

Oncology

Retroperitoneal extragonadal seminoma developed with acute lower inferior vena cava syndrome: A case report



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Introduction

Extragonadal germ cell tumors (EGCTs) are rare, accounts for approximately 5% of the germ cell tumors among male.¹ EGCTs often occur in the midline of the body and patients may present with various symptoms depending on the tumor locations and size. On several occasions, EGCT is difficult to diagnosis as a GCT without collecting a specimen. On the other hand, with a combination of chemotherapy and surgery, EGCTs are expected to be completely cured, though EGCT is more refractory than GCT occurring in testis. We present a young man with extragonadal seminoma developed with acute lower inferior vena cava (IVC) syndrome and complicated by a pulmonary embolism.

A case report

A 30-year-old man suffered from rapidly progressing left leg pain and edema for two weeks and was referred to our hospital. On physical examination, his left leg was swelling, and skin color turned dark red. On a blood test, lactate dehydrogenase (LDH) elevated (1082 IU/L) and also showed coagulation disorders (D-dimer rose to 18.2 µg/mL). Computed tomography (CT) scan showed a 65mm retroperitoneal tumor that invaded IVC and bilateral common iliac artery and vein (Fig. 1a and b). It also showed venous thrombus in the left iliac vein and right pelvic lymph node swelling. He was admitted to our hospital and performed an open biopsy, and the pathological examination revealed seminoma (Fig. 2). There was no evidence of testicular abnormality, and we diagnosed with retroperitoneal extragonadal seminoma.

His baseline serum LDH and human chorionic gonadotropin (hCG) levels increased abnormally (939 IU/L and 8898 mIU/mL, respectively), and alpha-fetoprotein (AFP) levels were normal (1.1 ng/mL) before to treatment. Immediately, he received anticancer chemotherapy with full dosage of bleomycin, etoposide, and cisplatin (BEP). On the

day 5 of 1st cycle, he developed sudden onset of dyspnea, and at the same time, his leg edema exacerbated. CT showed pulmonary embolism and massive residual leg thrombus (Fig. 1c), he was treated with a continuous infusion of heparin and IVC filter insertion.

Retroperitoneal tumor and thrombus drastically diminished on CT after four cycles of BEP chemotherapy. The fluorine-18 deoxyglucose positron-emission tomographic CT (FDG-PET/CT) and whole body Magnetic Resonance Imaging (MRI) could not reveal findings of viable tumors. At the same time, LDH and hCG levels decreased to within normal range (291 IU/L and 1.5 mIU/mL, respectively).

After a detailed explanation of treatment options for the patient: surgical resection or surveillance, the patient and we decided to keep under strict surveillance. Unfortunately, however, bilateral leg edema and back pain developed gradually, and three months after starting surveillance, CT showed a 45mm retroperitoneal tumor that located in the same area in the first diagnosis and liver tumors (Fig. 1d and e). He was diagnosed with recurrence and metastasis of seminoma and has been treated with paclitaxel, ifosfamide, and cisplatin (TIP) chemotherapy.

Discussion

EGCTs are reported to account for approximately 5% of GCTs in male, and among them, retroperitoneum is the second most common site of origin (13–45%) after mediastinum (35–54%).^{1,2} Therefore, retroperitoneal GCT is rare; however, it is necessary to always keep in mind as a differential diagnosis of retroperitoneal masses even with normal bilateral testes. Primary retroperitoneal masses originated within the retroperitoneal space encompass various types of neoplasms with often non-specific imaging findings.⁴ A previous study reported that abdominal pain is the most common symptom among retroperitoneal EGCTs (29%), following back pain (14%). Weight loss (9%),

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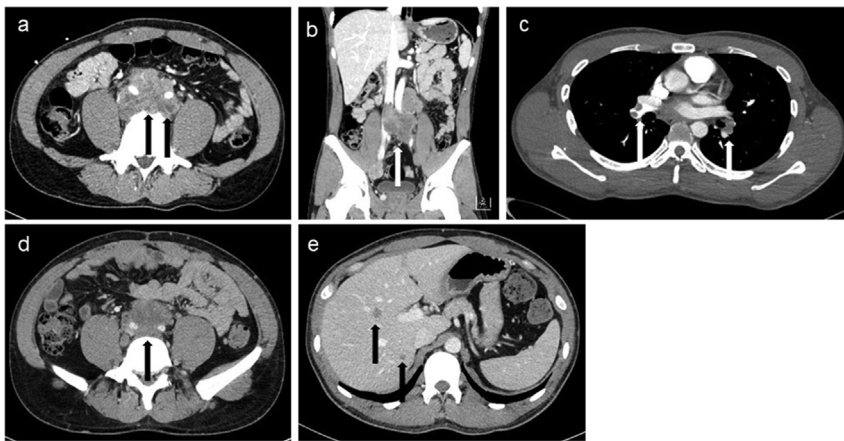


Fig. 1. Abdominal CT before treatment shows a 65mm retroperitoneal tumor that invasive IVC and bilateral common iliac artery and vein. (arrow) (a), axial CT image. (b), coronal CT image. Chest CT during the first course of the chemotherapy shows bilateral pulmonary embolism (c). Abdominal CT at the time of recurrence shows a 45 mm retroperitoneal tumor that located in the same area in the first diagnosis (d) and liver tumors (e).

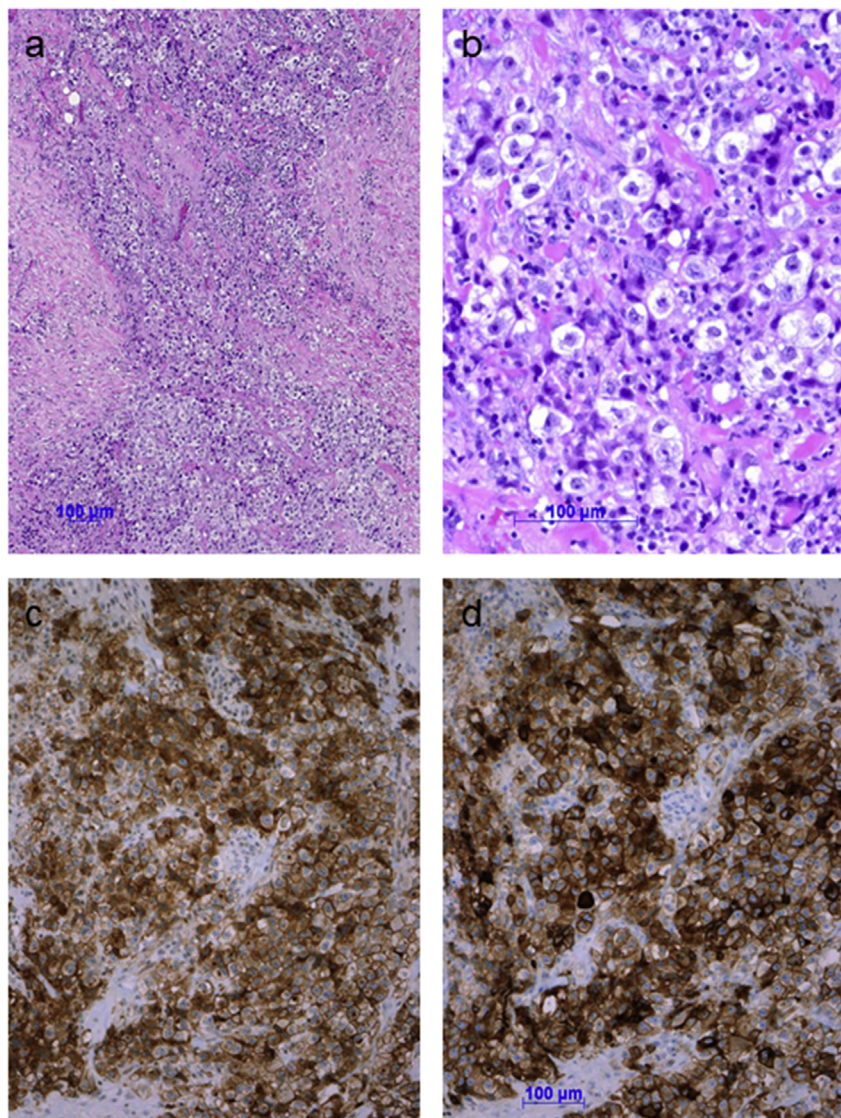


Fig. 2. Pathological findings. The biopsy specimen shows spheroidal cells with abundant granular to clear cytoplasm, a large centrally located nucleus and clumped chromatin pattern which are infiltrated by lymphocytes. (Haematoxylin & Eosin section) (a),(b). The biopsy specimen is positive for c-kit (c) and PLAP (d). These findings agree with seminoma.

venous thrombus (9%), fever/night sweats (8%), and a palpable abdominal mass (6%) also reported in rare cases.² Therefore, definitive diagnosis is difficult because of the variety of tumor sites with often non-specific imaging findings and symptoms. Tumor biopsy is necessary for definitive diagnosis of retroperitoneal masses. Thus, we performed tumor biopsy, and pathological examination showed seminoma in the present case. The baseline serum hCG levels were abnormally elevated (8898 mIU/mL) in this case. The cause of extremely high hCG level was uncertain; however, there was a possibility that a large number of syncytiotrophoblast cells existed or choriocarcinoma existed elsewhere other than the biopsy specimen.

Retroperitoneal tumors can give rise to IVC obstruction by compressing the IVC from the outside, by invading the venous wall, or by growing inside as tumor thrombus.³ The endovascular approach with stent placement for IVC obstruction is the treatment modality of choice for chronic IVC obstruction; however, reports on the interventional treatment of malignant inferior vena cava obstructions are rare.⁵ In the present case, the chief complaints were rapidly progressive leg edema and pain due to the IVC obstruction and thrombus, therefore, we didn't select an endovascular approach for the first time. Venous thromboembolism is a potentially lethal event. Anticoagulation is the cornerstone of treatment. IVC filters may be used in circumstances when anticoagulation is contraindicated or as an adjunct to anticoagulation. Accepted indications for IVC filters are in patients with acute venous thromboembolism and absolute contraindications to anticoagulation or in patients who have failed adequate anticoagulation.⁵ The increased use of IVC filters is not supported by high-quality evidence, and none of the international guidelines comment on the use of IVC filters in patients with malignancy despite being commonly used; however, the present case was treated with anticoagulation therapy and IVC filter insertion because of the pulmonary embolism with dyspnea and the massive residual leg thrombus. Patients with malignancy were said to be less likely to undergo IVC filters retrieval and had a reduced rate of retrieval success.⁵ The patient has also kept under surveillance without removing the IVC filter.

In view of the recurrence in a short period and the rapid growth of recurrent tumors during three months, intensive multidisciplinary therapy will be needed for the present case even though the pathological diagnosis was seminoma of well treatment responsive pathological

findings.

Conclusion

We reported a case of retroperitoneal seminoma developed with acute IVC syndrome. EGCTs are rare, however, should be considered in the differential diagnosis of retroperitoneal masses, particularly in the case of rapidly progressing its symptoms.

Conflicts of interest

The authors declare no conflict of interest in submitting this report.

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Appendix A. Supplementary data

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.eucr.2018.07.004>.

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