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Case Report

Giant retroperitoneal schwannoma in a 52-year-old man

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ABSTRACT

Schwannoma in the retroperitoneal space is rare, and it is extremely rare in patients with no history of neurofibromatosis. We present a case of giant retroperitoneal schwannoma in a 52-year-old man who did not have neurofibromatosis. Because malignant transformation would be extremely rare in this circumstance, close imaging follow-up could avert the necessity for complete resection. The possibility of schwannoma should be considered when evaluating retroperitoneal tumors with the characteristic findings, even if there is no connection between the tumor and the intervertebral foramina.

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1. Introduction

Schwannomas are nerve sheath tumors that usually affect the head, neck, and the flexor surfaces of the extremities. However, schwannoma in the retroperitoneal cavity is rare, accounting for about 0.75%–2.6% of all schwannomas, and for 4% of all retroperitoneal tumor [1–3]. Histologically, schwannomas are distinguished by the presence of areas of high and low cellularity called Antoni A and Antoni B tissue, respectively [4]. Patients with retroperitoneal schwannomas usually do not

suffer from any symptoms, or exhibit nonspecific symptoms, such as abdominal pain, abdominal discomfort, constipation, and deep vein thrombosis [5]. The lack of specific symptoms sometimes makes it difficult to accurately diagnose preoperatively. Schwannomas are usually benign, and solitary malignant schwannomas are extremely rare in patients without any history of neurofibromatosis [6,7]. Considering the postoperative complications, it is sometimes very difficult to determine to treat this tumor [8]. Herein, we report a case

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of giant retroperitoneal schwannoma and review the relevant literature.

2. Case report

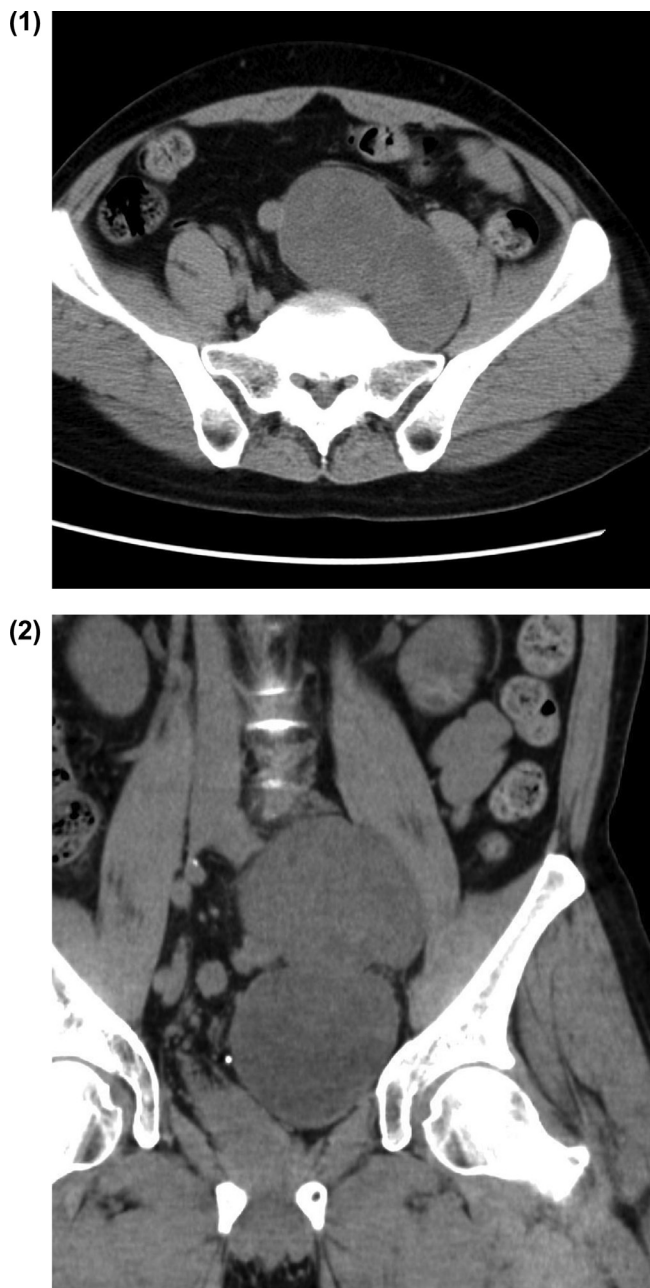
A 52-year-old male came to the hospital after a retroperitoneal tumor, and deviation of the bladder was found during a health examination. The patient had no complaints or symptoms, and there was no past medical history. Physical examination showed the temperature 36.8°C, pulse 86 bpm, respiration 16 bpm, and blood pressure 156/94 mmHg. A blood biochemical test and a tumor marker test showed no abnormalities: carcinoembryonic antigen 1.0 ng/ml, carbohydrate antigen 19-9 17.3 U/ml, soluble interleukin-2 receptor 279 U/ml, and neuron-specific enolase 13.9 ng/ml.

Computed tomography showed that the retroperitoneal tumor consisted of three circumscribed round and oval tumors extending craniocaudally. The tumor had slightly low density, compared with muscle, and an overall maximum diameter of 130 mm (Figs. 1 and 2). Magnetic resonance imaging (MRI) revealed that the density inside the tumor was heterogeneous and consisted of two compartments. First compartment had a heterogeneous high signal intensity on T2-weighted imaging, with restricted diffusion and strong enhancement indicating a solid compartment. The other compartment had high signal intensity on T2-weighted imaging with no enhancement, indicating a cystic compartment (Figs. 3-6). Ultrasonography showed a circumscribed hyper-echoic mass with several echo lucent areas inside the mass, indicating a cystic component (Fig. 7). We suspected that it was a neurogenic tumor with cystic degeneration, mainly suspecting schwannoma. However, it was difficult to determine the origin of the tumor because there was no connection between the tumor and the foramen intervertebral. Since it was large, and exhibited strong, restricted diffusion, we performed open biopsy, considering the potential of malignancy (Fig. 8). The pathologic diagnosis based upon hematoxylin and eosin staining showed two compartments with compact hypercellular Antoni A areas, and myxoid hypocellular Antoni B areas (Fig. 9). The immunohistopathologic findings were as follows: strongly positive for S-100 and negative for cytokeratin AE1/AE3, neurofilament, desmin, and α -smooth muscle actin (Fig. 10). The final pathologic diagnosis was schwannoma, and there was no malignancy. As there was no malignancy, and no evidence of other malignant tumors mimicking benign schwannoma, we chose to follow up the tumor closely.

3. Discussion

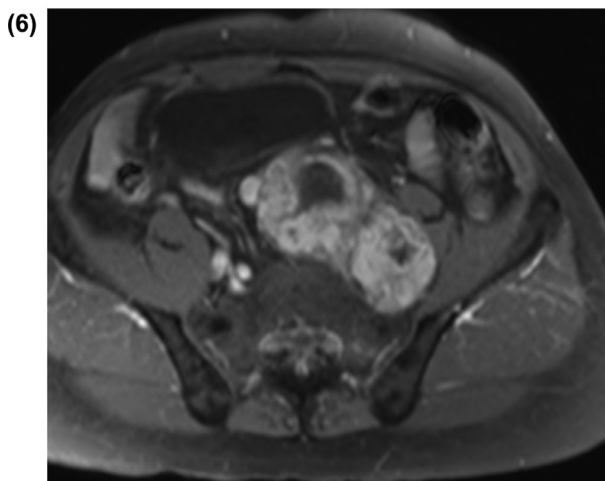
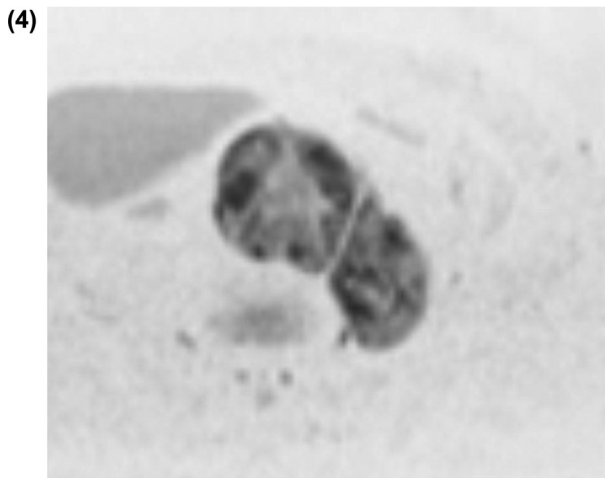
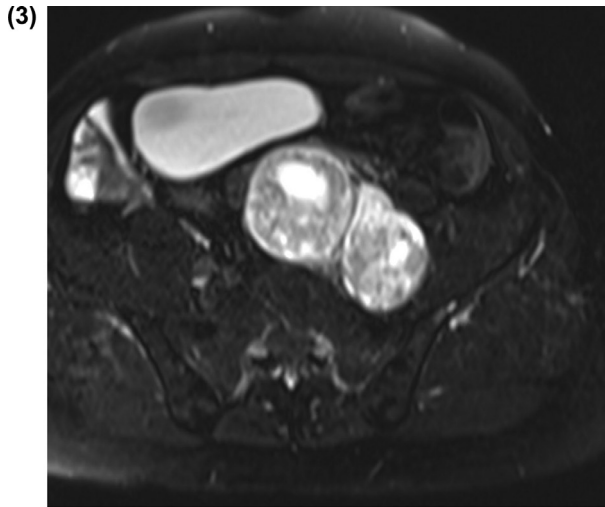
Primary retroperitoneal neoplasms are rare, and include tumors arising from major organs and/or arising beside major organs. If the tumor is huge, it is often difficult to even find the origin of the tumor, which can lead to misdiagnosis [9].

The findings that led us to suspect that the tumor was neurogenic are as follows: the tumor was a well-circumscribed spindle shape, extending along the nerve tracts, no calcifi-



Figs. 1-2 – Computed tomography revealed that the retroperitoneal tumor consisted of three circumscribed round and oval tumors, extending craniocaudally with a slightly low density compared with muscle, and a maximum diameter of 130 mm.

cation or subtle calcification, slightly low density compared with soft tissue on computed tomography because of the high density of myelin, including fat [10,11]. In reference to schwannoma in the retroperitoneal space, a connection between the tumor and foramen intervertebral is important to support the diagnosis, but a lack of evident connection is insufficient to deny the diagnosis of schwannoma [8]. The classical finding of schwannoma, called target sign, can be seen on MRI as a T2WI hyperintense rim surrounding a



Figs. 3-6 – Magnetic resonance imaging revealed that the tumor was heterogeneous, and that it consisted of two compartments: a compartment with heterogeneous, high signal intensity on T2-weighted imaging (Fig. 3) with restricted diffusion (Figs. 4 and 5) and strong enhancement (Fig. 6), indicating a solid compartment; and the other compartment with very high signal intensity on T2-weighted imaging with no enhancement, indicating a cystic compartment.



Fig. 7 – Ultrasonography showed a circumscribed hyperechoic mass with echo lucent inside the mass, indicating a cystic component.

central, low-signal area [12]. Fascicular sign characterized by multiple, small, ring-like structures with peripheral hyperintensity representing the fascicular bundles within the nerves is also seen on T2WI [11]. In our case, the shape and the size were round and oval, and the tumor roughly extended along the nerve tract, which supported the diagnosis of schwannoma. Typical findings, as “target signs” or “fascicular signs,” were not seen on MRI in our case.

While histological malignancy is also an important factor, in terms of malignancy, it must be taken into account that malignant schwannomas are extremely rare in patients without neurofibromatosis [6,7]. Findings on diffusion-weighted imaging have been reported to have the potential to differentiate pathologically malignant tumors from benign tumors. However, there have been very few reports concerning retroperitoneal schwannoma. In their report of diffusion-weighted imaging on peripheral nerve sheath tumors throughout the body, Demehri et al. reported that only minimum apparent diffusion coefficient (ADC) was useful [13]. However, contrary to this, Chhabra et al. reported that minimum ADC was not useful [14]. Kim et al. reported that texture analysis of ADC can be useful to diagnose malignancy of myxoid-containing tumors, such as schwannoma [14]. Demehri et al. also reported that bigger tumor, ill-defined or infiltrative margins, peritumoral edema, and early arterial enhancement are factors that can predict malignancy. However, all malignant peripheral nerve sheath tumors in their report occurred in patients with neurofibromatosis [13].

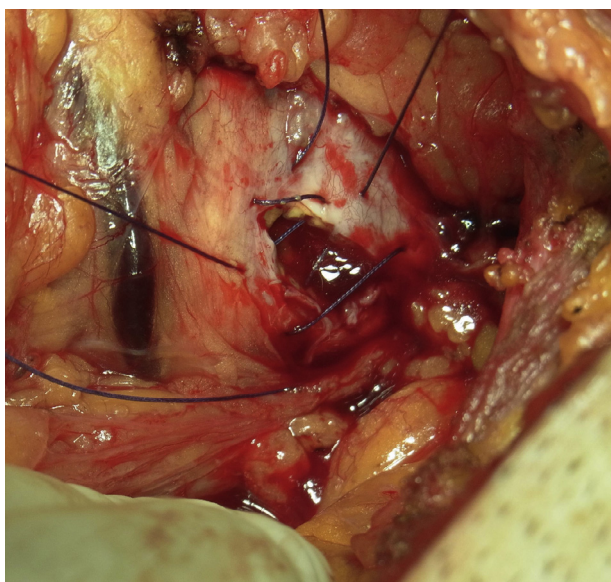


Fig. 8 – Open biopsy revealed a white circumscribed tumor, and leakage of mucinous liquid from inside, after cutting the tumor.

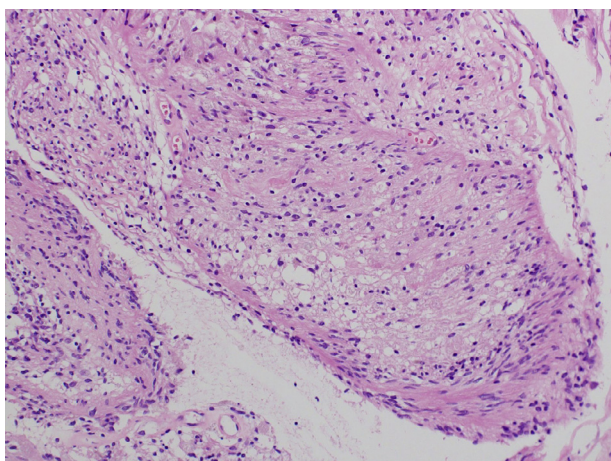


Fig. 9 – Pathologic diagnosis based upon hematoxylin and eosin staining showed two compartments: compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas.

Recurrences after resection are also known to be an unfavorable course of schwannoma. Schwannoma recurrence often occurs when it has malignant potential pathologically [15–17]. After confirming malignancy in pathologic sections, the recurrence rate after marginal excision is 72%, compared with 11.7% after wide surgical resection [18]. Benign schwannomas also have the potential for recurrence if the resection is incomplete and the rate has been reported to be 5%–10% [19]. Therefore, if there is the possibility of malignancy, a clinically wide surgical margin will be needed. When there is very little possibility of malignancy, marginal or partial resection is not recommended.

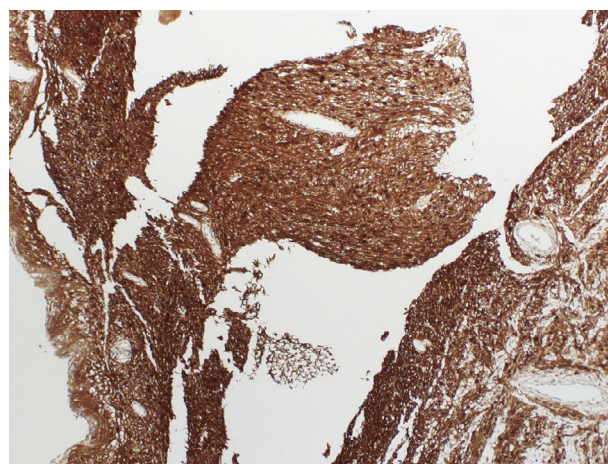


Fig. 10 – Strong expression of S-100 protein was observed.

Symptoms after surgery have also been investigated previously. Many reports have indicated that there are no residual symptoms. However, some reported no improvement or subtle improvement of symptoms after surgery [20–24], and some reported recovery of residual symptoms several months after surgery [25–27]. Since there appears to be no relationship between the type of operation, size of the tumor, and connection with lumbar vertebrae, this requires further investigation.

4. Conclusion

In conclusion, we experienced a case of giant retroperitoneal schwannoma. When the tumor is large, if the patient has neither a history of neurofibromatosis nor any evidence of malignancy, close follow-up may be a treatment choice.

Drug names and instrument names

MRI: 1.5 T Magnetom Avanto (Siemens, Erlangen, Germany).

Units

No alternative units.

Embedded math equations

No equations.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2018.04.021.

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