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CASE REPORT

Ewing's Sarcoma with Extension into Superior Vena Cava and Right Atrium

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Abstract

Ewing sarcoma is rarely shown to develop this intravascular extension so the decision of the initial treatment is more difficult. We report a 7-year-old boy of this sarcoma with extension into superior vena cava (SVC) and right atrium (RA), who was successfully treated with initial surgery. Intravascular extension was observed from the azygous vein to SVC and finally RA. The removal of the intravascular extension was done, 7 days before chemotherapy was started. The initial surgery for the intravascular extension may have decreased a risk of pulmonary tumor embolism and this made the chemotherapy done safe in this patient.

Keywords Ewing's sarcoma · Intravascular extension · Tumor embolism · Initial treatment

For tumors with an intravascular extension, the initial treatment decision is complicated. Wilms tumor is known to develop intravascular extensions, and preoperative chemotherapy is also known to be effective [1]. On the other hand, sudden death during chemotherapy has also been reported [2]. Furthermore, there are few reports of extraskeletal Ewing's sarcoma (EWS) extension into vessels [3–12], making the decision regarding initial treatment for EWS more complicated than for Wilms tumor. The present report concerns a pediatric case of extraskeletal EWS with extension into the superior vena cava (SVC) and right atrium (RA) that was removed by surgery as the initial treatment.

Case Report

A 7-year-old boy was admitted into our hospital with back pain that began 19 days before the date of admission. Ten days from the onset date, a limp and gait disturbance developed. Leg weakness and dysuria developed on the next day. At the previous hospital, magnetic resonance imaging (MRI) showed an intraspinal tumor compressing the spinal cord. Emergency spinal decompression surgery was done. Six days later, he was referred to our center for further treatment.

MRI showed a large tumor extending from the right posterior mediastinum into the epidural space and impinging on the spinal cord. Intravascular extension reached the azygous vein, SVC, and RA. Echocardiography showed an intravascular tumor in the azygous vein and SVC extending into the RA. The histology of the tumor extracted from the spinal canal showed the preliminary findings of undifferentiated round cells and *EWSR1-FLI1*. The final diagnosis was of Ewing's sarcoma. Distant metastases were confirmed in the right temporalis muscle and the right gluteus maximus.

Surgery was chosen due to the high probability of sudden death resulting from a tumor embolism during the initial chemotherapy. In the embolectomy under cardiopulmonary bypass, the SVC appeared swollen due to the tumor, which was detached from the azygous vein following dissection of the SVC and RA. The tumor did not adhere to the intima or the tricuspid valve. By pulling the adherent portion away from the azygous vein, the inlet portion to the SVC was able to be ligated (Fig. 1). Although incomplete resection of the tumor was performed, the postoperative course was good, and chemotherapy consisting of vincristine, doxorubicine, ifosfamide, and etoposide (VIDE) was started based on the EuroEwing99 protocol [2] on postoperative day 7. After six courses of VIDE, the patient underwent high-dose chemotherapy

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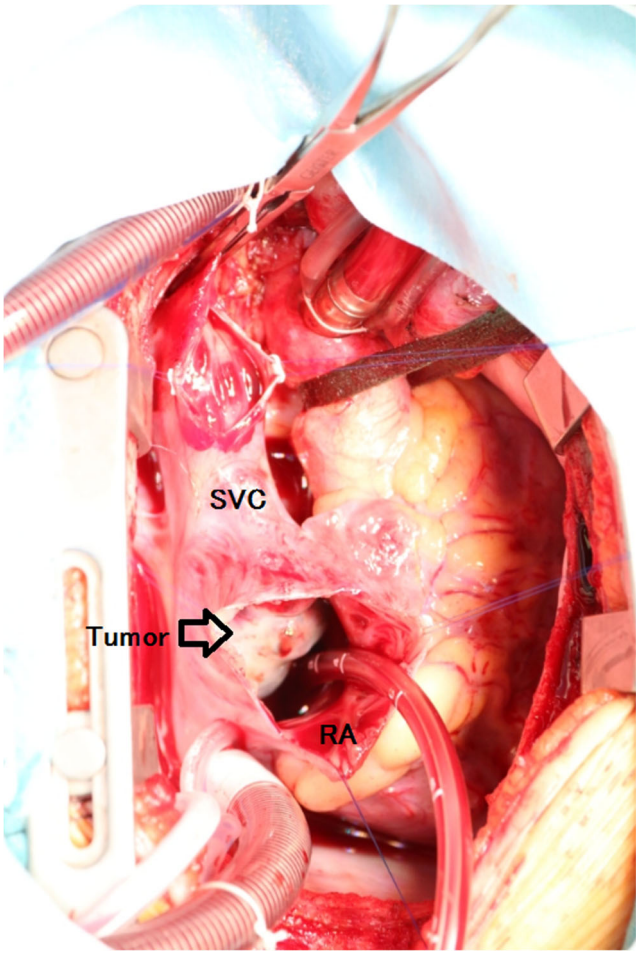

Fig. 1 The inlet portion to the SVC

(busulfan and melphalan) and autologous bone marrow transplantation. For the metastases to the head, electron beam treatment was administered (45Gy/25fr). The patient remains disease-free one-and-a-half years after the end of the treatment, and is now able to walk with the aid of crutches.

Discussion

For intravascular extension of a tumor, choosing an initial treatment is difficult. The present case report is the first to describe Ewing's sarcoma extending from the azygous vein into the heart. The preoperative chemotherapy was chosen in some cases of EWS invading the heart [4] because these patients could not underwent tumor resection without heart transplantation. Mete UK et al. [5] showed a case with EWS with tumor thrombus extending from inferior vena cava to RA. This tumor spread across the diaphragm, thus this case was managed with chemotherapy for the initial treatment. The site and spread of the tumor are important factor for choice the initial treatment. On the other hand, unlike Wilms tumor, in previous reports of EWS cases with intravascular extension, tumor resection was chosen as the initial treatment in some cases [6–12] and the progress of treatment was reportedly favorable. The review reported by Mcmahon et al. [13] showed that the effect of chemotherapy is an important factor in the choice of the initial treatment thus the difference between EWS and Wilms tumor is thought to come from the difference of the effect of chemotherapy. The number of cases is small and future study is needed.

In the present case, we first consulted the surgeons and intensive care physicians regarding the initial treatment. The delay in postoperative chemotherapy or complications arising during cardiopulmonary bypass was thought to be critical. On the other hand, initial chemotherapy carried the risk of sudden death due to pulmonary embolisms from the tumor in the RA. The tumor resection was considered to be the safer alternative and was performed as the initial treatment under cardiopulmonary bypass. Seven days after the surgery, chemotherapy was performed safely without delay.

In EWS, particularly in cases of tumors extending into the heart, we should take potential complications and the possibility of delaying postoperative chemotherapy into account and choose the least invasive form of treatment.

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Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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