

Prenatally diagnosed large mediastinal lymphangioma: A case report

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Key Clinical Message

Thoroughly planned labor with immediate postnatal resuscitation and percutaneous decompression is mandatory for a large mediastinal lymphangioma that compromises the cardiopulmonary function, and close observation with the administration of Eppikajutsuto (TJ-28), a Japanese herbal medicine, may be a reasonable treatment, especially when resection of the lesion seems technically challenging.

KEY WORDS

mediastinal lymphangioma, prenatal diagnosis

The incidence of lymphangioma is 1 in 6000 pregnancies, with most lesions occurring on the neck and axilla. Lymphangioma confined to the mediastinum is quite rare and mostly remains asymptomatic until adulthood.¹ We herein report a case in which a large mediastinal lymphangioma that was prenatally diagnosed and which required immediate postnatal resuscitation due to respiratory failure, in which the patient was successfully treated with the prompt percutaneous drainage of the lesion.

A 29-year-old mother (gravida 1, para 1) was referred to the Department of Obstetrics in our hospital because fluid collection in the thoracic cavity was detected by fetal ultrasonography at the 33 weeks of gestation. Fetal magnetic resonance imaging (MRI) revealed a large cystic lesion in the anterior mediastinum, surrounding the heart and large vessels (Figure 1A). It occupied more than half of the thoracic cavity, compressing the bilateral lungs backward. As the fetal ultrasonography demonstrated a few septums inside the cystic lesion, lymphangioma was most likely the diagnosis (Figure 1B). Judging from the size and location of the lesion, postnatal respiratory distress seemed inevitable. Based on a discussion among gynecologists, neonatologists, and our team, we planned to deliver the baby by Caesarian section around the term period (if the well-being of the fetus

was maintained), with prompt postnatal resuscitation with ultrasonography-guided percutaneous drainage on site. We also discussed the possibility of thoracotomy under extracorporeal membrane oxygenation (ECMO) beforehand and simulated the procedures, in case the tube drainage failed to decompress the lesion adequately. On MRI, the fetal trachea seemed straight without a mass effect caused by the lesion, so endotracheal intubation immediately after birth seemed feasible. The delivery was planned for the 37 weeks of gestation. The lesion gradually became enlarged, and the mother was hospitalized at 35 weeks and 6 days of gestation. The fetal cardiac output measured by echocardiography decreased from 504 mL/kg/min (gestational age: 33 weeks) to 246 mL/kg/min (gestational age: 36 weeks), and a small amount of pleural effusion and ascites was noted. The baby was delivered by Caesarian section with neonatologists and our team standing by at 37 weeks and 2 days of gestation. While the male neonate with apnea and bradycardia was resuscitated by the neonatologists, our team—divided into two subteams—scanned his left and right chest with ultrasound and noticed large fluid collection, and marked the appropriate puncture sites. Percutaneous tube drainage was performed under local anesthesia at the left and right anterior axillary lines at 2 and 6 minutes after birth,

respectively, using 6-Fr Argyle™ Trocar Aspiration Kits (Cardinal Health Inc., Dublin, OH, USA). Approximately 250 mL of serous content was drained soon after the procedures. While the content was drained, the neonate pinked up well. The Apgar scores at 1 and 5 minutes were 5 and 8 points, respectively. He was transferred to the Pediatric Intensive Care Unit in a stable condition. The drainage was minimal thereafter, and cytology indicated that the content was lymphocyte-dominant. The left and right mediastinal drains were removed at 2 and 9 days of age, respectively. His cardiopulmonary function was quite stable and he was weaned off the ventilator at 6 days of age. Contrast-enhanced computed tomography taken after the two drains were removed revealed a small amount of fluid collection in his anterior mediastinum. Follow-up studies with ultrasound and chest X-ray showed that the volume of mediastinal fluid collection was consistent, and he was discharged to home at 20 days of age. Treatment with Eppikajutsuto (TJ-28; TSUMURA & CO., Tokyo, Japan), at a dose of 0.2 g/kg/d, was started at 29 days of age, and the patient remained clinically stable without an increase in fluid volume during 20 months of follow-up.

Our literature search revealed only 13 pediatric cases with prenatally diagnosed lymphangioma that were mainly confined to the mediastinum.¹⁻⁷ The clinical features of the 14 cases, including ours, are presented in Table 1. The lesions in those cases were first detected in 20-35 weeks of gestational age. In our case, the lesion occupied more than half of the bilateral thoracic cavities and was the largest lesion reported in the literature. Any mass in the thoracic cavity in a fetus requires precise evaluation with fetal ultrasonography and MRI, and its differential diagnoses, other than lymphangioma, include congenital diaphragmatic hernia, teratoma, thymoma, pulmonary sequestration, and bronchogenic and neurogenic cyst.⁵ In our case, fetal MRI showed a large mediastinal cyst without a solid component, but did not visualize the septum, which could only be detected by ultrasonography.

Based on these findings, lymphangioma was the most likely diagnosis. The predominance of lymphocytes in the drained fluid supported this diagnosis. Hydrops fetalis was noted in two of the fetuses in the literature (cases 3 and 5), possibly due to the mass effect of the lesions. One of the fetuses required prenatal drainage of the lesion; the other required immediate intubation and surgery soon after birth. Our case did not develop hydrops fetalis; however, serial echocardiograms revealed a decreased cardiac output, possibly due to impaired dilatation. Such cases of mediastinal lymphangioma require careful attention during follow-up, especially when the lesion widely surrounds or causes the deviation of the heart. If the lesion had caused a fetal circulatory problem, intrauterine treatment with percutaneous drainage to reduce its volume would be necessary, as was performed in one of the reported cases (case 3).

The treatment strategies for the reported cases of prenatally diagnosed mediastinal lymphangioma varied. Our patient and three of the reported cases (cases 1, 5, and 12) developed respiratory distress and required ventilator support immediately after birth. The probability of postnatal respiratory decompensation can be assessed by the mass effect of the lesion on the airway, which would be visualized by fetal MRI. If the trachea is too narrowed or deviated for safe intubation, resuscitation including tracheostomy during *ex utero* intrapartum treatment (EXIT) should be planned, as is in cases of congenital upper airway anomaly. Resection was performed in six cases (cases 1, 5, 7, 9, 11, 12) at 1 day to 19 months of age. One of these cases required a second surgery to treat its recurrence (case 9). One of the resected cases developed diaphragmatic nerve palsy after surgery (case 3). Prenatal drainage followed by two resections in infancy (case 3) and sclerotherapy (cases 9 and 13) was performed in 1 and 2 cases, respectively. Two of the reported cases received no treatment for the lesion: one had stable disease thereafter (case 8), the other showed complete resolution within 6 months. The follow-up period of 20 months in our case is

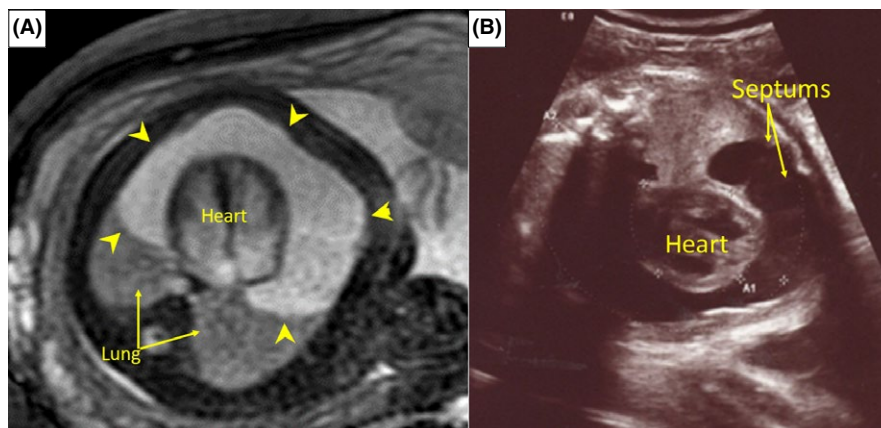


FIGURE 1 Fetal imaging studies at the time of the diagnosis of mediastinal lymphangioma (gestational age: 33 wk). (A) An axial view of fetal magnetic resonance imaging (B-TFE mode) showing a large lesion with water density and no solid component (arrowheads)

TABLE 1 The reported cases of prenatally diagnosed mediastinal lymphangioma

Case #	Year	Author	Gender	GA (wk) at diagnosis	Location	Size (cm) at prenatal diagnosis	Prenatal problem	GA (wk) at birth	Mode of delivery	Body weight at birth (g)	Problem after birth	Treatment	Outcome
1	1992	Zalel	Male	29	Between left atrium and spine	2.7*2.0*1.4	No	39	Spontaneous	2600 (first twin)	Respiratory distress and congestive heart failure, intubated	Resection at 40 d of age	Well at age of 3 mo
2	1993	Giactalone	Female	25	Posterior mediastinum	4*3.5	Increased size, mediastinal shift	N/A	Medical termination ^a	1360	N/A	N/A	N/A
3	1993	Muraskas	N/A	22	Anterior mediastinum	N/A	Cardiac displacement, hydrops fetalis	37	C/S	3880	No	Two prenatal US-guided percutaneous decompressions, and two resections at 5 d and 19 mo of age	Well at 6 mo after the second operation
4	1995	Wu	Male	31	Superior mediastinum	5.1*2.4	No (gradual decrease in size)	31	N/A	N/A	No	No	Complete resolution at 6 mo of age
5	2000	Jung	Male	23	Along right wall of heart	2.5*1.2	Increased size, hydrops fetalis	35	C/S	2800	Respiratory distress, intubated	Resection at 5 d of age ^b	Well at 18 mo of age
6	2002	Chen	Female	28	Anterior mediastinum over heart and upper lungs	3.8*4.8	Pleural effusion	38	C/S	2818	Nuchal cystic hygroma developed	No (partial excision of nuchal hygroma at 5 d of age)	Well at 7 mo of age with partial regression
7	2007	Bemasconi	N/A	20	Along the anterolateral aspect of the right heart	1.9*1.3	No	N/A	N/A	N/A	Slowly growing	Resection at 19 mo of age	N/A
8	2007	Ono	Male	28	Posterior mediastinum, surrounding thoracic descending Ao	4.6*2.6*3.0	No	39	C/S	2474	No	No	Stable, follow-up period N/A

(Continues)

TABLE 1 (Continued)

Case #	Year	Author	Gender	GA (wk) at diagnosis	Location	Size (cm) at prenatal diagnosis	Prenatal problem	GA (wk) at birth	Mode of delivery	Body weight at birth (g)	Problem after birth	Treatment	Outcome
9	2008	Comstock	N/A	25	Between heart apex and left chest wall, around SVC, upper mediastinum	3.3*0.7	No	37	C/S	N/A	No	Two resections at 6 wk and 4 mo of age	N/A
10	2008	Comstock	N/A	22	Upper right thorax, extending into right neck	N/A	Increased size	N/A	N/A	N/A	Slight deviated trachea, but no respiratory problem	Sclerotherapy with doxycycline twice	Asymptomatic at one year
11	2008	Ruano	Female	28	Left posterior mediastinum	5.9*4.0*3.6	No	39	C/S	3750	No	Resection at 5 d of age	Asymptomatic at one year
12	2012	Ballouhey	Male	34	Left hemithorax	N/A	No	N/A	Spontaneous	N/A	Respiratory distress, intubated	Resection at 1 d of age	Well
13	2012	Ballouhey	Male	35	Left hemithorax	N/A	No	N/A	Spontaneous	N/A	No	Sclerotherapy at 5 mo of age	Well
14	2018	Our case	Male	33	Anterior mediastinum	Occupied more than half of thoracic cavity	Decreasing cardiac output	37	C/S	3519	Respiratory distress, intubated	Percutaneous drainage right after birth	Stable at 20 mo of age

GA, gestational age; N/A, not available; C/S, Caesarian section.

^aDue to a possible poor prognosis.

^bComplicated with phrenic nerve palsy.

still short; however, the relatively stable postnatal course in the three cases may indicate that immediate postnatal radical surgery (as was performed in cases 3, 5, 11, and 12) might not be necessary for mediastinal lymphangioma.

Our case was the first in which planned percutaneous drainage was performed during postnatal resuscitation for mediastinal lymphangioma; this approach gave the treating team enough time to evaluate imaging and improve the cardiopulmonary function in preparation for a curative operation. Before delivery, we repeatedly held conferences with obstetricians and neonatologists, and planned the location of delivery, resuscitation of the baby, equipment for drainage (two sets), ultrasonography and the assignments of each of the personnel in a single operating room. An elaborate plan like this is essential for the successful rescue of a neonate with a life-threatening large mediastinal lymphangioma.

Eppikajutsuto (TJ-28), a Japanese herbal medicine, is reported to reduce redundant body fluids and have anti-inflammatory effects.⁹ Our literature search revealed that TJ-28 treatment led to the regression of lymphangiomas in 10 children; the locations included the head, neck, shoulder, chest,^{8,10} and retroperitoneum.⁹ The successful treatment of a mediastinal lymphangioma with TJ-28 has only been reported in a 2-year-old boy,¹⁰ in whom the lesion showed marked regression after 9 months of treatment. Although our follow-up period of 20 months is still too short to draw any conclusions with regard to the final outcome, we consider that TJ-28 may have contributed to stabilizing the size of the lesion in our case. We would treat the lesion by percutaneous drainage first if the lesion enlarges compromising cardiopulmonary function and may operate on it depending on the effectiveness of the drainage.

Thoroughly planned labor with immediate postnatal resuscitation and percutaneous decompression is mandatory for a large mediastinal lymphangioma that compromises the cardiopulmonary function, and close observation with the administration of TJ-28 may be a reasonable treatment, especially when resection of the lesion seems technically challenging.

INFORMED CONSENT

Informed consent was taken from the patient's parents for publishing the case.

AUTHORSHIP

HT and KM: wrote the manuscript. TA and HT: planned the perinatal management and analyzed the data. YS, KO, NS, and HK: conducted the postnatal surgical treatment with

analysis of the surgical specimens. All authors read and approved the final manuscript.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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