Concurrent cor triatriatum sinister and levoatriocardinal vein in an 11-year-old boy presenting with foudroyant pulmonary edema after appendectomy: A living tribute to the mal-incorporation theory

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Introduction

Cor triatriatum sinister (CTS) is a rare cardiovascular malformation, in which the left atrium (LA) is separated by a membrane into the proximal and distal LA chambers (1). Levoatriocardinal vein (LACV) is an even rarer vascular anomaly that connects the LA (or tributaries of pulmonary vein) with the left innominate vein (an embryologic derivative from the cardinal system) (2). Development of the pulmonary vein and systemic venous sinus are spatiotemporally correlated during embryogenesis (3). Here, we report a case of an 11-year-old boy unexpectedly presenting with acute pulmonary edema after appendectomy. Echocardiography and chest computed tomography (CT) revealed a combination of CTS and LACV, which were then surgically corrected. We briefly reviewed the English literature reporting the pediatric patients with concurrent CTS and LACV, compared their clinical profiles with ours, highlighted the pathomechanism of a delay onset of clinical manifestations in our patient, and provided an anatomical feature indicating the mal-incorporation theory for the embryogenesis of CTS.

Case Report

An 11-year-old boy was transferred to our hospital due to overt tachycardia and tachypnea two days after appendectomy, and admitted to the pediatric intensive care unit under the impression of acute cardiopulmonary failure. At the time of admission, his height was 135 cm, weight 61 kg, heart rate 126 bpm, respiratory rate 44 bpm, and blood pressure 141/93 mm Hg. Chest auscultation revealed moist rales. There was a grade II/VI systolic ejection murmur over the left upper sternal border. Preoperative chest radiogram showed only scoliosis (Fig. 1a). However, pulmonary edema was noted two days after appendectomy (Fig. 1b). The N-terminal pro-brain natriuretic peptide concentration was 2593 pg/mL (>450 pg/mL) and troponin-I concentration 0.09 ng/mL (>0.03 ng/mL). Two-dimensional echocardiography with color Doppler revealed a secundum atrial septal defect (ASD II), a limiting membrane in the LA indicating CTS (Fig. 1c), a 7.5-mm orifice in this membrane, and a pressure gradient of 10.8 mm Hg across the orifice (Fig. 1d). Pulmonary acceleration time was 80 ms, and pulmonary artery pressure 40 mm Hg. Chest CT showed a Lam type A1 CTS (Fig. 1e, 1f, and 1g) and a large LACV (Fig. 1h). After obtaining an informed consent, cardiovascular surgery was performed, including excision and removal of the limiting membrane within the LA, repair of the ASD II, and ligation of LACV. Follow-up chest radiogram and echocardiography showed regression of pulmonary edema and pulmonary hypertension, respectively. Meanwhile, normalization of N-terminal pro-brain natriuretic peptide and troponin-I were also achieved. The patient was discharged uneventfully.

Discussion

The most noteworthy point in the present case report is that there was a deep groove noted from the external aspect of the

Table 1. Seven pediatric patients with concomitant cor triatriatum sinister and levoatriocardinal vein

No/Age/Sex	Obstruction	Association	IAS	LACV		Outcome	Author (Reference)
				Origin	Drainage		
1/3 wk/F	CTS, CoA	CoA	ASD II	LA	INV	Expired	Pinto et al. (5)
2/4 wk/M	CTS	-	PFO	LA	RSVC	Survived	Bernstein et al. (2)
3/10 wk/F	CTS	-	PFO	LUPV	INV	Survived	Bernstein et al. (2)
4/3 mo/F	CTS	-	PFO	LA	RSVC	Survived	Bernstein et al. (2)
5/4 mo/F	CTS	-	Intact	LA	RSVC	Survived	Tosun et al. (4)
6/3 mo/F	CTS	VSD	Intact	LA	RSVC	Survived	Tosun et al. (4)
7/11 yr/M	CTS	-	ASD II	LUPV	INV	Survived	Lee and Tu (This report)

ASD II - secundum atrial septal defect; CoA - coarctation of the aortic arch; CTS - cor triatriatum sinister; F – female; IAS - interatrial septum; INV - innominate vein; LA - left atrium; LACV - levoatriocardinal vein; LUPV - left upper pulmonary vein; M – male; mo – month; PFO - patent foramen ovale; RSVC - right superior vena cava; VSD - ventricular septal defect; wk – week; yr - year

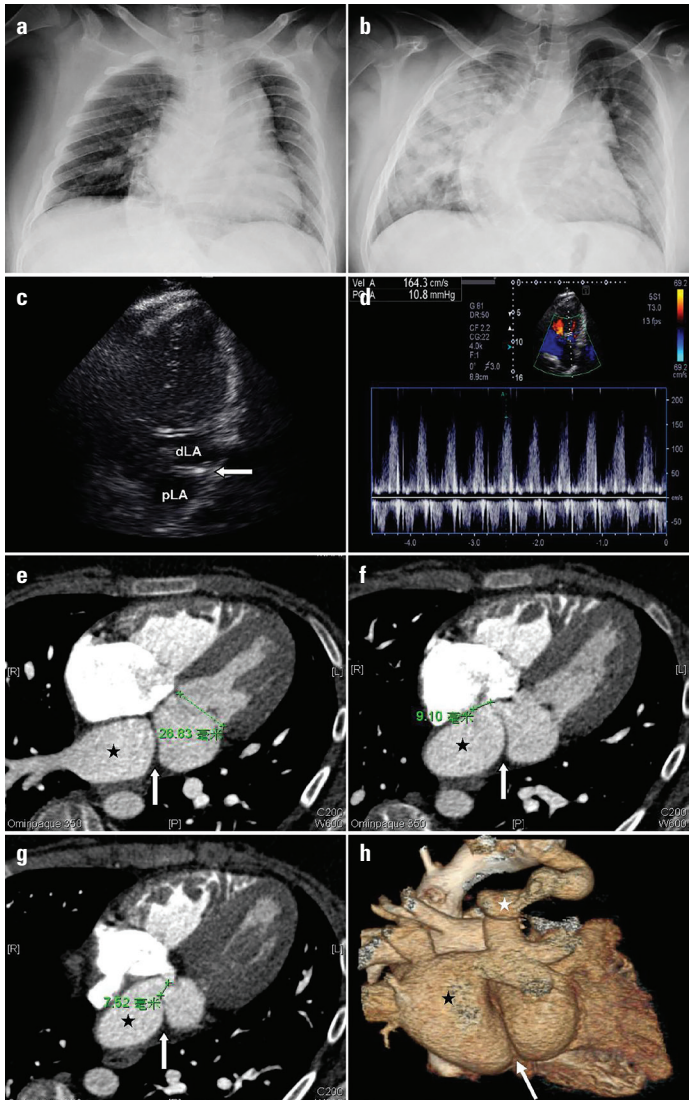

Figure 1. (a) Chest radiogram shows only scoliosis before appendectomy. (b) Chest radiogram shows pulmonary edema two days after laparoscopic appendectomy. (c) Two-dimensional echocardiography shows a membrane (arrow) that separates the left atrium (LA) into a proximal LA chamber (pLA) and a distal LA chamber (dLA). (d) Doppler echocardiography shows a pressure gradient of 10.8 mm Hg across the orifice of the limiting membrane. Chest computed tomography (CT) shows (e) a limiting membrane (white vertical arrow) that separates the left atrium into a proximal chamber (black star) and a distal chamber, and a 26.8-mm intact mitral valve, (f) a 9.1-mm secundum atrial septal defect between the proximal chamber (black star) and the right atrium, and (g) a 7.5-mm orifice in the limiting membrane (white vertical arrow), indicating the presence of Lam type A1 cor triatriatum sinister (CTS). (h) Three-dimensional chest CT, viewing from the posterior aspect, shows a large levoatriocardinal vein (white star) that connects the left upper pulmonary vein and the left innominate vein. This vein was measured approximately 20.0 mm in diameter at its junction with the left upper pulmonary vein and 10.0 mm at its junction with the left innominate vein. Of note is that, there is a deep groove (oblique white arrow) downstream the proximal LA chamber (black star). This external deep groove anatomically corresponds to the internal location of the limiting membrane downstream the proximal LA chamber (black star). This is a living tribute to the embryogenesis of the mal-incorporation theory indicating that CTS occurs due to a failure of incorporation of the common pulmonary vein into the primitive LA during the fifth embryonic week

chest CT. This external deep groove anatomically corresponds to the internal location of the limiting membrane that is downstream the proximal LA chamber. This is a living tribute to the embryogenesis of the mal-incorporation theory indicating that CTS occurs due to a failure of incorporation of the common pulmonary vein into the primitive LA during the fifth embryonic week (1). Albeit development of the pulmonary veins and systemic venous sinus are spatiotemporally correlated during embryogenesis (3), a combination of CTS and LACV is extremely rare (2). Till date, there are only six reported cases of pediatric patients with concomitant CTS and LACV in the English literature (2, 4, 5). The clinical and anatomical features of the seven pediatric patients, including our case, with concomitant CTS and LACV are tabulated in Table 1. Our patient, aged 11 years, was much older than the other six patients, who were aged from 3 weeks to 4 months. The clinical manifestations of our patient could be confounded by scoliosis, obesity, inadequate physical activity, low socioeconomic status, and reluctance to call for medical assistance, which may render a delayed diagnosis of CTS and/or LACV. In addition, three anatomical features were functionally contributive to decompress the LA, including a moderate-size ASD II (9.1 mm) above the membrane, a not-so-small orifice in the membrane (7.5 mm), and a large LACV (20.0 mm). Hemodynamically, there was a left-to-right shunt in our patient, which mimicked a shunt of a non-restrictive or large ASD II. Provided the pulmonary venous inflow is not significantly impeded, patients with concurrent CTS and LACV could be clinically asymptomatic. We highlight that obstructive left heart disease(s) should be scrutinized by echocardiography and/or chest CT in pediatric patients unexpectedly presenting with postoperative pulmonary edema or pulmonary hypertension.

Conclusion

Given that development of the pulmonary veins and systemic venous sinus are spatiotemporally correlated, a combination of CTS and LACV, though rare, is not impossible. Echocardiography and chest CT are useful examinations to unmask the Janus face of CTS and LACV in patients presenting with pulmonary edema and pulmonary hypertension.

Informed consent: Written informed consent was obtained from the parents of the patient for the publication of the case report and the accompanying images.

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