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## Giant retroperitoneal ancient schwannoma: Is preoperative biopsy always mandatory?



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## 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 contiguous 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 exsanguinating haemorrhage should be remembered and the necessary precautions activated.

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## 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.<sup>1</sup> 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

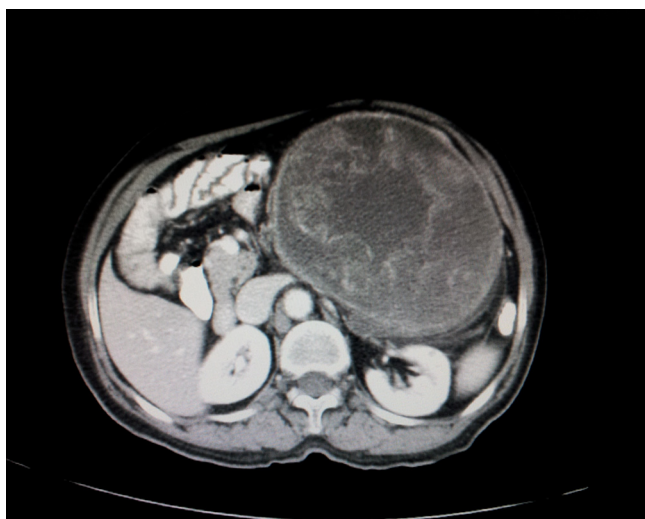
A 71-year-old female, presented with a slowly growing abdominal mass, associated with chronic constipation and pain, which she first observed 10 years ago. Gradually, by virtue of the weight of the mass, her mobility became limited. Relatively recently, she also suffered weight loss, vomiting, and lassitude.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

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

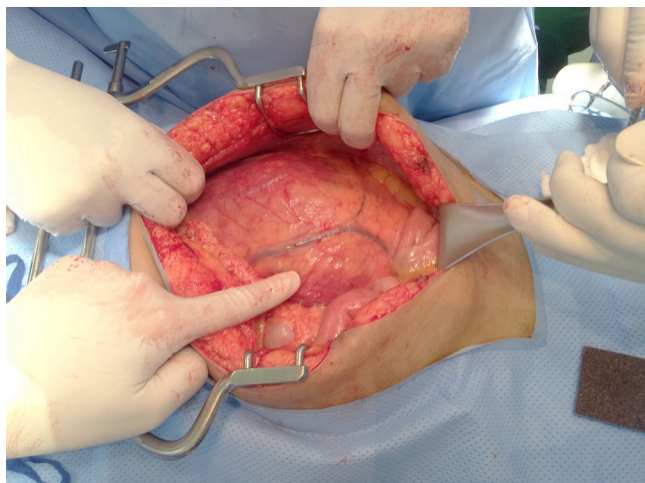
epigastrium just below the tail of the pancreas, down to the pelvis, displacing the descending colon laterally with no adherence of contiguous structures (Fig. 1). On magnetic resonance imaging (MRI), the mass showed heterogeneous hyperintense signals in T2 weighted images.

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 out-patient 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<sup>2</sup> 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.<sup>1</sup> 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.<sup>3</sup>

Although ancient Schwannoma may exhibit certain radiologic features,<sup>4–6</sup> which enables a tentative preoperative diagnosis to be reached at times,<sup>7</sup> none of them is specific. For this reason, the importance of preoperative needle biopsy has been stressed.<sup>8</sup> 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.<sup>8</sup>

The radiologic features of the tumour presented here were concordant with the recognized features of ancient schwannoma,<sup>4–7</sup> 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.<sup>8</sup> On the other hand, the role of fine needle aspiration cytology (FNA) is not well established,<sup>9</sup> as cellular pleomorphism in degenerated areas

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