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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.

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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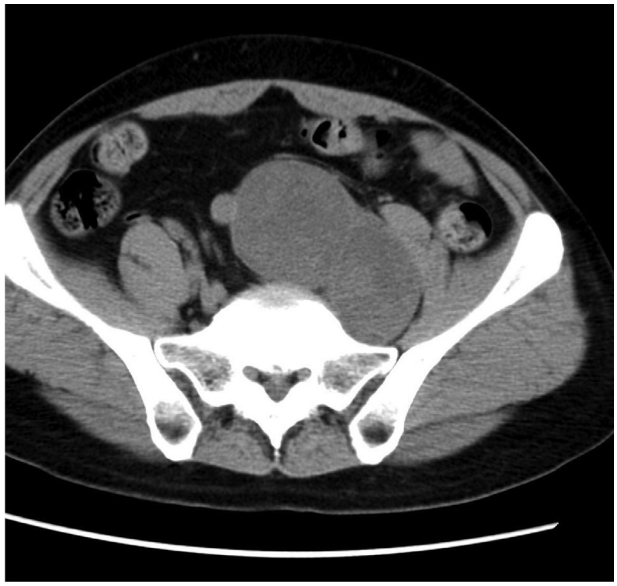
of giant retroperitoneal schwannoma and review the relevant literature.

2. Case report

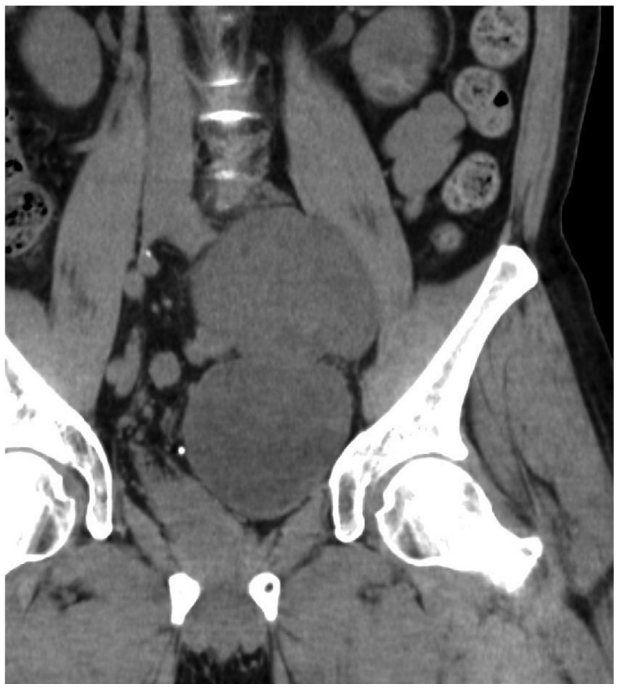
A 52-year-old male came to the hospital after a retroperitoneal tumor, and deviation of the bladder was found during a health examination. The patient had no complaints or symptoms, and there was no past medical history. Physical examination showed the temperature 36.8°C, pulse 86 bpm, respiration 16 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/m, 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 hyperechoic 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.

(1)



(2)



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.

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 calcification 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