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Advanced surgical strategy for giant mediastinal germ cell tumor in children

Shigehisa Fumino [a], Kohei Sakai [a], Mayumi Higashi [a], Shigeyoshi Aoi [a], Taizo Furukawa [a], Masaaki Yamagishi [b], Masayoshi Inoue [c], Tomoko Iehara [d], Hajime Hosoi [d], Tatsuro Tajiri [a,*]

[a] Department of Pediatric Surgery, Kyoto Prefectural University of Medicine, Kyoto, Japan
[b] Department of Pediatric Cardiovascular Surgery, Kyoto Prefectural University of Medicine, Kyoto, Japan
[c] Department of Thoracic Surgery, Kyoto Prefectural University of Medicine, Kyoto, Japan
[d] Department of Pediatrics, Kyoto Prefectural University of Medicine, Kyoto, Japan

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ABSTRACT

Purpose: Giant mediastinal germ cell tumor (MGCT) requires a well-planned advanced surgical approach. We retrospectively reviewed our surgical strategy for giant MGCT.
Methods: Five children (median age, 5 years) with giant MGCT were treated in our institute from 2012 to 2016.
Results: The initial diagnosis was made by tumor markers and image inspection in all cases. Benign teratomas (2 girls) and malignancies (3 boys) were treated with upfront surgery and radical tumorectomy after neo-adjuvant chemotherapy, respectively. After detailed 3D-CT, radical tumor excision was performed supported by a skilled pediatric cardiovascular surgeon. The basic approach was as follows: under cardiopulmonary support (CPS) or with CPS on standby, via median sternotomy, the pericardium and phrenic nerve were resected en bloc with the tumor, followed by diaphragmatic plication. Open biopsy was performed via lateral thoracotomy in 1 patient who showed dense adhesion and fistula formation in the lung; lobectomy via hemi-clamshell incision was required. No deaths or severe sequelae occurred in this series.
Conclusions: Resectability is the most important predictor of outcomes for MGCTs. Preoperative 3D-CT and CPS can enable complete resection and ensure surgical safety. Well-functioned surgical team is critical success factor in such advanced surgery.

1. Introduction

Germ cell tumors (GCTs) are rare tumors thought to be derived from totipotential primitive germ cells that either mismigrate along the urogenital ridge during early embryogenesis or are distributed physiologically. GCTs show a wide diversity of benign or malignant characteristics [1]. Among extragonadal GCTs, mediastinal germ cell tumors (MGCTs) in children are extremely rare and often form giant masses invading the surrounding vital organs and tissues [2]. Therefore, the treatment of giant MGCTs is challenging and requires a well-planned advanced surgical approach.

In this study, we retrospectively reviewed our experience and discussed our surgical strategy for giant MGCT.

2. Patients and methods

From 1989 to 2016, 74 children with germ cell tumors were treated at our institution. Among them, five patients had giant GCTs in the anterior mediastinum and surgically treated between 2012 and 2016. There were 3 boys and 2 girls, and the median age at the diagnosis was 5 years (range, 1–15). The medical charts were retrospectively reviewed for the collection of data on clinical symptoms, size, tumor marker, surgical intervention, perioperative complications, pathology, and outcomes.

* Corresponding author. Department of Pediatric Surgery, Kyoto Prefectural University of Medicine, 465 Kawaramachi-Hirokoji, Kamigyo-ku, Kyoto, 602-8566, Japan.
E-mail address: taji@koto.kpu-m.ac.jp (T. Tajiri).

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This study was performed in accordance with the Ethical Guidelines for Medical and Health Research Involving Human Subjects by the Ministry of Health, Labour, and Welfare of Japan in 2014 and complies with the Helsinki Declaration of 1964 (revised in 2013). The study was approved by the local ethics committee of our institution.

3. Results

The clinical characteristics of the five patients are listed in Table 1. All patients were symptomatic, with respiratory symptoms including cough and dyspnea in all five cases, a fever in four, and chest wall projection in one. These symptoms led them to visit our medical institution, and several imaging studies including X-ray, chest computed tomography (CT), and magnetic resonance imaging (MRI) revealed that they had a giant mass in the chest.

All tumors originated within the anterior mediastinum and extended into the left hemithorax in four patients and the right hemithorax in one. The tumors were composed of solid and cystic areas, fat densities, and irregular calcifications of varying degrees. The maximum tumor diameter ranged from 5.6 to 18.0 cm, and the malignant lesions were bigger than the benign ones. Tumor markers were determined in all cases; alpha-fetoprotein (AFP) was strongly elevated in the 3 malignant cases (12,520–116,657 ng/mL). Human chorionic gonadotropin (HCG) was within normal ranges in all cases. A pre-treatment biopsy was done in two cases suspected of having malignancy. One underwent an open biopsy via lateral thoracotomy (Case 5), and the other underwent a cervical lymph node biopsy (Case 3).

Two girls were diagnosed with a benign mature teratoma (Cases 1 & 2) based on CT and tumor marker findings and underwent upfront surgery. Three boys had a malignant tumor including a yolk sac tumor in one and a non-seminomatous combined tumor in two, and surgical extirpation of the residual mediastinal mass was performed after six to eight cycles of cisplatin-based neoadjuvant chemotherapy and when the tumor marker values decreased to within normal ranges.

Before surgery, detailed three-dimensional (3D)-enhanced CT was performed in all patients, and the vascular anatomy was precisely reconstructed (Fig. 1). A tumor board consisting of pediatric oncologists, radiologists, and the pediatric surgical team was convened for each surgery. The surgical team included pediatric surgeons and pediatric cardiovascular surgeons mainly, and an otolaryngologist and a thoracic surgeon joined in some cases. The surgical strategy was discussed and developed in these board meetings.

All tumor excision operations were supported by a skilled pediatric cardiovascular surgeon. Our surgical approach was as follows: under cardiopulmonary support (CPS) or with CPS on standby, via median sternotomy, we reached the anterior mediastinum. The tumor was dissected, and the attached thymus, the pericardium, and the phrenic nerve of the affected side were resected en bloc with the tumor, followed by diaphragmatic plication to prevent diaphragmatic eventration. If there were the other organs with adhesion or invasion by the tumor, such as lung, thoracic wall, or major vessels, they were resected together via collar incision or hemi-clamshell incision if necessary. Vascular reconstruction using an ePTFE graft was performed in Case 3.

In this series, no operative mortality was noted. Postoperative morbidity occurred in four patients, with one patient each developing pericardial effusion, hoarseness, pyothorax, and chylothorax. Although three complications were resolved with conservative therapy, the patient with chylothorax (Case 3) was intractable to conservative treatment such as fat restriction and octreotide administration. He ultimately underwent thoracoscopic sealing

Table 1
Summary of the cases with mediastinal germ cell tumor.

No.	Age (yr)	Sex	Diagnosis	Clinical symptoms	Size (cm)	AFP (ng/mL)	Biopsy	CPS	Surgical procedure		Outcomes			
									Skin incision	Findings	Operation time	Blood loss	Complication	
1	1	F	mature teratoma	fever, cough	8.0 × 5.0	6	–	on	mid-sternotomy	Excision of tumor with phrenic nerve + pericaridum+a part of lung	3 h 20 min	No count	–	well
2	5	F	mature teratoma	fever, cough	5.6 × 4.5	1	–	stand-by	mid-sternotomy	Excision of tumor with pericaridum	2 h 42 min	10 g	pericardial effusion	well
3	6	M	yolk sac tumor	fever, cough, chest wall projection	13.5 × 13.0	116,657	cervical lymph node	on	mid-sternotomy	Excision of tumor wth phrenic nerve + pericaridum + lt. Innominate v. with vascular reconstruction	4 h 20 min	358 g	chylothorax requiring thoracoscopic repair	well
4	15	M	malignant	fever, cough, dyspnea	17.0 × 18.0	16,880	–	on	mid-sternotomy + collar incision + inguinal incision	Excision of tumor wth phrenic nerve + pericaridum + bil. IJV + SVC without vascular reconstruction	12 h	2050 g	hoarseness	well
5	15	M	malignant	cough	13.0 × 7.7	12,520	open biopsy via thoracotomy	stand-by	mid-sternotomy + hemi-clamshell	Excision of tumor with phrenic nerve + pericaridum + lt. upper lobe of lung	10 h 34 min	840 g	pyothorax	recurrence with lung metastasis

CPS; cardiopulmonary support.
IJV; internal jugular vein.
SVC; superior vena cava.