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Case Research

Retroperitoneal schwannoma mimicking metastatic seminoma: case report and literature review

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Abstract

If a testicular cancer patient has a mass in the retroperitoneum, a metastasis is often the first suspicion, probably leading to improper diagnosis and overtreatment. Here we report a case of retroperitoneal schwannoma mimicking metastatic seminoma. A 29-year-old man, who had a history of seminoma, presented with a single retroperitoneal mass suspected to be a metastasis. Because the patient refused radiotherapy, 3 cycles of cisplatin, etoposide, and bleomycin were offered. Post-chemotherapy computed tomography scan revealed persistence of the retroperitoneal mass, with no change in tumor size or characteristics. Subsequently, retroperitoneal lymph node dissection was performed. The dissected tissue contained negative lymph nodes but a single mass in the attached fat. Pathology revealed retroperitoneal schwannoma, which was confirmed by immunohistochemistry. Thus, clinicians should be aware of retroperitoneal schwannoma and its distinction from metastatic seminoma to avoid misdiagnosis and ensure proper treatment.

Key words Retroperitoneal schwannoma, mimic, metastatic seminomatous tumor, retroperitoneal lymph node, retroperitoneal mass

Schwannoma is a benign neoplasm that originates from the Schwann cells of the peripheral nerve sheath. Most schwannomas occur in the head and neck or the extremities; rarely, they are seen in the retroperitoneal space. There have been occasional reports of retroperitoneal schwannoma mimicking metastatic seminoma[1]. Testicular germ cell tumors spread first to the retroperitoneal lymph nodes. When a patient with testicular cancer is found to have a mass in the retroperitoneum as shown on images, metastasis may be suspected initially. We report a patient with seminoma who had a mass in the retroperitoneum as shown on images. Radical orchiectomy was performed first. The pathologic diagnosis was seminoma limited to the testis and epididymis without vascular/lymphatic invasion; the clinical stage was IIB. The patient refused radiotherapy but underwent 3 cycles of cisplatin, etoposide, and bleomycin. Post-chemotherapy computed tomography (CT) scans revealed no change in the size or characteristics of the retroperitoneal mass. Retroperitoneal lymph node dissection was performed, and pathology and immunohistochemistry revealed retroperitoneal schwannoma. Awareness of this mimicking phenomenon is important because it can prevent erroneous diagnosis and ensure proper treatment.

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Case Report

A 29-year-old man presented with the right testis enlarged for 1 month without causing any discomfort. Physical examination revealed that the right testis

measured up to 6 cm × 5 cm × 4 cm, which was much bigger than the left testis. This patient's medical history was not unusual. Notably, his brother was diagnosed with seminoma 10 years ago but was still alive and in good health. CT of the chest and abdomen revealed a single abdominal mass (29 mm × 31 mm) located below the renal hilum along the para-aorta. The serum concentration of α-fetoprotein, human chorionic gonadotropin, and lactate dehydrogenase were normal. Right radical orchiectomy was performed. The section of the right testis contained a light white uncapsulated mass, measuring 5 cm ×4 cm. Microscope examination revealed seminoma limited to the testis and epididymis without vascular/lymphatic invasion, making the clinical stage IIB. The patient refused radiotherapy but underwent 3 cycles of cisplatin, etoposide, and bleomycin (PEB). Post-chemotherapy CT revealed persistence of the retroperitoneal mass, with no change in tumor size or characteristics (Figure 1).

Retroperitoneal lymph node dissection was performed, and the harvested tissue contained negative lymph nodes but a single mass in the attached fat. The mass was diagnosed pathologically as retroperitoneal schwannoma and confirmed as such with immunohistochemistry (positive for S-100 and CD56, negative for CD34 and CD117). Cell proliferation was evaluated with Ki-67 staining, and a low proliferation rate (Ki-67 < 5%) was reported, supporting the benign nature of the lesion. Six months after the initial surgery, the patient was alive and in good health without any evidence of relapse. Complications such as retrograde ejaculation, ascites, and intestinal obstruction were not observed.

Discussion

In the United States, estimated 8290 new cases of testicular cancer were diagnosed in 2011, with more than 60% being seminoma. Moreover, 350 men died of testicular cancer in 2011[2]. Both right-sided and left-sided testicular germ cell tumors spread first to the retro-

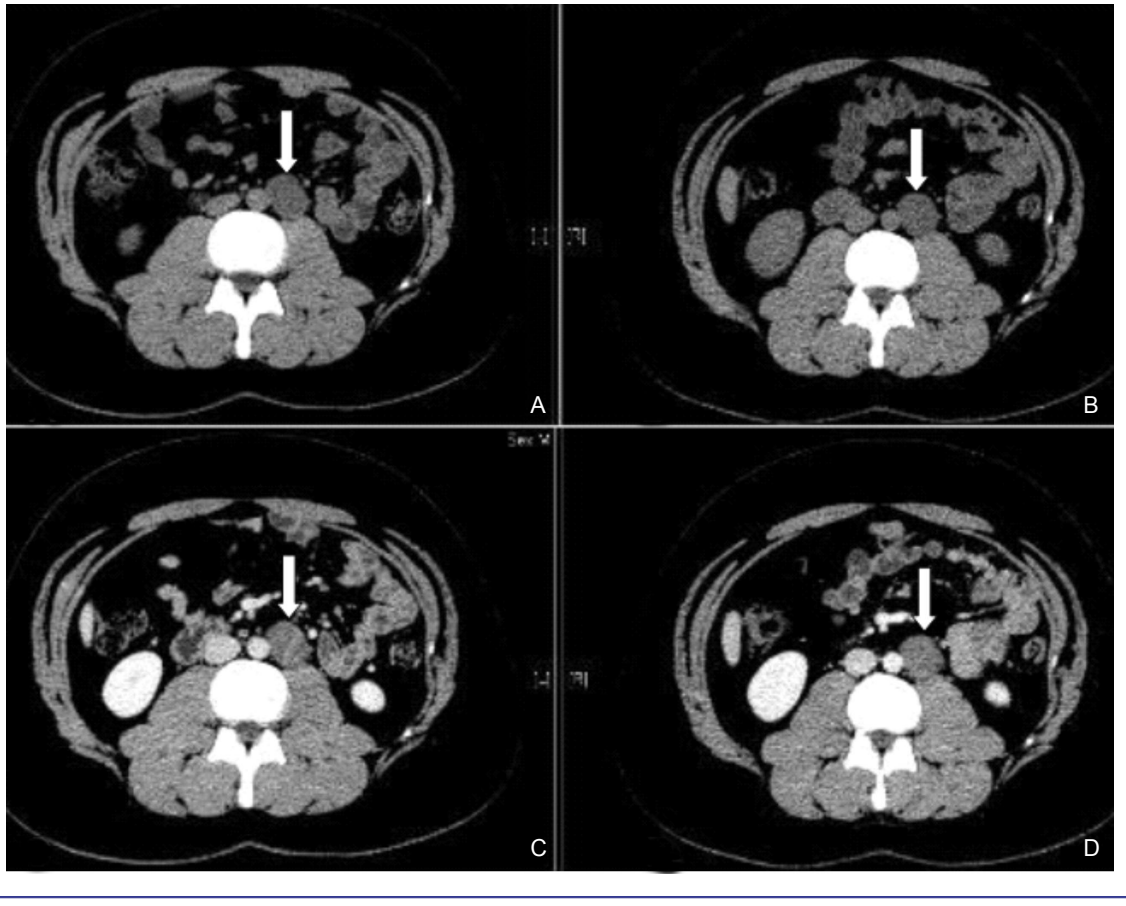

Figure 1. **Computed tomography (CT) of the tumor (denoted by arrow) before and after chemotherapy.** A and B, plain scans; C and D, contrast-enhanced CT scans; A and C, before chemotherapy; B and D, after chemotherapy. CT values: A, 20.1; B, 13.2; C, 26.6; D, 19.3.

peritoneal lymph nodes and then move superiorly along the thoracic duct. For patients with stage IA/IB seminoma, treatment comprises radiotherapy, chemotherapy, and surveillance. However, the first two options can potentially lead to morbidity, and surveillance is therefore the recommended treatment option. Three cycles of PEB therapy constitute over-treatment.

Schwannoma is a generally benign neoplasm arising from Schwann cells, which are found in peripheral nerve sheaths derived from the neuroectoderm. It can occur in any peripheral nerve sheaths except for those of cranial nerves I and II, where Schwann cells are absent. The retroperitoneal space is flexible; thus, the symptoms are due to organ displacement and are nonspecific. Schwannoma is usually discovered incidentally during investigation for unrelated symptoms, as in the case reported here.

To the best of our knowledge, there have been 5 reports (7 cases) of retroperitoneal masses mimicking metastatic testicular cancer in the English language literature [1,3-6]: 4 paragangliomas simulated metastatic non-seminoma (mixed germ cell tumors); 1 schwannoma simulated metastatic non-seminoma (positive teratomatous element); 1 subdiaphragmatic pulmonary sequestration mimicked metastatic seminoma; and 1 schwannoma mimicked metastatic seminoma, as in our case.

In these previous cases, the retroperitoneal masses were not located in the interaortocaval region below the level of the right renal vein. In one case reported by Xiao *et al.*[6], a mass was found in the anterior aspect of the right external iliac artery and aorta and in the paraportal/para-aortic area. The authors also reported another case, in which the mass extended from the bilateral iliac to the para-aortic area. Kopecky *et al.*[5] observed subdiaphragmatic pulmonary sequestration in the suprarenal para-aortic area. Takashi *et al.*[1] reported a case in which retroperitoneal schwannoma mimicked lymph node metastasis of seminoma. The schwannoma was located in the suprahilar interaortocaval region.

In the above 4 cases, the location of the retroperitoneal masses varied but none was found in the interaortocaval region below the level of the right renal vein, which is the initial location for metastasis from cancer of the right testis[7]. In our case, the retroperitoneal mass was below the renal hilum in the para-aortic area. Therefore, the locations of the primary tumor and retroperitoneal masses are useful indicators for differentiating lymph node metastasis of testicular cancer from primary tumors of other origins.

None of the four cases above responded to chemotherapy or radiotherapy[1,4-6], as was our patient. One report did not document this information[3]. In all 7 cases, again as in our case, there were no abnormal markers. These observations may help us avoid making incorrect diagnoses[1].

Preoperative diagnosis based on clinical examination is difficult because the symptoms are vague or nonspecific or the patient is asymptomatic. Imaging modalities such as CT and magnetic resonance imaging may be more helpful than ultrasonography and may be used to target tissue biopsy. CT- or ultrasonography-guided fine-needle aspiration for the preoperative diagnosis of schwannoma has been reported, but the accuracy of both biopsy targeting and diagnosis was poor[8]. A recent study demonstrated that use of endoscopic ultrasonography-fine needle aspiration to establish the diagnosis may help prevent unnecessary surgery[9]. However, preoperative biopsy carries a risk of disseminating tumor through needle-track seeding and is not widely used. Furthermore, it is easy to misdiagnose retroperitoneal masses as metastatic testicular cancer and especially to misdiagnose a retroperitoneal schwannoma as right-sided seminoma.

The present case report emphasizes that clinicians should be aware of retroperitoneal schwannoma and its distinction from metastatic seminoma to avoid misdiagnosis and ensure proper treatment. Unresponsiveness to chemotherapy or radiotherapy, as well as the location of the primary tumor and retroperitoneal masses might be useful indicators.

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Meeting Description

The Biennial Pathology Symposium of M. D. Anderson Cancer Center-Chinese Sister Institutions was first held in partnership with MD Anderson's sister institute, Fudan University Shanghai Cancer Center in Shanghai in 2005. Pathology of the 21st Century (P21) is an international program at MD Anderson that has been held six times, in the United States and elsewhere. P21 emphasizes the use of innovative diagnostic tools for neoplastic disorders and new neoplastic entities and classification systems. As one of MD Anderson's sister institutes in China, Sun Yat-sen University Cancer Center is honored to host this year's combined pathology and P21 conference in China.

The main theme of this conference is novel approaches to the pathologic diagnosis and classification of tumors. The conference topics will cover a wide spectrum of neoplastic diseases, such as gynecologic tumors, gliomas, thyroid neoplasia, melanocytic lesions, neuroendocrine tumors, colorectal cancer, and prostate cancer. Particular attention will be given to molecular and cytogenetic testing in pathologic diagnosis and personalized therapy. Relevant topics, such as the application of and approach to immunohistochemical analyses in tumor pathology and the training of pathologists in the 21st century, will also be presented.

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