CLINICAL CASE

Somatic transformation in germ cell tumors: a two-case report

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KEYWORDS
Mature teratoma; Malignant transformation; Germ cell tumor metastasis

Abstract
Background: We describe herein 2 cases of germ cell tumors with somatic malignant transformation (SMT), a rare phenomenon with a low survival rate.

Case 1: A 53-year-old man had a one-month progression of pain and hardening in the left testis. He was diagnosed with a tumor and underwent radical orchiectomy (RO). The histopathology report stated: teratoma with SMT into neurogenic sarcoma and rhabdomyosarcoma. The patient developed retroperitoneal disease and was treated with chemotherapy. The retroperitoneal tumor progressed and grew and so surgical exploration was carried out, identifying an unresectable tumor. A biopsy was taken and the result was a high-grade fusiform rhabdomyosarcoma extending to the greater omentum.

Case 2: A 45-year-old man was a user of NSAIDs. He underwent RO and retroperitoneal tumorectomy in 2004 that reported germ cell tumor at both sites. He received chemotherapy and adjuvant radiotherapy. In 2014 he was referred to our institution and a PET-CT scan identified a 3 x 4 cm intercaval-aortic tumor. Tumor markers were normal. Tumorectomy of 80% of the tumor (R2) was performed, reporting fusocellular sarcoma. Three cycles of doxorubicin/ifosfamide and radiotherapy were administered. The patient remained under surveillance with a 16 mm intercaval-aortic tumor. He presented with upper gastrointestinal bleeding and died.

Conclusions: SMT of germ cell tumors is rare and insufficiently understood.

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Introduction

Somatic malignant transformation (SMT) refers to the presence of mixed germ cell tumors (GCTs) and their development into malignant non-germ cell tumors. It is a very rare phenomenon, characterized by resistance to chemotherapy and in which surgical resection is the only potentially curative method. Survival rates for patients with SMT are 50-60%.1-4

Case 1

A 53-year-old man with an unremarkable past medical history presented one month prior with an enlarged and hardened left testis. It was associated with pain in the left iliac fossa and ipsilateral lumbar area. A testicular ultrasound (US) study was done that showed a 9 x 9 cm testicular mass dependent on the left testis, with heterogeneous echogenicity and microcalcifications in the contralateral testis. After diagnosis, the patient underwent left radical orchiectomy (RO) and the histopathology study reported a 6.5 x 4 cm intermediate-grade fusocellular sarcoma with positive surgical margins. In December of 2014, the patient was given 3 cycles of doxorubicin, ifosfamide, and cisplatin (TIP), with poor response according to the RECIST criteria. He continued to be stable under palliative care and then died 18 months after the initial diagnosis.

Case 2

A 45-year-old man had a past history of smoking, alcoholism, NSAID use, and high blood pressure in treatment with a beta blocker. His illness began with the finding of a retroperitoneal tumor for which retroperitoneal tumorectomy and right RO were performed in May 2004, reporting germ cell tumor at both sites. He underwent chemotherapy (regimen not known) and adjuvant 40 GY radiotherapy in 25 fractions. In 2014 the patient began to experience pain in the mesogastrium and hypogastrum. He was referred to the INCAN where he had a PET/CT scan that identified a 3 x 4 cm intercaval-aortic tumor and a maximum SUV of 3.9 (fig. 3). The tumor markers from October 2014 were: alphafetoprotein 1.97 ng/ml, beta fraction human chorionic gonadotropin below 1 IU/ml, and lactate dehydrogenase 152 U/l.

In November 2014, 80% of the tumor (R2) adhered to the vena cava and aorta was removed through tumorectomy and the histopathology study reported a 6.5 x 4 cm intermediate-grade fusocellular sarcoma with positive surgical margins. In December of 2014, the patient was given 3 cycles of doxorubicin and ifosfamide and 58 GY radiotherapy in 19 fractions, ending in April 2015. In June 2015 control tomography showed partial response with a 16 mm intercaval-aortic residual tumor and it was decided to put the patient under surveillance. In July 2015 he had sudden onset of upper gastrointestinal bleeding and died due to hypovolemic shock.
Figure 1  Retroperitoneal tumor conditioning left renal ectasia

Figure 2  Increased tumor volume after failed chemotherapy.
Discussion

SMT is characterized by a high rate of disease progression and mortality. Given its rarity, pathogenesis, prognostic factors, and ideal management are still poorly defined.

Sarcoma is the most frequently reported histologic malignancy. Other histologic types described in SMT are: intestinal adenocarcinoma, primitive neuroectodermal tumors (PNET), and hematologic neoplasias.1

The histologic type of the tumor affects survival in these patients, but tumor grade is one of the most significant prognostic factors. Therefore when there are sarcomatous or sarcomatoid elements in the GCTs, it is important to assign tumor grade. Patients with high-grade sarcomatoid elements should receive stricter follow-up.5

The treatment of germ cell tumors with SMT can be a challenge because they are usually very difficult to resect and complex surgical resections are often necessary for ridding the patients of the disease.

In addition, these tumors have a propensity for local recurrence.

Because they are resistant to chemotherapy, surgical resection is the treatment of choice.

Conclusion

SMT of germ cell tumors is a rare phenomenon. It is essential to make the correct diagnosis so that optimum therapy can be provided, as well as administering treatment as soon as possible. Greater evidence and experience are required that support the new chemotherapy and other treatment options for improving disease prognosis, especially in its advanced stages.

Ethical responsibilities

Protection of persons and animals. The authors declare that the procedures followed conformed to the ethical standards of the responsible committee on human experimentation and were in accordance with the World Medical Association and the Declaration of Helsinki.

Data confidentiality. The authors declare that they have followed the protocols of their work center in relation to the publication of patient data.

Right to privacy and informed consent. The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

Financial disclosure

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Conflict of interest

The authors declare that there is no conflict of interest.

References


