

## Case Report

## Retroperitoneal Giant Ancient Schwannoma

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## Abstract

Ancient schwannoma is a rare variant of benign peripheral nerve sheath tumor, often morphologically mimicking malignancy. Retroperitoneum is an uncommon location. We describe one case of giant retroperitoneal ancient schwannoma. Literature concerning this variant is also reviewed. A 65 year old male presented with altered bowel habits. Imaging findings revealed a large heterogeneous mass in the retroperitoneum. The patient was treated with complete excision. Grossly, the tumor was predominantly solid with focal cystic degeneration. Histological examination showed an encapsulated spindle cell tumor with nuclear palisading, very focal nuclear atypia and widespread foam cell infiltration - findings consistent with ancient schwannoma. A diagnosis of ancient schwannoma can be considered for a solid-cystic encapsulated mass in the retroperitoneum. Malignant transformation is very rare. Local recurrence is uncommon following complete excision. The authors have nothing to disclose.

**Key words:** Ancient, giant, schwannoma, S100

**Cite this article as:** Khandakar B, Dey S, Chandra Paul P, Medda S, Bhattacharya A, Datta S. Retroperitoneal Giant Ancient Schwannoma. *Arch Iran Med.* 2014; 17(12): 847 – 849.

## Introduction

Schwannoma or neurilemmoma arises from Schwann cells - the supporting glial cells of peripheral nerves.<sup>1</sup> These slow growing benign tumors occur predominantly in females of 2<sup>nd</sup> to 5<sup>th</sup> decades.<sup>2</sup> Ancient schwannoma, a rare morphological variant with degenerative histological changes, often poses significant diagnostic difficulty by mimicking malignancy. Schwannomas are usually located intracranially, in association with the eighth cranial nerve, commonly known as acoustic neuroma, though tumors at other sites have also been reported.<sup>3</sup> Retroperitoneal location is rare, accounting for 0.3% to 3.2% of these tumors, often detected incidentally.<sup>4</sup> The average diameter of Schwannomas usually does not exceed 5 to 6 cm, although tumors measuring up to 28 cm have been described.<sup>5</sup> We are reporting one case of giant retroperitoneal ancient Schwannoma managed surgically in our institution.

## Case Report

A 65-year-old male presented to the surgery out-patient department with chief complaints of altered bowel habit and abdominal discomfort for six months. On examination, a non-tender deep-seated midline swelling was palpated in the hypogastrium. An ultrasound and CT scan of abdomen revealed a large ( $\approx 13 \times 11.5$  cm) well demarcated retroperitoneal heterogeneous solid mass compressing the rectum and bladder (Figure 1a). The patient subsequently underwent complete surgical excision of the mass. The

specimen was sent for histopathological examination. The post-operative period was uneventful and the patient was discharged on the 5<sup>th</sup> post-operative day.

## Histopathology

Grossly, the specimen was grey-white, encapsulated, oval, firm, measuring  $\approx 15 \times 10 \times 8$  cm. The cut surface of the tumor was yellowish, focally myxoid, soft to firm and predominantly solid. An area of cystic degeneration ( $6 \times 5 \times 4$  cm) was identified near the central part of the mass containing hemorrhagic fluid (Figure 1). On microscopy, the tumor was well circumscribed, moderately cellular with focal myxoid areas. The tumor cells were arranged in short fascicles, with nuclei organized in palisades at places (Figure 2a). The individual tumor cells displayed minimal pleomorphism with indistinct cell borders, moderately eosinophilic cytoplasm and spindle wavy vesicular nuclei with inconspicuous nucleoli (Figure 2b); however, rare bizarre tumor cell nucleus was also identified. Some areas showed extensive infiltration by inflammatory cells, comprising predominantly of foam cells and occasional siderophages (Figures 2c and 2d). Mitotic count was low ( $<1/10$  HPF). The histomorphological features suggested an initial diagnosis of benign peripheral nerve sheath tumor, Schwannoma, with degenerative features. Immunohistochemistry revealed diffuse S100 positivity in the tumor cells (Figures 2e and 2f). A final diagnosis of 'retroperitoneal giant ancient schwannoma' was offered. At nine months follow-up, the patient is doing fine without any complaints.

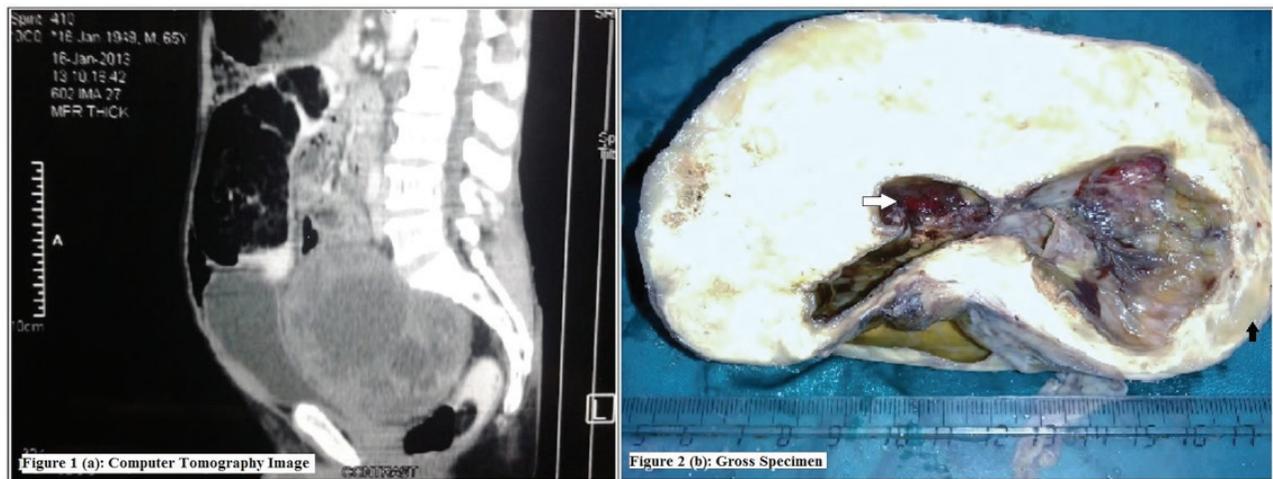
## Discussion

Schwannomas are characterized by their benign biological behavior; however, malignant cases have also been described. Computer tomography imaging of benign schwannoma reveals well delineated heterogeneous mass without any infiltration.<sup>6</sup> These

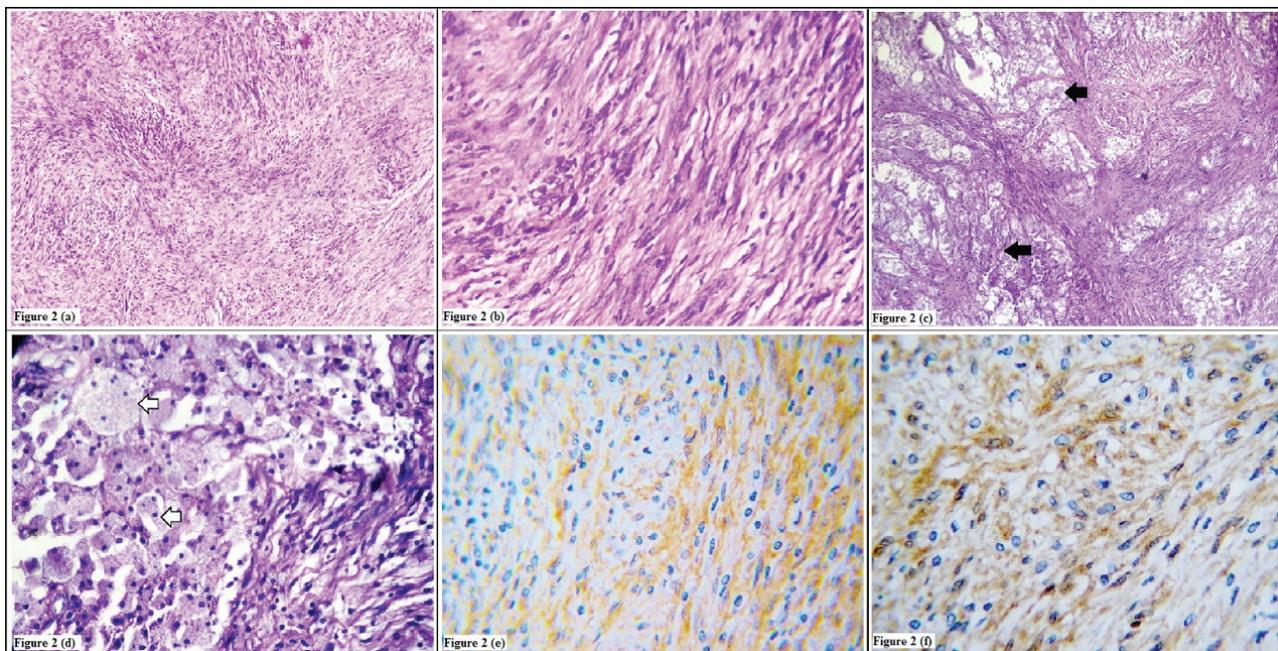
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Accepted for publication: 18 June 2014



**Figure 1. (a)** Contrast Enhanced Computer Tomography image showing large well demarcated heterogeneous solid mass mass compressing the bladder and rectum. **(b)** Cut surface of the tumor showing focal myxoid (black arrow) areas. Central part showing cystic cavity (white arrow).



**Figure 2. (a)** Tumor cells in short fascicles and nuclei arranged in palisades focally (Hematoxylin and Eosin stain, 100 $\times$ ). **(b)** Individual tumor cells have indistinct cell borders, eosinophilic cytoplasm and spindle wavy vesicular nuclei with inconspicuous nucleoli displaying minimal pleomorphism (Hematoxylin and Eosin stain, 400 $\times$ ). **(c)** Extensive infiltration by foam cells (black arrow) (Hematoxylin and Eosin stain, 100 $\times$ ). **(d)** Foam cells at higher magnification (white arrow) (Hematoxylin and Eosin stain, 400 $\times$ ). **(e) & (f)** Immunohistochemistry for S100 showing diffuse and strong S100 positivity in the tumor cells (Hematoxylin counter stain; 100 $\times$ ).

tumors commonly arise in head-neck region.<sup>7</sup> Retroperitoneum is a rare location and retroperitoneal schwannomas often attain a large dimension due to non-specific vague clinical symptoms leading to a delay in diagnosis. A definite preoperative diagnosis at this site is usually difficult, as schwannomas constitute only 4% of all tumors in the retroperitoneum.<sup>8</sup> Due to the absence of any pathognomonic radiological features, erroneous imaging diagnosis is not uncommon.<sup>6</sup> Use of preoperative fine-needle aspiration cytology (FNAC) in diagnosis is controversial. Preoperative true cut biopsy may be helpful in suspected malignant lesions.<sup>1</sup> Laparotomic excision is considered the treatment of choice, as most tumors are encapsulated, and non-invasive.<sup>3</sup>

On microscopy, schwannoma is characterized by alternating are-

as of hyper- (Antoni A) and hypo- (Antoni B) cellularity. Antoni A areas contain clusters of parallelly arranged spindle cells forming palisades, commonly known as Verocay's bodies. In the Antoni B areas, cells are loosely arranged in a myxoid background. The cells are typically immuno-positive for S-100, neurone specific enolase and vimentin, while immuno-negative for smooth muscle actins.<sup>1</sup>

A rare variety of peripheral nerve sheath tumor, Ancient schwannoma, often morphologically simulates malignancy but is in fact benign.<sup>1</sup> Ancient schwannomas are usually deep seated and can acquire a large size. Ackerman and Taylor were the first to introduce the term "ancient neurilemmoma". They reported long-standing schwannomas with less Antoni A areas and more hypo-

cellular areas.<sup>9</sup> Ancient schwannomas with areas of hyalinization, calcification, necrosis, hemorrhage, cystic and fatty degeneration, focal to extensive infiltration by siderophages and histiocytes have been described in literature. All these changes are thought to occur due to aging of the tumor. Sometimes such tumors are erroneously diagnosed as malignant because of nuclear atypia and pleomorphism.<sup>10</sup> In doubtful cases, mitotic count becomes crucial as it is always low in this variant. A proper diagnosis of malignant schwannoma is essential as they have a much worse prognosis and metastasize distantly. Combined clinical, radiological, and histological findings are helpful. Clinically, pain is the most important feature. These malignant ones have irregular infiltrating margins invading surrounding tissues. Histomorphologically, these show marked nuclear atypia and frequent mitoses.<sup>11</sup> Malignant schwannomas are treated aggressively with excision and adjuvant therapy.<sup>1</sup>

We reported a case of giant retroperitoneal ancient schwannoma treated by surgical excision. Ancient schwannoma is a rare variant of a common tumor and most cases are benign. A thorough search for mitosis is crucial. Local recurrence and malignant behavior is extremely rare.

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