

**PRODUCTION OF BETA-HCG BY SPERMATIC CORD LEIOMYOSARCOMA: A
PARANEOPLASTIC SYNDROME?**

A Case Report

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CASE REPORT

20 A 28-year-old man presented with a leiomyosarcoma of the spermatic cord after three weeks of increasingly painful swelling of the left upper hemiscrotum. The tumor was high-grade, Stage III pT2b N0 M0 (American Joint Committee on Cancer, 2002). A firm, nonreducible mass was palpable over the left upper hemiscrotum and distal inguinal canal. He denied any local trauma or unexpected weight loss and had no gynecomastia or vomiting. Four years before admission, he had

25 undergone ipsilateral inguinal hernia repair, during which no tumor mass was noted. An ultrasonographic examination of the scrotum was performed using a linear 7.0 MHz transducer. This revealed an extratesticular, hypoechoic mass-like lesion extending cranially into the left inguinal canal. Color Doppler ultrasonography showed minimal vascularity. The left epididymis and testis were normal. The patient underwent *en bloc* resection, revealing a solid, round tumor

30 measuring $6 \times 6 \times 4.5 \text{ cm}^3$, separate from the ipsilateral testis and epididymis. Pathology demonstrated a high-grade paratesticular leiomyosarcoma stemming from the spermatic cord (Figs 1, 2). Comprehensive studies did not show any metastases in the lungs, liver or lymphatic system. Definitive therapy with radical inguinal orchiectomy and wide excision of surrounding soft tissue confirmed a negative surgical margin of the initial *en bloc* resection.

35 Preoperative serum beta human chorionic gonadotropin (β -hCG) concentration was 43.88 ng/mL (immunoradiometric assay, CIS bio), whereas the postoperative β -hCG level decreased to

normal (less than 0.15 ng/mL). Immunohistochemistry for β -hCG (DAKO, 1:200) revealed focal staining (Fig. 3) in the resected tumor. Two courses of systemic chemotherapy with regimen of BEP (Bleomycin 30mg Day 2nd, 9th and 16th; Etoposide 100 mg/ M² Day 1~5; Cisplatin 20 mg/M² Day 1~5) were given as adjuvant therapy. Postoperatively, the patient has been free of recurrence or disease progression for seven months.

Most reports advocate radical inguinal orchiectomy with wide excision of the surrounding soft tissue as the standard treatment. Most distant metastases of spermatic cord leiomyosarcomas are either hematogenous to the lungs or liver, or pass via the lymphatic system to para-aortic lymph nodes. It appears that hematogenous metastasis is more frequent than lymphatic spread. Therefore, retroperitoneal lymph node dissection is not regularly recommended for patients with a spermatic cord leiomyosarcoma. Compared with surgery, the definite roles and therapeutic effects of adjuvant therapy, including chemotherapy and radiation therapy, are unknown. However, most physicians recommend radical surgery followed by radiation therapy or chemotherapy to achieve complete tumor regression. For this patient, we chose systemic chemotherapy rather than radiation therapy to abrogate any possibly hematogenous micrometastases after confirmation of a negative surgical margin. Cisplatin-based chemotherapy was used to treat the leiomyosarcoma, as there may also have been a syncytiotrophoblastic germ cell tumor with leiomyosarcoma differentiation.

Small intestine leiomyosarcomas that secrete β -hCG have been reported.³ The female

55 patient presented with “hyperemesis gravidarum”, a paraneoplastic syndrome caused by elevated level of serum β -hCG. HCG is a heterodimeric glycoprotein with a half-life of about 24–36 hours, assembled from two noncovalently linked α and β subunits which are encoded by two different genes. The α subunit of hCG is common to LH, FSH and TSH. The β subunit of hCG confers hormonal specificity and serves as a tumor marker in the diagnosis and management of germ-cell
60 tumors in men and of choriocarcinomas in women. Nongonadal cells producing β -hCG is associated with tumors of poor prognosis such as cancers of the lungs, pancreas, and liver. It may be secreted following metaplasia of the carcinomatous tissues into a tissue similar to poorly differentiated trophoblast cells.

Spermatic cord leiomyosarcomas with β -hCG secretion demonstrated by
65 immunohistochemistry stain have been reported in the German literature.⁴ In our patient, rapid normalization of β -hCG after tumor resection and positive immunohistochemistry stains of β -hCG in the resected tumor provide strong evidences of paraneoplastic secretion of β -hCG. To our knowledge, this has not been reported in the English medical literatures. Because β -hCG could be a paraneoplastic manifestation in spermatic cord leiomyosarcomas, we recommend serum β -hCG
70 as a biological marker to monitor and predict early recurrence in selective cases.

Acknowledgement

This study was supported in parts by C.Y. Foundation for Advancement of Education, Sciences

and Medicine.

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FIGURE LEGENDS

A: Photomicrography of tumor reveals round to spindle nuclei with moderate atypia. Two mitotic figures are identified (arrows). H & E, reduced from x 400.

B: Actin, reduced from x 400.

90 C: Immunoactivity of beta-hCG, reduced from x 400.

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