

## Case Report

## Recurrence of Testicular Germ Cell Tumor as An Angiosarcoma: A Case Report and Review of the Literature

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### Abstract

Testicular germ cell tumor (TGCT) is the most common malignant tumor among young males. TGCT containing a teratoma component may show differentiation into various histopathological subtypes and may rarely recur as angiosarcoma. The present case had TGCT containing a teratoma component, and tumor recurrence occurred first as cartilage tissue and then as an angiosarcoma in the retroperitoneal area. After the diagnosis of angiosarcoma was established, the tumor was considered unresectable. The patient received chemotherapy and radiotherapy. The current study presents a rare case of TGCT, and the researchers also conducted a literature review.

### Introduction

Testicular germ cell tumor (TGCT) is the most common tumor type encountered in young males aged between 15-35 years. The tumor responds well to chemotherapy. The five-year overall survival rate is 96%. The cure rate is 80% even in the presence of metastatic disease (1, 2). Somatic malignant tumors such as carcinoma or sarcoma may occur in the primary tumor or in metastases from TGCT. Teratomas account for 5% to 10% of TGCT and may be resistant to the chemotherapy. Teratomas have the ability to differentiate into various types of tissues, and somatic malignant tumors arising in TGCT are mostly derived from the teratoma component of the tumor (3).

Angiosarcoma is one of the rare somatic malignancies arising in TGCT. The current case had TGCT containing a teratoma component that occurred first as cartilage tissue and then as an angiosarcoma in the retroperitoneal area.

### Case Report

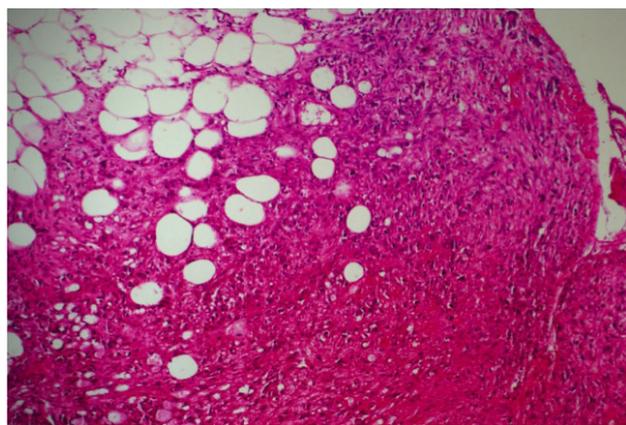
A 21-year-old male patient presented with painless swelling in the right testis. Scrotal ultrasonography revealed a tumor mass within the testis. The patient then underwent right radical inguinal orchiectomy. The

pathological examination showed “testicular mixed germ cell tumor (embryonal carcinoma, yolk sac tumor, teratoma)”. Abdominal CT showed two lymphadenopathies, the largest measuring 1.8 cm in diameter. The patient was diagnosed with a Stage IIA tumor, and he received three cycles of BEP (bleomycin, etoposide, cisplatin) chemotherapy. The patient was then put on follow-up.

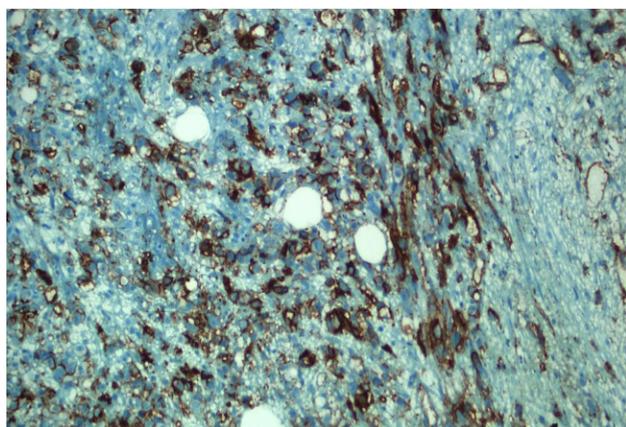
The patient developed a recurrent intra-abdominal tumor after a four year disease-free follow-up period. A 3x2 cm mass lesion was detected within the abdomen in the retroperitoneum in the left paraaortic area, and grade 2 hydronephrosis due to compression by the tumor mass was detected in the left kidney. The patient received three cycles of TIP (paclitaxel, cisplatin, ifosfamide) chemotherapy. The mass lesion within the abdomen showed shrinkage. The patient underwent retroperitoneal lymph node dissection. The pathological examination revealed “areas of mature and immature cartilage tissue (consistent with a teratoma compo-

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**Figure 1.** Light microscopy image of angiosarcoma (H&E stain, X100).



**Figure 2.** CD34 expression in the angiosarcoma tumor cells (CD34 stain, X200).

ment)”. The decision to continue follow-up was made.

Recurrence developed within the abdomen six months after. A 4x3 cm mass lesion was detected in the retroperitoneal area at the level of the L3-L5 vertebrae. The patient developed grade 3 hydronephrosis due to compression by the mass lesion. The patient reported pain radiating to the left leg. During the laparotomy, the mass lesion was deemed unresectable and only a biopsy was obtained from the mass lesion. The pathological examination was reported as “Malignant mesenchymal tumor (morphological and immunohistochemical findings consistent with Angiosarcoma)”. Palliative radiotherapy was applied to the area between the L3 and L5 vertebrae. The patient was administered with 6 cycles of GEMOX (gemcitabine, oxaliplatin) chemotherapy. The mass lesion in the abdomen showed regression after the therapy. The symptoms of the patient disappeared. The patient still continues attending follow-up visits at our clinic.

## Discussion

Primary or metastatic lesions of TGCT rarely may show differentiation into angiosarcoma. The association between TGCT and angiosarcoma was shown for the first time by Ulbright et al. in 1985 (4). Hughes and Steele presented cases with angiosarcoma arising from testicular teratoma in 1991 and 2000, respectively (5, 6). Lee et al. reported a case of testicular seminoma that was treated and later developed angiosarcoma (7). Sahoo et al. presented a case with angiosarcoma in the metastasis from mature testicular teratoma (8). In the present case, cartilage tissue first and then angiosarcoma developed arising from TGCT four years after the initial therapy. The present case differs from other reported cases with angiosarcoma due to recurrence of disease in two different forms.

The development of secondary somatic (non-germ cell) malignant component in germ cell tumors (GCT) is a known phenomenon and may occur in all GCTs located in the testicles, ovaries, mediastinum, or in the cranium (3, 9). Sarcomas are the most common somatic malignancy arising in GCTs (10). Various hypotheses have been postulated to explain the occurrence of sarcomas in GCTs. One of the postulated hypotheses is the malignant mesenchymal transformation of the teratoma component of GCT (9, 10). However, sarcomas have been observed in some germ cell tumors despite the absence of a teratoma component. True et al. reported five cases of spermatocytic seminoma that developed sarcoma despite the absence of teratoma component (11). Malagon et al. presented a case of pure seminoma that developed rhabdomyosarcoma (12). Aberrant differentiation of the primitive germ cells has been postulated to explain for this occurrence (11, 12).

Guo et al. examined 33 cases with TGCT containing a sarcoma component, and rhabdomyosarcoma (24 cases) was shown to be the most common histological subtype. This study reported only one case of angiosarcoma (13). In another study, Contreras et al. examined 12 cases with a mediastinal germ cell tumor containing angiosarcoma component, and teratoma was shown to be the most common component arising in GCTs. This study also showed that the angiosarcoma component in GCT was associated with poor prognosis (14). Idrees et al. provided evidence through genetic analysis that metastases from TGCT containing an angiosarcoma component are derived from the same clonal origin with the primary tumor (15).

GCTs differentiating into angiosarcoma are resistant to standard therapy and are therefore associated with poor prognosis. Surgical therapy remains the mainstay of treatment. Chemotherapy possesses limited efficacy. Of patients with mediastinal germ cell tumor containing angiosarcoma component in the study by Contreras et al., 80% showed metastasis, 20% showed local recurrence, and 70% died in approximately five years (14). The current case showed recurrence in the form of angiosarcoma, and the tumor was deemed unresectable. The patient underwent radiotherapy followed by rescue chemotherapy as administered in patients with testicular cancer; the patient received benefit from this therapy.

It should be kept in mind that TGCT containing a teratoma component may show recurrence in the form of angiosarcoma. Radiotherapy and chemotherapy may show efficiency in cases where surgery is not feasible.

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