

## PARATESTICULAR LEIOMYOSARCOMA WITH ELEVATED SERUM BASIC FETOPROTEIN: A CASE REPORT

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A case of paratesticular leiomyosarcoma associated with elevated serum basic fetoprotein level is reported.

A 45-year-old male visited our hospital with a 6-month history of a painless left scrotal mass. High orchiectomy was performed under the diagnosis of left testicular tumor. Macroscopic and microscopic examination of the resected tumor demonstrated it to be paratesticular leiomyosarcoma. The value of serum basic fetoprotein, which is a newly identified oncofetal protein and useful as a tumor marker in some kinds of malignant neoplasm, was elevated before therapy, and its fluctuation was correlated with his clinical course. In spite of chemotherapy and resection of metastatic lesions in the lung, his condition gradually deteriorated with increasing serum basic fetoprotein level, and he died with carcinomatosis 2 years and 6 months after his admission. We suggest that basic fetoprotein is a useful tumor marker in this disease.

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**Key words:** Paratesticular leiomyosarcoma, Basic fetoprotein

### INTRODUCTION

Of the 90% of intrascrotal non-testicular tumors which originate from the spermatic cord<sup>1)</sup>, 70% are benign and 30% are malignant. Paratesticular leiomyosarcoma is a very rare malignant condition, for which there is no useful tumor marker. On the other hand, basic fetoprotein (BFP), an oncofetal protein, is useful as a tumor marker in some types of malignant tumors<sup>2)</sup>. We report herein a case of paratesticular leiomyosarcoma with elevated serum BFP, and discuss the utility of BFP as a tumor marker in this case.

### CASE REPORT

A 45-year-old male was admitted to our hospital on August 18, 1987 with a painless left scrotal mass swelling which had persisted for 6 months. Physical examination revealed a firm mass about 5.0 cm in diameter in the left scrotum. Laboratory data revealed significant elevation of serum BFP (514 ng/ml, normal range, <75 ng/ml), while the levels of  $\alpha$ -fetoprotein (AFP),  $\beta$ -human chorionic gonadotropin

( $\beta$ -hCG), and lactate dehydrogenase (LDH) were within normal limits. Ultrasonography of the scrotal contents revealed a hypoechogenic mass containing a small hyperechogenic area with unclear borderline and the testes.

Chest X-ray demonstrated a few metastatic nodules in the bilateral lungs. Computerized tomography revealed no swelling of the retroperitoneal lymph nodes. High orchiectomy of the left testis was performed under the diagnosis of left testicular tumor. Macroscopically, a firm white tumor was observed. The tumor measured 12×7×6 cm, weighed 350 g, and was encapsulated by a hard white capsule. Microscopic examination revealed the firm fibrous capsule located between the tumor and testes and epididymis, and spindle-shaped tumor cells containing bizarre multinucleated forms with the nucleus (Fig. 1). Mitotic figures were frequent, with an average of 6 per high power field. The tumor was diagnosed as paratesticular leiomyosarcoma, but because it was so large its origin could not be ascertained. Post-operatively, multiple lung metastases were

observed. Although one course of anti-neoplastic chemotherapy (actinomycin-D; 1 mg/m<sup>2</sup>, day 1, vincristine; 1 mg/m<sup>2</sup>, day 1, VP-16; 100 mg/m<sup>2</sup>, day 1-3, ifosphamide; 1 g/m<sup>2</sup>, day 1-3, bleomycin; 10 mg/body, day 1-3 and cisplatin; 100 mg/m<sup>2</sup>, day 4) was administered for the lung metastasis, the response was poor. Therefore, lung metastatic nodules were resected; the histology of the resected tumors was similar to that of the left testicular tumor. The serum BFP level decreased after orchietomy and resection of the lung metastatic nodules. Although the patient was discharged from the hospital in a good condition, lung metastasis was observed 14 months after orchietomy. Therefore, two courses of another anti-neoplastic chemotherapy (methotrexate 20 mg/m<sup>2</sup>, day 1, vincristine; 0.6 mg/m<sup>2</sup>, day 1, cyclophosphamide; 500 mg/m<sup>2</sup>, day 1, adriamycin; 20 mg/m<sup>2</sup>, day 1, bleomycin; 30 mg/body, day 1 and cisplatin; 50 mg/m<sup>2</sup>, day 2) was administered, the effect was also poor. His condition gradually deteriorated and

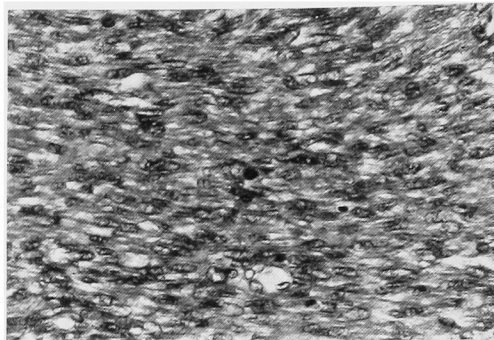


Fig. 1. Microscopic findings for the tumor. Proliferation of spindle-shaped tumor cells with frequent mitosis is seen. Elongated spindle cells contain oval, blunt-ended nuclei and bizarre, giant, strap-like forms. H&E stain, reduced from  $\times 200$

serum BFP value increased, and he died with carcinomatosis 30 months after the first admission (Fig. 2).

#### DISCUSSION

Increased mitotic rate is an important criterion in assessing malignancy in lei-

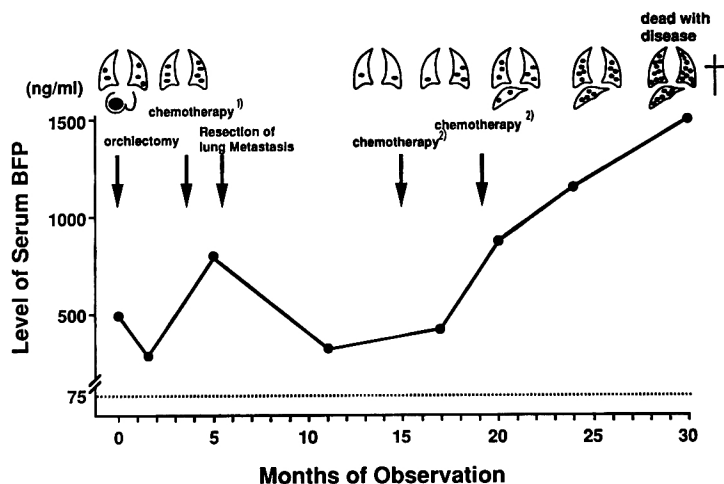


Fig. 2. Clinical course of paratesticular leiomyosarcoma. Serum basic fetoprotein (BFP) was significantly elevated at admission (514 ng/ml; normal range,  $<75$  ng/ml). The value was decreased after orchietomy and after resection of lung metastatic tumors, but the value increased with recurrence of the disease. The patient died after systemic metastases with significantly elevated level of serum BFP were noted. The value of serum BFP was well correlated with clinical course in this patient. 1) Actinomycin D; 1 mg/m<sup>2</sup> and vincristin; 1 mg/m<sup>2</sup> was administered at day 1, VP-16; 100 mg/m<sup>2</sup>, ifosphamide; 1 g/m<sup>2</sup>, and bleomycin; 10 mg/body was administered at day 1-3 and then cisplatin; 100 mg/m<sup>2</sup> was administered at day 4. 2) Methotrexate; 20 mg/m<sup>2</sup>, vincristin; 0.6 mg/m<sup>2</sup>, cyclophosphamide; 500 mg/m<sup>2</sup>, adriamycin; 20 mg/m<sup>2</sup> and bleomycin; 30 mg/body was administered at day 1, and then cisplatin; 50 mg/m<sup>2</sup> was administered at day 2

myosarcoma<sup>3)</sup>. In our case, as 6 mitotic figures per high-power field and large bizarre nuclei were observed, the tumor was thought to be malignant.

Recently, Russo et al. analyzed the clinical features in 43 patients with genitourinary sarcoma treated at Memorial Sloan-Kettering Cancer Center (MSKCC) using the MSKCC sarcoma staging system, which is based on tumor stage, grade, size, and depth, and presence or absence of metastasis<sup>4)</sup>. They found that high grade, advanced-stage tumor, size greater than 5.0 cm, and metastatic sarcoma were unfavorable factors. On the other hand, favorable factors associated with significantly increased survival were size less than 5.0 cm., low histological grade, paratesticular or bladder tumor site, and complete surgical resection. Moreover, they found that chemotherapy was not effective in advanced stage leiomyosarcoma. In our case, as the tumor cells showed frequent mitosis and large bizarre nuclei, the tumor was classified as high grade, and the tumor was larger than 5.0 cm. Cytotoxic chemotherapy was not effective in our case.

In consideration of several reports, in which investigators mentioned that cytotoxic chemotherapy was not effective for the high-stage disease<sup>4,5)</sup>, we performed resection of the metastatic nodules in bilateral lungs in our patient after one course of chemotherapy, and his condition temporarily improved. He was discharged with decreasing serum BFP level, but he died with carcinomatosis 30 months after the first admission, prior to which level of serum BFP had increased.

BFP is an oncofetal protein first detected by M. Ishii et al<sup>2)</sup>. This protein has a molecular weight of 100,000 and isoelectric point of 9.3<sup>2)</sup>. In patients with certain types of malignant tumors, the value of serum BFP is elevated<sup>2,6)</sup>. Among patients with urogenital malignant diseases<sup>7,8)</sup>, the value is elevated in 71% of those with seminoma and 47% of those with renal cell carcinoma. In our case, the values of common tumor markers in testicular

tumor, such as AFP,  $\beta$ -hCG and LDH, were within the normal limits. Serum BFP value was significantly elevated in our case, and the value decreased with treatment and increased with recurrence of the disease. These findings demonstrate that BFP is a potentially useful marker in this case (paratesticular leiomyosarcoma).

Due to the small number of cases reviewed to date, no firm conclusions can be drawn regarding therapy and prognosis. We think that further analysis of many cases is necessary to understand the biology of paratesticular leiomyosarcoma and to establish a more effective therapy for this tumor. Moreover, we suggest that serum BFP is a useful tumor marker in this disease.

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## 和文抄録

## 血清 Basic Fetoprotein が高値を呈した傍精巣平滑筋肉腫の1例

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血清 basic fetoprotein が高値を呈した傍精巣平滑筋肉腫の1例を報告する。

患者は45歳男性で6カ月前より左陰嚢内容の無痛性腫大を自覚し当科を受診した。

左精巣腫瘍の診断の下に左高位除辜術が施行された。肉眼的および病理組織学的所見より摘除腫瘍は傍精巣平滑筋肉腫であった。血清 basic fetoprotein は新しく同定された胎児性蛋白で、ある種の悪性腫瘍で

高値を示す。自験例では術前血清 basic fetoprotein が高値を示し、その値は患者の病状とよく相関した。化学療法および、肺転移巣切除にもかかわらず患者の全身状態が悪化するとともに血清 basic fetoprotein 値は上昇し、患者は入院後2年6カ月目に癌死した。血清 basic fetoprotein は自験例では有用な腫瘍マーカーであると思われた。

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