Congenital Aortocaval Fistula to the Superior Vena Cava : A Case Report

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Abstract

Various systemic arteriovenous fistulas have been described. The arteriovenous fistula arising from the ascending aorta and draining separately into the superior vena cava is very uncommon. The authors report a case of congenital aortocaval fistula to the superior vena cava in a 22 year-old woman in whom the fistula was closed successfully.

Key word : Fistula, Aortocaval

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Aortocaval fistula is an abnormal communication between the aorta and vena cava. The communication between the abdominal aorta and inferior vena cava (IVC) has been described in patients with ruptured abdominal aortic aneurysm, mycotic aneurysm and trauma[1-3]. The superior vena cava (SVC) is an uncommon site of drainage of this fistula. The authors report a case of congenital aortocaval fistula draining to SVC.

A 22 year-old woman was admitted for cardiac evaluation. She had been in a normal state of health until 6 months before admission, when she

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began experiencing shortness of breath and chest discomfort. She denied having had palpitations, orthopnea and paroxysmal nocturnal dyspnea. She had no history of chest trauma and did not recall being febrile. Her family history was unremarkable and she had not had any significant childhood illness.

Physical examination revealed a young woman in no distress. There was no clubbing, cyanosis, or edema. She was afebrile. The heart rate was 80 bpm and regular, and her blood pressure was 140/70 mmHg. All peripheral pulses were normal. Cardiac findings revealed mild cardiomegaly, normal S_1 and S_2 and a grade IV/VI continuous murmur in the second and third right intercostal spaces. The initial clinical diagnosis was a ruptured sinus of valsalva or coronary arteriovenous fistula. An electrocardiogram showed left ventricular hypertrophy as per voltage criteria. A chest X-ray revealed left ventricular hypertrophy, enlarged pulmonary trunk and increased pulmonary vasculature (Fig. 1). Transthoracic and transesophageal echocardiography showed an abnormal extracardiac continuous color and Doppler flow. The origin of this abnormal flow was not demonstrated by echocardiography but the left and right ventricles were enlarged. Oxymetry showed increased oxygen saturation of 90.3 per cent in SVC. Qp/Qs was 1.84 : 1. The coronary arteries were normal.

The communication between ascending aorta and superior vena cava was demonstrated by aortogram on the left anterior oblique at 45 degree. The origin of the fistula was near the brachiocephalic trunk (Fig. 2). The aortocaval fistula was demonstrated pre-operatively using gadolinium-enhanced, three dimensional magnetic resonance angiography (MRA) (Fig. 3). The origin of this fistula was 6 cm

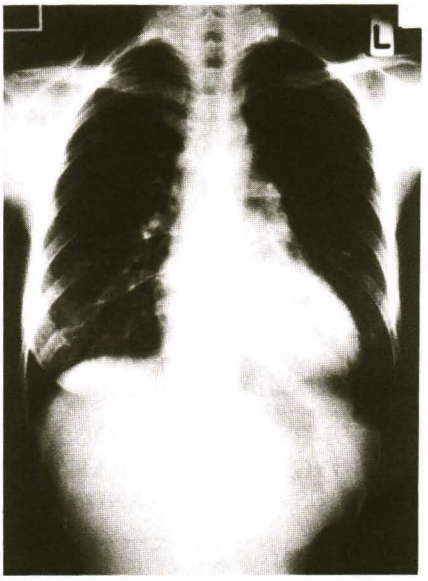

Fig. 1. Chest X-ray reveals mild cardiomegaly and increased pulmonary vasculature.

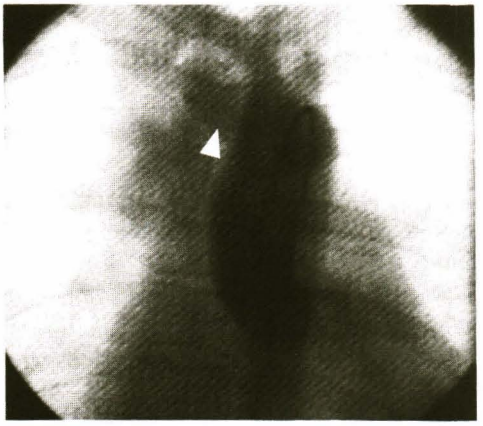

Fig. 2. Aortogram shows a fistula between the ascending aorta and superior vena cava (arrow), which is very dilated.

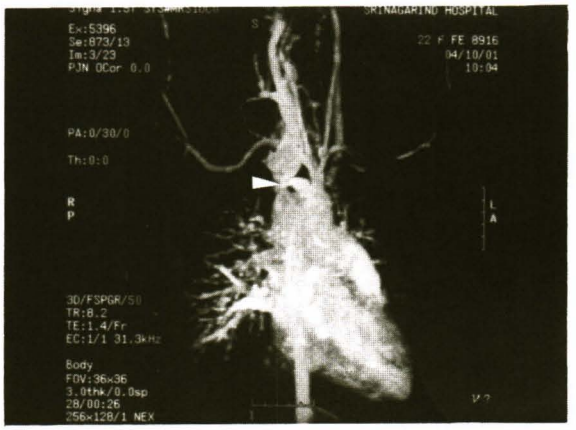

Fig. 3. MRA demonstrates a fistula between the ascending aorta and SVC (arrow).

above the aortic root. The major branches of the aortic arch including the brachiocephalic trunk, left common carotid artery and left subclavian artery were normal.

Intra-operatively, the SVC and right atrium were dilated. A 10-mm in diameter fistula connecting the SVC to the ascending aorta was visible. The SVC end of the fistula was excised and closed by direct suture. The other end of the fistula was excised from the ascending aorta and the aortic wall was subsequently repaired by vascular graft. Post-operative course was uneventful and the patient was discharged on day 7.

DICUSSION

Communication between the aortic root and the right heart chambers has been described and is commonly a result of a coronary arteriovenous fistula or a ruptured sinus of valsalva. Most of these fistulas drain into the right atrium, right ventricle, or coronary sinus; hence fistulous communication to the pulmonary artery, left heart chambers, and SVC is less frequent. Indeed, an aortocaval fistula is an unusual vascular abnormality, which can drain into either the SVC or IVC. Physical findings are similar to those in patients with patent ductus arteriosus and coronary artery-cardiac chamber fistula, except that the location is different.

Approximately 150 cases of aortocaval fistula to the inferior vena cava have been reported[1]; common causes being ruptured abdominal aortic aneuysm and trauma. Kuint et al reported a rare case of congenital fistula between the distal aspect of the descending aorta and inferior vena cava in a newborn[4].

Aorto-SVC fistula is even less frequent. Soler et al reported an unusual case of congenital systemic arteriovenous fistula between the descending aorta, azygos vein, and superior vena cava[5]. Morris et al described the vascular complications of dissecting thoracic aortic aneurysms such as aorto-SVC, and aorto-pulmonary arterial fistual formation[6]. Semiz et al reported a patient with an arteriovenous fistula between the brachiocephalic trunk and the superior vena cava- the result of a gunshot wound[7]. Oomman reported a case of congenital aortocaval fistula to the SVC[8]; in which the origin of fistula the was just above the right coronary sinus and was demonstrated pre-operatively by echocardiography.

The presented case had distinctive anatomical features; namely that the fistula originated in the ascending aorta near the origin of the brachiocephalic trunk. This was very difficult to demonstrate by echocardiography but efficiently evaluated by MRA. The authors conclude that MRA may be a promising method for evaluating aorto-SVC fistula.

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ทางติดต่อระหว่างเอออร์ตา และหลอดเลือดดำ superior vena cava ที่เป็นมาแต่-กำเนิด : รายงานผู้ป่วย

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Aortocaval fistula to the superior vena cava เป็นความผิดปกติที่พบได้น้อยมากในทางคลินิก รายงานผู้ป่วยหญิงไทย อายุ 22 ปี มีอาการสำคัญคือเหนื่อยมา 6 เดือน ตรวจร่างกายพบว่ามี continuous murmur ที่ช่องระหว่างกระดูก-ซี่โครงที่ 2 และ 3 ทางด้านขวา การตรวจด้วยคลื่นเสียงสะท้อนหัวใจไม่สามารถให้การวินิจฉัยโรคได้ การตรวจสวนหัวใจและเอ็ม อาร์ไอ พบว่ามีทางติดต่อระหว่างเอออร์ตา และหลอดเลือดดำ superior vena cava ผู้ป่วยมีอาการดีขึ้นหลังได้รับการผ่าตัด

คำสำคัญ : ทางติดต่อ, เอออร์ตา, หลอดเลือดดำ superior vena cava

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