

Video-assisted Thoracic Surgery for Ewing's Sarcoma of the Mediastinum in a 3-year-old Girl

Kiyoshi Koizumi, MD,¹ Shuji Haraguchi, MD,¹ Iwao Mikami, MD,¹
Hirotohi Kubokura, MD,¹ Daisuke Okada, MD,¹ Shigeki Yamagishi, MD,¹
Hiroyasu Kinoshita, MD,¹ Yutaka Enomoto, MD,¹ Kazuo Shimizu, MD,¹
and Miho Maeda, MD²

We report of the experience of video-assisted thoracic surgery (VATS) for a Ewing's sarcoma at the posterior mediastinum in a 3 year old girl. The patient developed common cold like symptoms and developed rapid lower limb paraplegia. A chest computed tomography (CT) showed a solid mass adjacent to the vertebrae. This was diagnosed as a Ewing's sarcoma histopathologically by CT-guided needle biopsy (CT-NB). The mass was pressing on a nerve root in the vicinity of a vertebral foramen. As the tumor reduced after induction chemotherapy, with improvement of paraplegia, the patient underwent thoracoscopic examination followed by complete removal of the tumor by VATS. After surgery, the patient underwent postoperative chemo-radiotherapy. Five years has passed after operation without tumor recurrence. VATS could be an option as combined therapy for Ewing's sarcoma when well controlled and localized under induction therapy. (Ann Thorac Cardiovasc Surg 2005; 11: 117–20)

Key words: children, Ewing's sarcoma, mediastinal cyst, thoracoscopic surgery

Case Report

A three year old girl suffered from dysbasia followed by continuous common cold like symptoms in October, 1998. It became impossible for the girl to stand up at the beginning of November, 1998 and she rapidly developed lower limb paraplegia. The patient presented at the Department of Pediatrics Nippon Medical School Hospital, her height was 104 cm, and body weight 16 kg. The patient showed no anemia or jaundice on admission. The blood examination and the biochemistry findings were within the normal range, and there was no elevation of the tumor markers (CEA, 0.6 ng/ml; NSE, 10 ng/ml; AFP, 1.7 ng/ml). Behind the left side of the heart, a pyriform-like mass shadow was found on a chest X-ray (Fig. 1). A chest CT

From ¹Division of Thoracic Surgery, Department of Surgery, and ²Department of Pediatrics, Nippon Medical School, Tokyo, Japan

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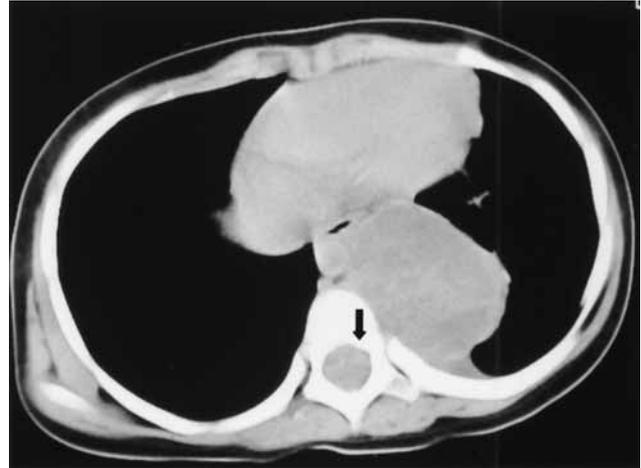
Address reprint requests to Kiyoshi Koizumi, MD: Division of Thoracic Surgery, Department of Surgery, Nippon Medical School, 1-1-5 Sendagi, Bunkyo-ku, Tokyo 113-8603, Japan.

scan revealed a mass shadow of 4×5×5 cm in the posterior mediastinum adjacent to the descending aorta and the thoracic vertebra (Fig. 2A). The tumor was spreading into the spinal canal though the neural foraminae at the 6th to 8th vertebral body, and pressed on the cord from the left side. MRI revealed a well encapsulated mass showing a heterogeneous cystic pattern (Fig. 3A). A CT-guided needle biopsy (CT-NB) was carried out and the tumor was diagnosed as a small round-cell tumor (Ewing's sarcoma) as shown in Fig. 4. Immunohistochemical examination revealed positive studies for PAS, NSE, and vimentin. Chemotherapy was undertaken using 750 mg/m² of cyclophosphamide, 1 cycle; 1.5 mg/m² of vincristine, 1 cycle; 30 mg/m² of adriamycin, 2 cycles; 45 mg/m² of cis-platin, 2 cycles. A chest CT scan on December 22, 1999 (Fig. 2B) and MRI on January 5, 1999 (Fig. 3B) showed cystic change of the tumor material. Improvement of paraplegia of the inferior limb was evident. The patient was referred to the division of thoracic surgery for thoracoscopic assessment, with a view to total excision of the tumor. On January 19, 1999, she underwent a thoracoscopic (5 cm) assessment. The patient was placed



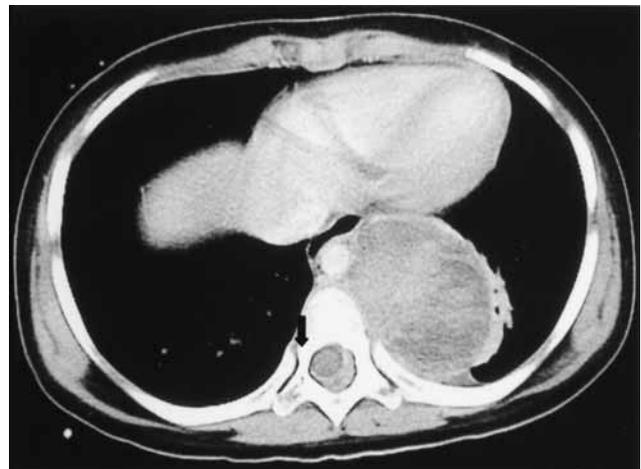
Fig. 1. Preoperative chest X-ray showed a pyriform-like mass shadow behind left side of the heart.

in the left lateral position under general anesthesia. Ventilation was maintained using a 4Fr. endotracheal tube. The first port was made in the fifth intercostal space of the anterior axillary line and the second port was made with the seventh intercostal space of the posterior axillary line (Fig. 3C). Thoracoscopic observation revealed a tumor localized in the posterior mediastinum. This was observed to be a cystic tumor adjacent to the thoracic vertebra without invasion of surrounding tissue (Fig. 3D). It was planned to perform total removal of tumor by video-assisted thoracic surgery (VATS). When parietal pleura was dissected and exposed, the tumor seemed to easily dissect from the surrounding tissue. There was fibrinous adhesion around the aperture but no tumor invasion. We taped the vagus nerve and started extraction of the tumor. The sixth intercostal nerve was unified within the tumor, and the base of the tumor seemed to continue toward the neural foramen. The tumor was drawn up very carefully from the aperture. The duration of surgery was 250 minutes and the blood loss was 113 ml. The cut surface of the cystic tumor showed an irregular lobular pattern involv-



A

1998. 11. 26



B

1998. 12. 22

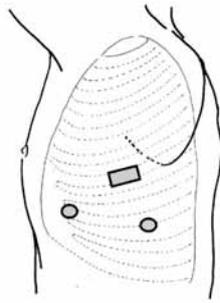
Fig. 2. A: Preoperative chest CT scan revealed a mass shadow of inside heterogeneity of 4×5×5 cm in the posterior mediastinum adjacent to the descending aorta and the thoracic vertebra, which was spreading into the spinal canal through neural foramenae at the level of the 6th to 8th vertebra, and oppressed the spinal cord from the left side (arrow).

B: After induction chemotherapy, the tumor slightly reduced in size and the content changed to cystic appearance (arrow).

ing hemorrhages, necrosis and fibrosis. On pathological examination, the cystic wall was completely fibrotic and the content showed necrosis with hemorrhages, thrombosis and deposition of hemosiderin. The remnant small round-cell tumor was confirmed in cytological analysis of cyst contents. The postoperative course was uneventful, and postoperative chemotherapy was started on the 14th postoperative day. The postoperative chemotherapy consisted of 1.5 gr/m² of dactinomycin, on day 8, 12, 16, 20; 1.5 gr/m² of vincristine, on day 8, 10, 12, 14, 16, 20; 1.5 gr/m² of cyclophosphamide, on day 5; 2 gr/m²/day of



1998. 12. 1



1999. 1. 5



A	C
B	D

Fig. 3. A: Before induction chemotherapy, MRI showed a well encapsulated mass, which revealed a heterogeneous cystic pattern. B: MRI after induction chemotherapy showed cystic change. C: Location of the minithoracotomy and two ports. D: Thoracoscopic observation revealed the cystic tumor. The parietal pleura were dissected along the tumor. T, tumor; P, parietal pleura.

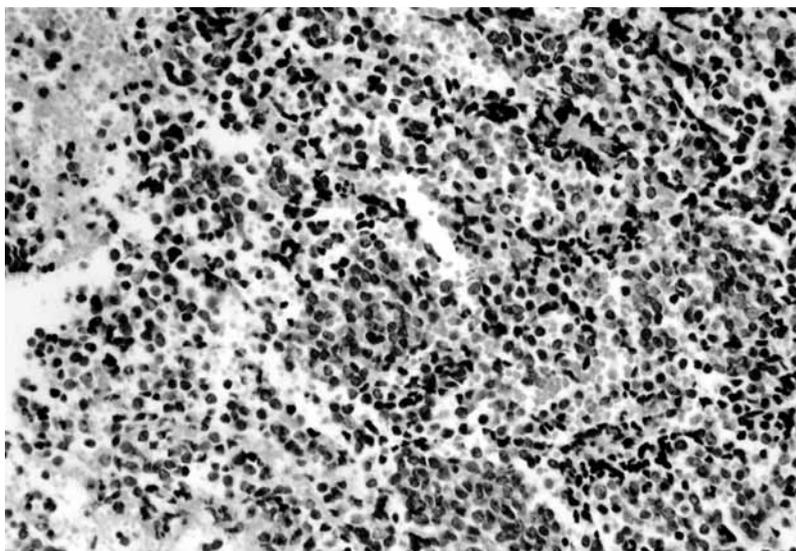


Fig. 4. Pathological examination by a CT-NB showed a small round-cell tumor (Ewing's sarcoma).

ifosfamide on day 1, 2, 3; 45 mg/m²/day of doxorubicin, on day 5 and 150 mg/m²/day of etoposide on day 1, 2, 3, respectively. Additional radiotherapy by 18 Gy was given over ten days. Five years has now passed after her surgery and the patient remains healthy and tumor-free.

Discussion

Pediatric thoracoscopy was introduced by Rodgers in 1979. Thoracoscopic surgery has been applied to malignant chest disease and benign chest disease.¹⁾ The advantage of thoracoscopy was ease of diagnosis.^{2,3)} In treatment of malignant disease, thoracoscopy was reported to be useful in establishing a quick and reliable diagnosis, and assisting appropriate treatment planning about chemotherapy, radiotherapy and/or surgery.⁴⁻⁸⁾ With these advantages, Smith et al.⁶⁾ emphasized that rapid postoperative recovery in VATS allowed rapid use of postoperative chemo-radiotherapy in comparison with thoracotomy. The benefits of tri-modality therapy are recognized in treatment for malignant neoplasm in childhood. Regarding chemotherapy, Marina, et al. reported the feasibility of dose-intensification for pediatric patients with Ewing's family of tumors and desmoplastic small round-cell tumors.⁹⁾ They emphasized the efficacy of dose-intensification when a tumor was well localized, and showed longevity of survival comparable to patients with metastasis. It is speculated that the disease derives from a peripheral original neuroectodermal germ cell. It is hypothesized that the tumor metastasis through the neural foramen. If the tumor is less than 8 cm without apparent metastasis, successful chemotherapy and excision can be expected.⁹⁻¹¹⁾ In this case, the tumor was 5 cm in diameter, without metastasis. The patient obtained tumor reduction and improvement of paraplegia of the inferior limb with chemotherapy. In summary, when induction therapy offered reduction of tumor and symptomatic improvement, the VATS procedure would be considered as an option as minimally invasive surgery for the localized mediastinal Ewing's sarcoma in children.

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