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## Journal of Pediatric Surgery CASE REPORTS

journal homepage: [www.jpascasereports.com](http://www.jpascasereports.com)Mediastinal extracardiac fetal rhabdomyoma; case report<sup>☆</sup>Go Miyano<sup>a,\*</sup>, Hiromu Miyake<sup>a</sup>, Masakatsu Kaneshiro<sup>a</sup>, Keiichi Morita<sup>a</sup>, Hiroshi Nouse<sup>a</sup>, Masaya Yamoto<sup>a</sup>, Koji Fukumoto<sup>a</sup>, Yasuo Horikoshi<sup>b</sup>, Kazuko Kudo<sup>b</sup>, Minoru Hamazaki<sup>c</sup>, Naoto Urushihara<sup>a</sup><sup>a</sup> Department of Pediatric Surgery, Shizuoka Children's Hospital, Shizuoka, Japan<sup>b</sup> Department of Hematology and Oncology, Shizuoka Children's Hospital, Shizuoka, Japan<sup>c</sup> Department of Pathology, Shizuoka Children's Hospital, Shizuoka, Japan

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## ABSTRACT

A 9-month-old girl with a small suprasternal mass was referred for treatment after fine needle aspiration of the mass elsewhere suggested rhabdomyosarcoma. On admission, magnetic resonance imaging (MRI) identified a 58 × 42 mm oval mass in the anterior mediastinum close to the common carotid artery and aortic arch. Surgical intervention was initially considered to be too risky, but after a trial of chemotherapy failed to reduce the size of the tumor, complete local excision was planned through a vertical median thoracic incision with median sternotomy. Surgery was performed when she was 10 months old. Due to severe adhesions, the left clavicle, first rib and the sternoclavicular joint needed to be partially resected to allow the tumor to be completely excised with safety. Operating time was 3 h; intraoperative blood loss was 123 mL requiring intraoperative transfusion. Histopathology and immunohistochemistry were consistent with an intermediate form of fetal rhabdomyoma. The postoperative course was uneventful and she is currently alive with no evidence of disease 4 years later. Fetal rhabdomyoma is rare and our case presented with a small, asymptomatic, suprasternal mass. While the distinction between rhabdomyoma and rhabdomyosarcoma can be difficult, a multidisciplinary approach enabled surgery to be performed successfully.

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Rhabdomyoma is a rare benign neoplasm of soft tissue, accounting for only 2% of all tumors with skeletal muscle differentiation; i.e., tumors with cells differentiated as skeletal muscle cells with cytoplasmic cross-striations [1]. Most patients are between 40 and 70 years old, although the age range extends from 2 to 82 years, and the male to female ratio is 3–4 to 1 [2]. There are two main types, cardiac and extracardiac, with extracardiac consisting of three clinical and histologic subtypes: adult, fetal, and genital accounting for 50, 40, and 10% of tumors, respectively [1]. Fetal rhabdomyomas arise in striated muscle, particularly in children under 3 years of age and more frequently in boys. Head and neck lesions are generally located near the aerodigestive tract or in the retroauricular region [3]. In children, approximately two dozen cases of cervicofacial extracardiac fetal

rhabdomyoma have been reported in the literature to date, and a few cases have been identified in other anatomic sites, such as a lower extremity [4], the abdominal wall [5], or retroperitoneum [6], with only one case of extracardiac fetal rhabdomyoma being reported in the posterior mediastinum. This is the first reported case of fetal type rhabdomyoma presenting in the anterior mediastinum which was surgically resected through a median sternotomy.

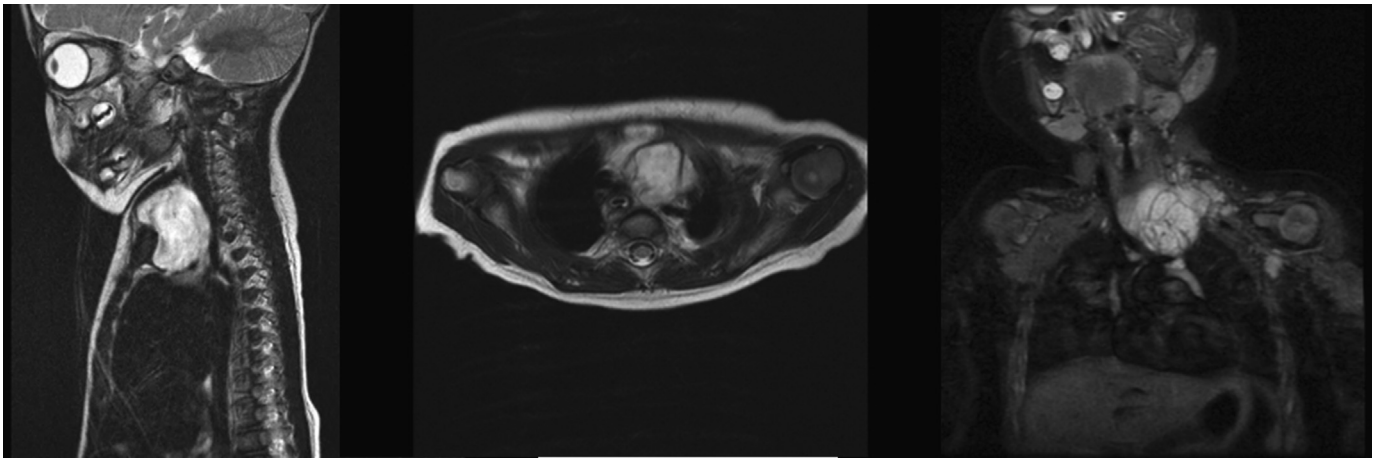
## 1. Case report

A 9-month-old girl was referred to the Department of Pediatric Surgery at Shizuoka Children's Hospital for investigation of a small suprasternal mass identified at a regular check-up. On assessment she was noted to be in mild respiratory stress and a 10 mm swelling was noted to be present superior to the sternum. She was admitted for further investigation despite normal laboratory tests. Ultrasonography identified a small nodule, and magnetic resonance imaging confirmed the presence of a 58 × 42 mm oval mass with heterogeneous signal intensity located in the anterior mediastinum, extending from the superior aspect of the aortic arch to the

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**Fig. 1.** Preoperative MRI confirmed the presence of an oval mass located in the anterior mediastinum, extending from the superior aspect of the aortic arch to the base of the neck, displacing the trachea and mediastinal vessels.

base of the neck, displacing the trachea and mediastinal vessels. The mass appeared to be mainly mediastinal (Fig. 1).

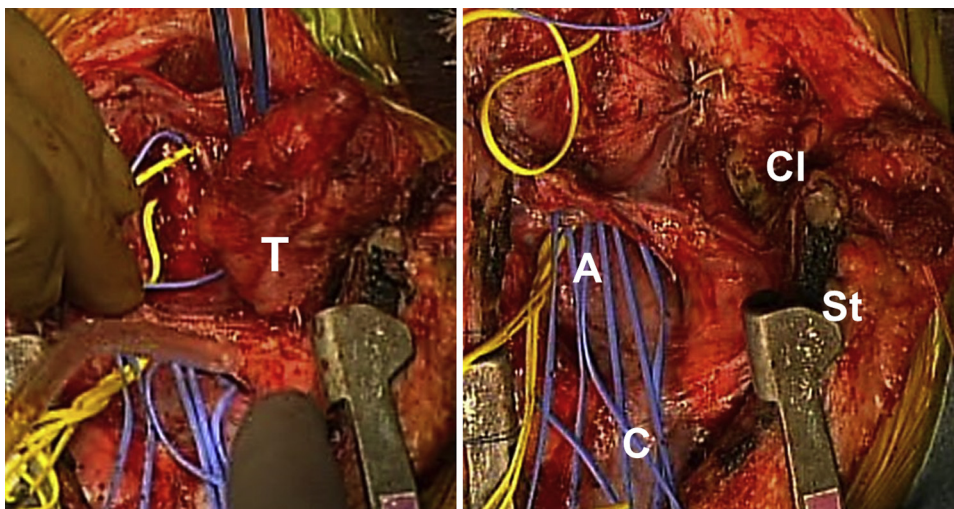
Because fine needle aspiration performed elsewhere was suggestive of rhabdomyosarcoma, and the behavior of the tumor was also suggestive of malignancy, a provisional diagnosis of rhabdomyosarcoma had been made elsewhere before she was referred. Owing to the proximity of the tumor to major vessels such as the common carotid artery and the aortic arch and a high risk for mortality associated with complete surgical excision, a trial of chemotherapy, including cyclophosphamide, cisplatin, and vincristine was chosen as the initial treatment after consultation with colleagues from pediatric oncology, radiology, and pathology. However, after 4 courses, there was no reduction in tumor volume and chemotherapy was abandoned.

The child underwent complete local excision of the mass at 10 months of age. A vertical median thoracic incision with median sternotomy was chosen for access initially but required extension toward the left clavicle later on. The tumor was found to be so strongly adhered to the left clavicle and the left first rib that it appeared to originate from these structures. On the posterior side of the sternum, there were no remarkable adhesions present. The clavicle, first rib, and sternoclavicular joint were all partially

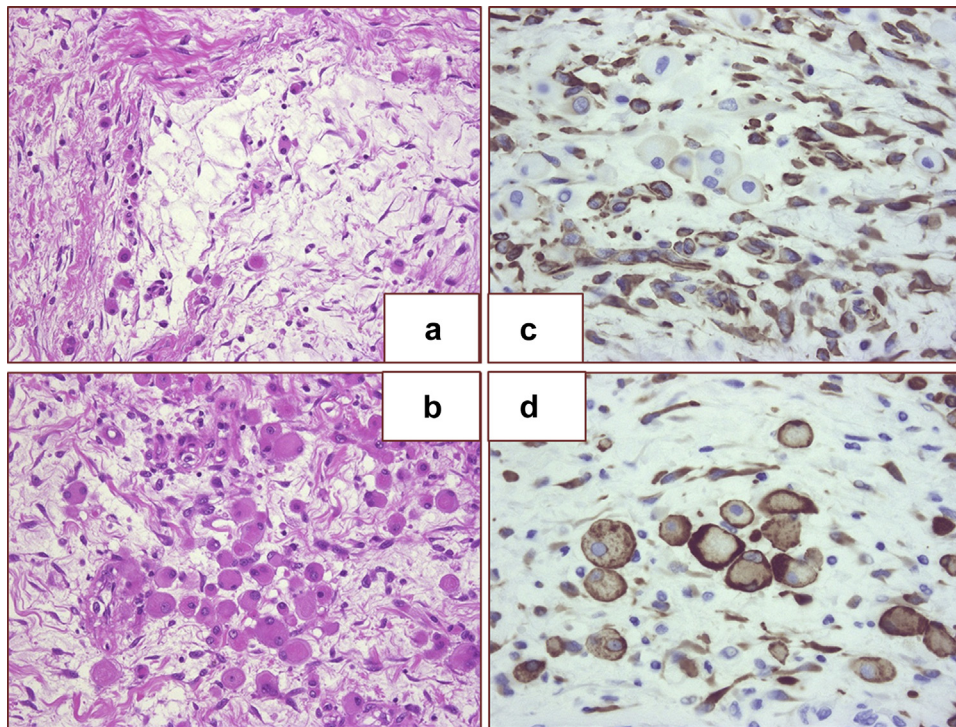
resected before the tumor could be excised completely in safety because of the seemingly highly malignant nature of the tumor on preoperative assessment and on direct observation (Fig. 2). Operating time was 3 h. There was 123 mL of hemorrhage, requiring intraoperative transfusion.

Histologic findings were of a well-circumscribed cellular lesion composed of oval to spindle-shaped cells with indistinct cytoplasm set in a variable fibromyxoid stroma (Fig. 3-a). Some ganglion like rhabdomyoblasts were noted (Fig. 3-b). Occasional cells showed cytoplasmic cross-striations suggestive of rhabdomyoblastic differentiation. Mitotic figures were absent, and neither immature areas nor cell atypia were seen. Immunohistochemistry showed a positive cytoplasmic reaction to vimentin (Fig. 3-c) and to muscle antigens, such as desmin (Fig. 3-d). Approximately 1%–5% of the tumor cell nuclei marked with Ki-67. Histopathologic findings were consistent with a myxoid form of fetal rhabdomyoma, so a diagnosis of rhabdomyoma of fetal type was made.

The postoperative course was uneventful and she was discharged home on the 10th postoperative day. Repeat chest radiograph 1 month after discharge and a computerized tomography scan 3 months after discharge were normal (Fig. 4). Currently she is well with no evidence of recurrence, 4 years after surgery.



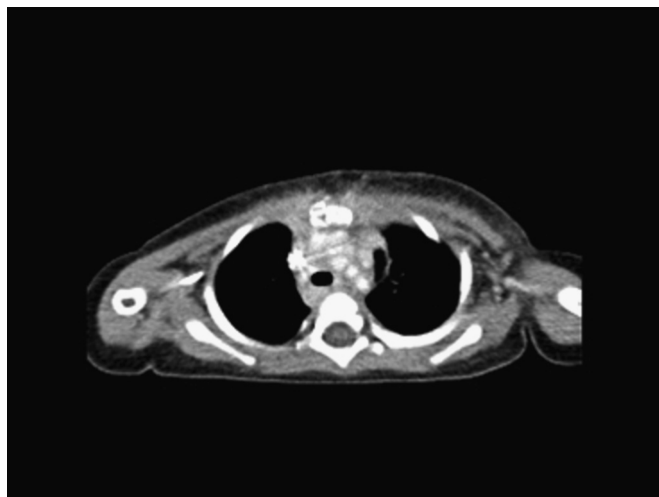
**Fig. 2.** Complete local excision of the tumor (T) was performed safely through a vertical median thoracic incision and median sternotomy. A: Aortic arch, Cl: Left clavicle, St: Sternum, C: Pericardium.



**Fig. 3.** Histologic findings were that of a well-circumscribed cellular lesion composed of oval to spindle-shaped cells with indistinct cytoplasm set in a variable fibromyxoid stroma; which was consistent with an myxoid form of fetal rhabdomyoma (3-a). Some ganglion like rhabdomyoblasts were noted (3-b). Immunohistochemistry showed a positive cytoplasmic reaction to vimentin (3-c) and to muscle antigens, such as desmin (3-d).

## 2. Discussion

There are only five adult cases of extracardiac rhabdomyoma located in the mediastinum reported in the literature [1,7–10]. Two were in the anterior mediastinum, three were in the posterior mediastinum, and one case was multifocal. The age range of these five cases was 52–80 years old. Clinical information is only available for four of these cases, and all were symptomatic with recurrent fever, dyspnea, respiratory airway infection, and dysphagia. In children, there is one case of adult rhabdomyoma arising in the anterior-superior mediastinum in a 9-year-old girl [11], and a case of fetal rhabdomyoma arising in the posterior mediastinum in a 6-



**Fig. 4.** No recurrence of tumor was identified on chest CT 3 months after resection.

year-old girl [6]. Although rare, no instance of aggressive local tumor growth or metastasis has been documented. Local tumor recurrence has been reported, usually attributed to incomplete resection [12] and surgical intervention continues to be the treatment of choice for fetal rhabdomyoma.

Although, it is extremely important to make an accurate diagnosis for efficiency of treatment, particularly in difficult locations such as our case, fetal rhabdomyoma can be confused with malignant growths of similar histopathology. The most serious condition with grave sequelae is rhabdomyosarcoma but distinction can be complicated because fetal rhabdomyoma can be of variable cellularity with differing degrees of skeletal muscle differentiation and have a range of cell types [13]. In fact, the first biopsy performed elsewhere was more suggestive of rhabdomyosarcoma because the tumor was noted to be invading surrounding structures which is a distinctly malignant trait. Use of chemotherapy is unusual for benign tumors, but can be proposed for aggressive lesions such as desmoid fibromatosis or when surgical excision is considered too risky [14]. In the present case, the location of the tumor near vital structures and the provisional diagnosis of rhabdomyosarcoma prompted us to err on the side of caution and use chemotherapy to try and reduce the size of the tumor to facilitate excision, but this proved to be unsuccessful and radical surgical resection was performed after 4 weeks of chemotherapy.

To the best of our knowledge, our patient is the first pediatric case of extracardiac fetal rhabdomyoma arising in the anterior mediastinum, treated by radical resection and partial resection of the first rib and clavicle through a median sternotomy. As such, it is an extremely rare condition that can be difficult to plan treatment for because of problematic diagnosis but fetal rhabdomyoma should be included in the differential diagnosis of large, asymptomatic, anterior mediastinal mass.

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