Magnetic resonance angiographic and three-dimensional computerized tomographic identification of scimitar syndrome in an 8-month-old infant

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Scimitar syndrome is a rare congenital abnormality. Clear anatomic definition is an important issue before operation. We report definition of both abnormal pulmonary venous drainage and anomalous systemic arterial supply by gadolinium-enhanced magnetic resonance angiography and three-dimensional computerized tomography in an eight month-old infant with scimitar syndrome. The presented case study confirms that these noninvasive diagnostic tools can successfully be used to provide valuable information about vascular anatomy in infants with scimitar syndrome.

Key words: scimitar syndrome, magnetic resonance angiography, three-dimensional computerized tomography, infancy.

Scimitar syndrome is a rare congenital abnormality consisting of partial abnormal pulmonary venous drainage from the right lung to the inferior vena cava with or without hypoplasia of the right lung, with dextraposition of the heart and anomalous systemic arterial supply from the abdominal aorta to a basal portion of the right lung[1]. Presentation in infancy is usually complicated with severe congestive heart failure and pulmonary hypertension[2].

The diagnosis of scimitar syndrome has usually been based on echocardiography and catheterization/angiography[1,2]. We report definition of both abnormal pulmonary venous drainage and anomalous systemic arterial supply by gadolinium-enhanced magnetic resonance angiography in an eight-month-old infant. The three-dimensional imaging reconstructed from the contrast-enhanced chest computed tomography also clearly showed both anomalous vascular connections in this patient.

Case Report

An eight-month-old girl presented with dyspnea, profuse sweating, feeding difficulty, and failure to thrive. Physical examination revealed growth retardation, tachycardia with gallop rhythm, tachypnea, congestion rales, and hepatomegaly. Chest X-ray showed prominent cardiomegaly with enlarged right atrium shadow and remarkable pulmonary venous congestion. There was no scimitar sign in chest X-ray. Electrocardiography revealed marked right QRS axis deviation, right atrial enlargement and right ventricular hypertrophy with strain. Two-dimensional and Doopler echocardiography demonstrated marked enlargement of right heart chambers with right ventricular hypertrophy, enlarged main pulmonary artery, and ostium secundum type (5 mm) atrial septal defect. Systolic pulmonary artery pressure was estimated as 74 mmHg from moderate degree tricuspid regurgitation. We could not delineate the connection of right pulmonary veins to the left atrium. Color flow echocardiography showed abnormal turbulent flow to the enlarged suprahepatic segment of the inferior vena cava.

Magnetic resonance angiography clearly demonstrated the scimitar vein, which drained whole segments of the right lung. Magnetic resonance angiography also revealed an aberrant

artery from the celiac trunk which was supplying the mediobasal segment of the right lung (Fig. 1). Computed tomography examination was planned to demonstrate parenchymal abnormalities and to exclude the pulmonary sequestration. The examination was performed using an 8-channel multidetector computerized tomography in angiography mode. MIP reconstructions of the images clearly delineated the presence of anomalous pulmonary veins forming two main branches and draining the middle and lower lobes totally and the upper lobe partially to the inferior vena cava at the subdiaphragmatic level, as seen in magnetic resonance angiography (Fig. 2). In addition to the vascular findings, computerized tomography examination also revealed a right upper lobe bronchial branching abnormality and fissure variations in both lungs.

Preoperative cardiac catheterization revealed severe pulmonary hypertension (pulmonary artery pressure: 83/36 (mean: 58) mmHg, pulmonary vascular resistance: 12.6 U/m^2), and a left-to-right shunt ratio of 1.6:1.0. Selective right pulmonary artery angiography showed scimitar vein draining into the inferior vena cava in levophase (Fig. 3a), and abdominal aortography demonstrated the aberrant artery supplying the part of the right lower lung lobe (Fig. 3b).

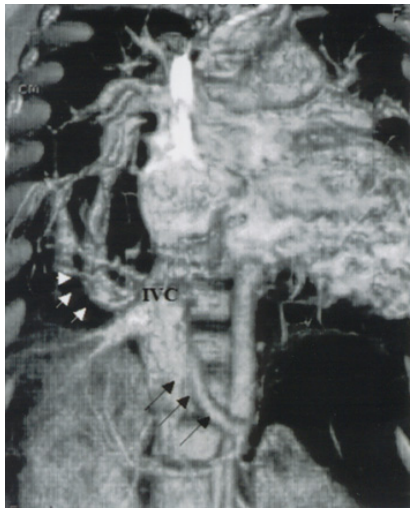

Fig. 1. Gadolinium-enhanced magnetic resonance angiography shows a scimitar vein (ScV) that drains the right lung into the enlarged suprahepatic segment of the inferior vena cava (IVC). An aberrant artery originating from the abdominal aorta (Ao) supplies the right lower lobe (left); horizontal view demonstrates the connection of the ScV to the IVC just posterior to the right hepatic vein (RHV) (right). MHV: middle hepatic vein, LHV: left hepatic vein.

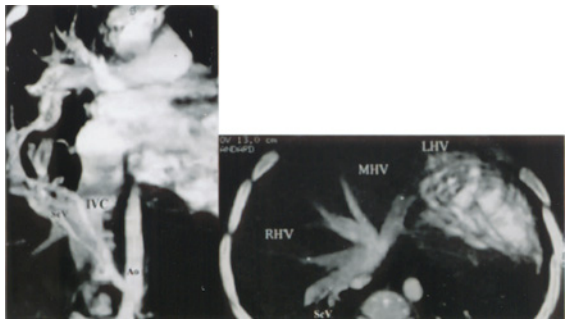

Fig. 2. Three-dimensional imaging in the anteroposterior view, reconstructed from contrast-enhanced computed tomography clearly shows a scimitar vein connected to the inferior vena cava (IVC) (white arrows) Note an anomalous systemic artery from the abdominal aorta, which supplies the right lower lobe (black arrows).

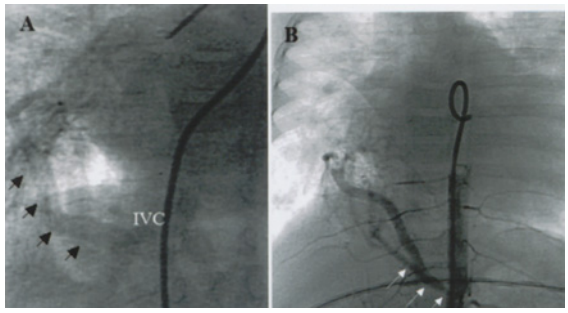

Fig. 3. a) Selective right pulmonary artery angiography shows scimitar vein draining into the inferior vena cava (IVC) in levophase in the anteroposterior view (arrows). b) Abdominal aortography in the anteroposterior view shows an anomalous systemic artery originating from the celiac trunk, dividing into two branches in its subsequent course, suppliying the right lower lobe (arrows).

The repair of the anomalous pulmonary venous return was carried out by repositioning abnormal vein from its point of entry into the inferior vena cava through the right atrium to the left atrium by the way of an enlarged atrial septal defect. Ligation of the aberrant artery supplying the right lower lobe was also accomplished during the operation.

Discussion

Patients with scimitar syndrome presenting in infancy usually suffer from severe congestive heart failure and severe pulmonary hypertension and should be operated without delay. Clear anatomic definition is crucial preoperatively in scimitar syndrome[2,3]. Although diagnosis of scimitar syndrome is

carried out by two-dimensional and Doppler echocardiography, abnormal systemic arterial supply is easily overlooked. The incidence of anomalous systemic arterial supply varies and it must be identified before operation since it might aggravate the degree of left-to-right shunt and the degree of pulmonary hypertenison[1-3].

To the best of our knowledge demonstration of both an anomalous pulmonary venous drainage and an abnormal systemic arterial supply with either magnetic resonance or computerized tomographic angiography was not previously performed in an infant with scimitar syndrome. Both gadolinium-enhanced magnetic resonance angiography[4] and contrast-enhanced computerized tomography of the chest[5] have been used previously to evaluate scimitar syndrome in adults. The presented case study confirms that these non-invasive diagnostic tools can successfully be used to provide valuable information about vascular anatomy in infants with scimitar syndrome and they may replace conventional angiography.

REFERENCES

1. Huddleston CB, Exil V, Canter CE, Mendeloff EN. Scimitar syndrome presenting in infancy. Ann Thorac Surg 1999; 67: 154-160.
2. Gao YA, Burrows PE, Benson LN, Rabinovitch M, Freedom RM. Scimitar syndrome in infancy. J Am Coll Cardiol 1993; 22: 873-882.
3. Najm HK, Williams WG, Coles JG, Rebeyka IM, Freedom RM. Scimitar syndrome. Twenty years' experience and results of repair. J Thorac Cardiovasc Surg 1996; 112: 1161-1168.
4. Gilkeson RC, Lee JH, Sachs PB, Clampitt M. Gadolinium-enhanced magnetic resonance angiography in scimitar syndrome diagnosis and postoperative evaluation. Tex Heart Inst J 2000; 27: 309-311.
5. Inoue T, Ichihara M, Uchida T, Sakai Y, Hayashi T, Morooka S. Three-dimensional computed tomography showing partial anomalous pulmonary venous connection complicated by the scimitar syndrome. Circulation 2002; 105: 663.