[a], Mushabab Al Shahrani[a], Howaida El Hakeem[c], Mubarak Al Shraim[d], Abdul Rahman M. Al Amri[e], M. Ezzedien Rabie[a,*]

[a] Department of Surgery, Armed Forces Hospital, Southern Region, Saudi Arabia
[b] Department of Radiology, Armed Forces Hospital, Southern Region, Saudi Arabia
[c] Department of Pathology Armed Forces Hospital, Southern Region, Saudi Arabia
[d] Department of Pathology, King Khalid University, Abha, Saudi Arabia
[e] Department of Surgery, Najran University, Najran, Saudi Arabia

ARTICLE INFO

Article history:
Received 1 July 2014
Accepted 14 August 2014
Available online 11 December 2014

Keywords:
Schwannoma
Retroperitoneum
Giant

ABSTRACT

INTRODUCTION: Schwannoma is the term given to tumours arising from Schwann cells of any nerve sheath. It may arise in the retroperitoneum, where it can attain enormous sizes and cause considerable diagnostic and therapeutic difficulties. A variety of incapacitating symptoms may arise, depending on its size alone and the related contagious organs.
PRESENTATION OF CASE: A 71-year-old female, who was incapacitated by a giant abdominal mass, associated with weight loss, immobility, general weakness and constipation. Radiologically, the presence of a huge pelviabdominal tumour was confirmed. A preoperative tissue diagnosis was entertained but omitted, and we resorted to direct surgical excision instead. During surgery, significant bleeding from the surrounding lumbar vessels was encountered, but it was controlled and the tumour was excised intact. Histopathologically, it showed the histologic features of ancient schwannoma.
DISCUSSION: In the patient presented here, who was rendered immobile by the tumour, total excision or at least debulking seemed appropriate, regardless of any biopsy result.
CONCLUSION: The diagnosis of retroperitoneal schwannoma and its variant "ancient schwannoma" should be considered when a huge pelviabdominal tumour is encountered. Although CT guided biopsy may be helpful in reaching a preoperative diagnosis, this might not change the decision for the need of total tumour excision or at least debulking, in the presence of incapacitating symptoms. With large tumours, the possibility of perioperative exanginating haemorrhage should be remembered and the necessary precautions activated.

1. Introduction

Schwannoma is a tumour which arises from Schwann cells in the nerve sheath. It is usually solitary and encapsulated, arising focally on the nerve, which becomes stretched over it. The sheath of any nerve may get affected and, rarely, the tumour may be found in the retroperitoneum where it can attain enormous dimensions. Small nerves of the skin or internal viscera may also be affected.[1] Ancient schwannoma is a rare subtype which exhibits certain radiologic and histologic features which are thought to occur with the passage of long time.

2. Case presentation

A 71-year-old female, presented with a slowly growing abdominal mass, associated with chronic constipation and pain, which she first observed 10 years ago. Gradually, by virtue of the weight of the mass, her mobility became limited. Relatively recently, she also suffered weight loss, vomiting, and lassitude. Her past history was otherwise irrelevant.

On examination, she looked cachectic but her vital signs were normal. Abdominal examination revealed a distended abdomen with a large 30 cm × 30 cm, firm, non-tender mass, occupying almost the entire abdomen.

Her blood picture and laboratory investigations as well as her abdominal X ray were all normal.

Abdominal ultrasound (US) and computerized tomography (CT) scan showed an encapsulated hypodense solid mass with cystic components and areas of enhancement, extending from the

* Corresponding author at: Department of Surgery, Armed Forces Hospital, Southern Region, PO Box 101, Khamis Mushait, Saudi Arabia. Tel.: +966 50775 3692.
E-mail address: ezzedien@hotmail.com (M. Ezzedien Rabie).

http://dx.doi.org/10.1016/j.ijscr.2014.08.015

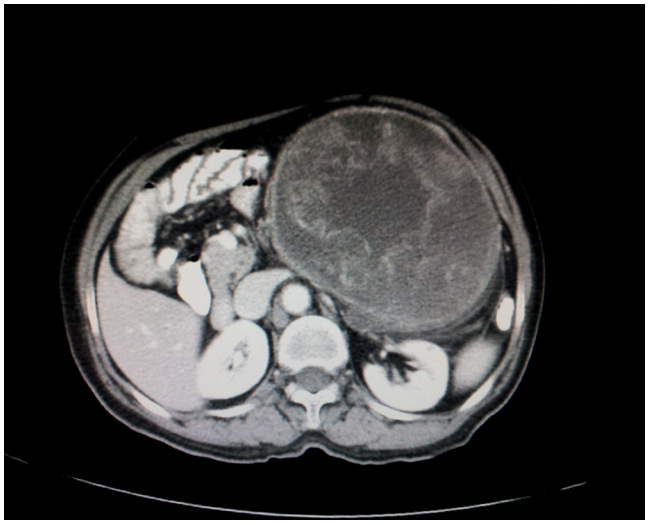

Fig. 1. CT scan showing heterogenous mass with cystic spaces and a well developed capsule.

epigastrium just below the tail of the pancreas, down to the pelvis, displacing the descending colon laterally with no adherence of contagious structures (Fig. 1). On magnetic resonance imaging (MRI), the mass showed heterogeneous hyperintense signals in T2 weighted images

Percutaneous needle biopsy was entertained, but omitted to avoid likely injury to an overlying or nearby vascular structure. As there was a definite capsule seen on the CT scan and owing to the marked symptoms the patient had, a decision was taken to explore the abdomen with the intention to excise the mass en toto or, if excision is unfeasible, to obtain an incisional biopsy while debulking the tumour.

At laparotomy, a huge retroperitoneal mass was seen, extending from just below the pancreas down into the pelvis (Fig. 2). The posterior peritoneum was opened over the mass and the left colon was mobilized to expose its anterior surface. Meticulous sharp and blunt dissection managed to release the mass from the adherent surrounding structures.

Significant bleeding was encountered from the surrounding lumbar vessels and was controlled by suture ligation.

Finally, the mass was excised intact within its fibrous capsule. It measured 29 cm × 30 cm × 13 cm, and weighed 3200 kg (Fig. 3).

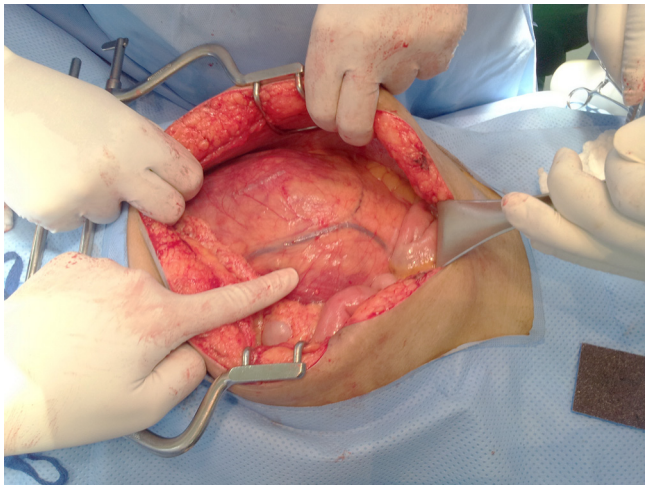

Fig. 2. Intraoperative view of the swelling with the overlying dilated vessels.

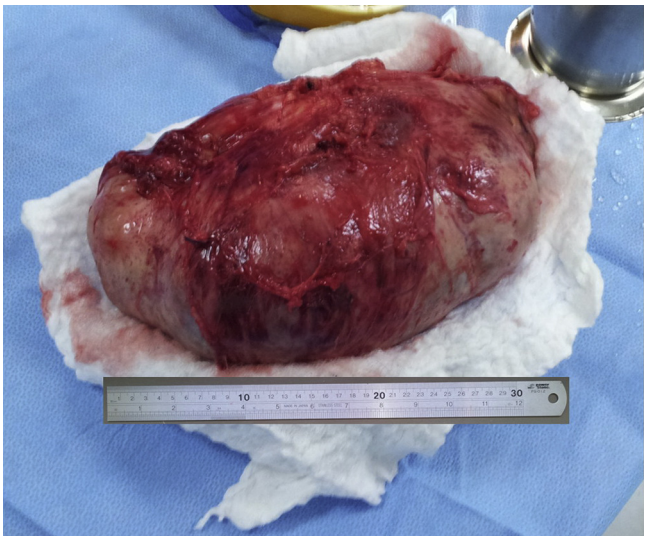

Fig. 3. The intact swelling measuring 29 cm × 30 cm × 13 cm.

Haemostasis was achieved with mass closure of the abdomen after suction drain insertion.

Postoperatively, the patient tolerated surgery well and was introduced to regular diet in time. In the third post-operative day, the drain was removed. Two weeks later, she appeared in the outpatient clinic in good condition.

Microscopically, there was palisade arrangement of spindle cells with extensive oedema of the matrix and the tumour tested positive to S-100 stain (Fig. 4A and B), features characteristic of ancient schwannoma.

3. Discussion

The discovery of a huge abdominal mass while investigating patient's symptoms, generates much interest regarding its nature management.

The term schwannoma was coined by Masson in 1932[2] to denote tumours which arise from Schwann cells in the nerve sheath. Microscopically, the tumour is composed of elongated cells, arranged in a palisade fashion and taking one of two patterns. In Antoni type A, the cells are arranged in an organized compact pattern. In Antoni type B, the cells are scattered loosely in an oedematous matrix. Both patterns may coexist in the same tumour and malignant changes are rare.[1] Ancient schwannoma, a term which was first utilized by Ackerman and Ackerman and Taylor in 1951, is a subtype characterized by hypocellular and degenerative areas which is believed to occur with the long term progression of the tumour.[3]

Although ancient Schwannoma may exhibit certain radiologic features,[4–6] which enables a tentative preoperative diagnosis to be reached at times,[7] none of them is specific. For this reason, the importance of preoperative needle biopsy has been stressed.[8] Moreover, the presence of heterogeneity and areas of degeneration on CT or MRI, may raise the possibility of malignancy, an additional reason behind the advisability of obtaining tissue diagnosis before embarking on surgical resection.[8]

The radiologic features of the tumour presented here were concordant with the recognized features of ancient schwannoma,[4–7] but, as is usually the case, the diagnosis was not considered preoperatively.

Obtaining a preoperative tissue biopsy may be problematic. In this regard, CT guided needle biopsy has proved its utility.[8] On the other hand, the role of fine needle aspiration cytology (FNA) is not well established,[9] as cellular pleomorphism in degenerated areas

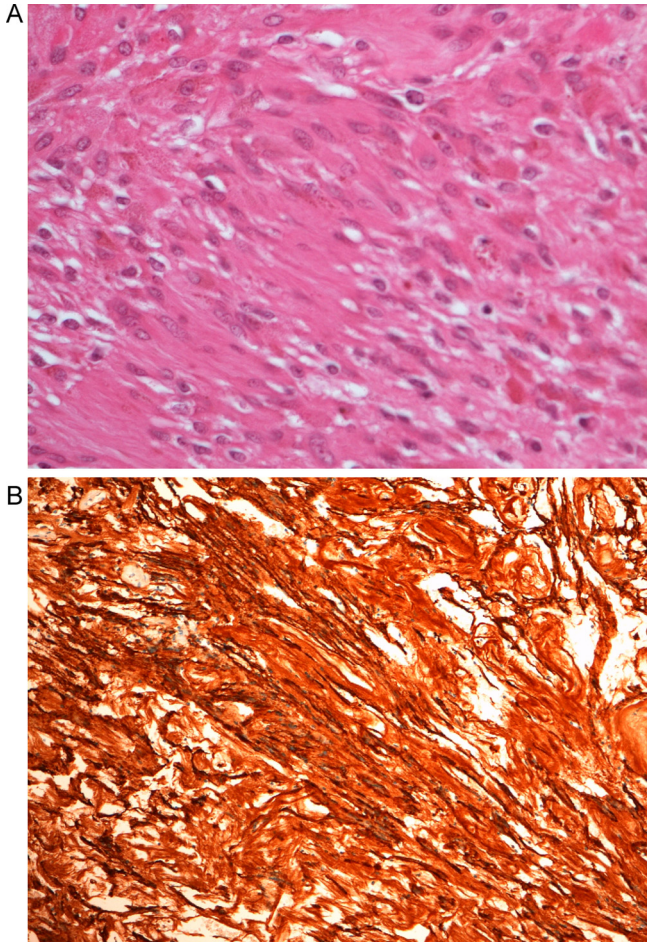

Fig. 4. A: Microphotograph showing Antoni A region with pallisade arrangement of cells (H&E ×100). B: On immunohistochemistry, the tumour tested positive for S-100 protein.

can be misinterpreted as malignancy.[10] In conditions where less invasive biopsy techniques fail, staged excision with preoperative incision biopsy was advocated by some (Ohene-Yeboah [11]).

However, we opted to omit a preoperative histologic diagnosis for several reasons. Firstly, such enormous tumours may harbour large vessels, either on the surface or within the tumour substance. Secondly, because almost all of the patient's symptoms were caused by the tumour bulk, its resection or at least debulking was necessary to alleviate the patient's symptoms. Additionally, despite the radiologically demonstrated heterogeneity, which raised the possibility of malignancy, the tumour demonstrated an intact smooth capsule, with no involvement of the surrounding organs. This gave some evidence of its benign nature and the feasibility of a safe dissection. Moreover, in our experience as well as the experience of others,[12] there is gross radiologic similarity between many of these intra abdominal tumours of different pathological nature, notably between schwannoma and gastrointestinal stromal tumours (GIST) (Fig. 5). Percutaneous biopsy, which may be the only practical way for preoperative tissue harvesting, has been discouraged in GIST.[13] In this context, one stage resection without preoperative biopsy has also been previously reported.[14]

Complete excision should be the aim. If not feasible, partial or subtotal resection should suffice, to avoid injuring contingent organs or neurovascular structures, although tumour recurrence may be inevitable in such cases.[15,16] In this regard, however, recurrence several years after a seemingly radical excision of a benign schwannoma has also been reported.[17]

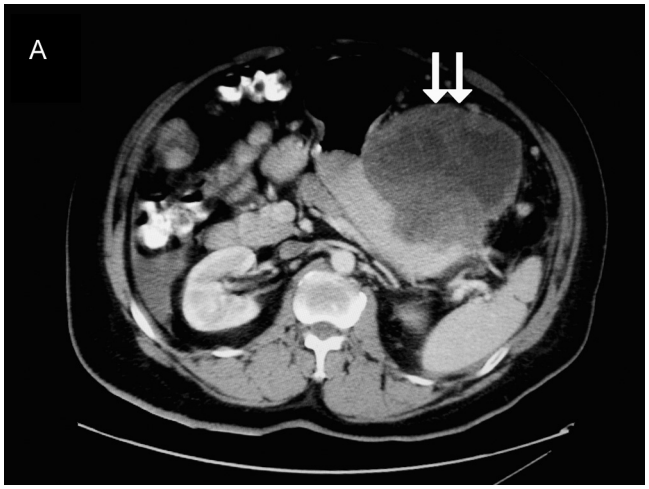

Fig. 5. GIST radiologic appearance is not dissimilar from that of schwannoma. Compare to Fig. 1. (First authors photo archive).

During tumour dissection, extensive bleeding may be encountered from the surrounding vessels. This may necessitate massive transfusion of blood and its products. Packing proved to be extremely beneficial under these circumstances, with the pack removed after 24–48 h.[8] These facts should be considered before contemplating surgery, and appropriate measures taken. In the patient presented here, excessive bleeding was encountered from the neighbouring lumbar vessels but, fortunately, it was secured with relative ease.

Despite our success in excising the intact tumour, demonstrating the nerve of origin was not possible, which is usually the case in such conditions.[8]

In our Pubmed search, utilizing the terms retroperitoneal ancient schwannoma in the title and abstract, we were able to retrieve only 9 reports, which reflects the extreme rarity of this condition.

4. Conclusion

The diagnosis of retroperitoneal schwannoma and its variant "ancient schwannoma" should be considered when a huge pelvi-abdominal tumour is encountered. Although CT guided biopsy may be helpful in reaching a preoperative diagnosis, this might not change the decision for the need of total tumour excision or at least debulking, in the presence of incapacitating symptoms. With large tumours, the possibility of perioperative exanginating haemorrhage should be remembered and the necessary precautions activated.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

The medical research ethics committee has approved the publication of this report, under the code AFHSRMREC/2014/General Surgery Department/018 in June 24, 2014.

Conflict of interest

The authors have no conflict of interest to disclose.

References

1. Walter JB, Talbot IC. *Structure and effects of some common neoplasms. Walter and Israel general pathology*. 7th ed New York: Churchill Livingstone; 1996. p. 435–70 (Chapter 26).
2. Masson P. Experimental and spontaneous schwannoma. *Am J Path* 1932;**8**:367; Foote MN, Luongo V, Marino ER. Benign giant retroperitoneal neurilemmoma. *Ann Surg* 1963;**157**(5):719–24.
3. Ackerman LV, Taylor FH. Neurogenous tumors within the thorax: a clinicopathological evaluation of forty-eight cases. *Cancer* 1951;**4**:669–91; Isobe K, Shimizu T, Akahane T, Kato H. Imaging of ancient schwannoma. *Am J Roentgenol* 2004;**183**(2):331–6.
4. Wong C, Chu TY, Tam KF. Retroperitoneal schwannoma: a common tumour in an uncommon site. *Hong Kong Med J* 2010;**16**:66–8.
5. Beaman FD, Kransdorf MJ, Menke DM. Schwannoma: radiologic pathologic coorelation. *Radiographics* 2004;**24**:1477–81.
6. Beaman FD, Kransdorf MJ, Andrews TR, Murphey MD, Arcara LK, Keeling JH. Superificialsot-tissue masses: analysis, diagnosis, and differential consideration. *Radiographics* 2007;**27**:509–23.
7. Isobe K, Shimizu T, Akahane T, Kato H. Imaging of ancient schwannoma. *Am J Roentgenol* 2004;**183**(2):331–6.
8. Schindler OS, Dixon JH, Case P. Retroperitoneal giant schwannomas: report on two cases and review of the literature. *J Orthop Surg (Hong Kong)* 2002;**10**(1):77–84.
9. Sofia L, Currò G, Iapichino G, Melita G, Lorenzini C, Cucinotta E. Retroperitoneal giant schwannoma: a case report and review of the literature. *J Chir Ital* 2008;**60**(1):141–6.
10. Daneshmand S, Youssefzadeh D, Chamie K, Boswell W, Wu N, Stein JP, et al. Benign retroperitoneal schwannoma: a case series and review of the literature. *Urology* 2003;**62**:993–7.
11. Ohene-Yeboah M, Adjei E, Stalsberg HA. Giant gutealschwannoma with extension into the pelvis: a case report. *West Afr J Med* 2009;**28**(3):185–8.
12. Choi JW, Choi D, Kim KM, Sohn TS, Lee JH, Kim HJ, et al. Small submucosal tumors of the stomach: differentiation of gastric schwannoma from gastrointestinal stromal tumor with CT. *Korean J Radiol* 2012;**13**(4):425–33, http://dx.doi.org/10.3348/kjr.2012.13.4.425.
13. Gastrointestinal Stromal Tumors Treatment (PDQ®)-National Cancer Inst. http://www.cancer.gov/cancertopics/pdq/treatment/gist/HealthProfessionls (last modified 07.09.2011).
14. Hettiarachchi JA, Finkelstein MP, Schwartz AM, Johnson GB, Konno S, Choudhury MS. Benign retroperitoneal schwannoma presenting as a giant adrenal tumor. *Urol Int* 2003;**71**(2):231–2.
15. Pongsthorn C, Ozawa H, Aizawa T, Kusakabe T, Nakamura T, Itoi E. Giant sacral schwannoma: a report of six cases. *Ups J Med Sci* 2010;**115**(2):146–52.
16. Foote MN, Luongo V, Marino ER. Benign giant retroperitoneal neurilemmoma. *Ann Surg* 1963;**157**(5):719–24.
17. Bunc G, Kramberger S, Kovacic S, Seruga T, Vorsic M. Recurrent giant invasive thoracolumbar schwannoma. *Wien Klin Wochenschr* 2004;**116**(Suppl. 2):93–6.