□ Case report □

A case of congenital ductus arteriosus aneurysm

Sheng Wen Wang, M.D., Ji Eun Kim, M.D.
Young Seok Lee, M.D. and Young Ah Lee, M.D.

Department of Pediatrics, Dong-A University College of Medicine, Busan, Korea

Aneurysmal dilatation of the ductus arteriosis has been considered a rare but potentially fatal abnormality. The mechanism of ductal aneurysmal formation remains uncertain. Plain chest radiography has proven helpful in the diagnosis of ductus arteriosus aneurysm (DAA), before the application of transthoracic echocardiography. The transthoracic echocardiography is an important tool for the diagnosis and follow-up of DAA. We present a case of congenital ductus arteriosus aneurysm in a newborn, that was an incidental discovery. The diagnosis was made by echocardiography, three-dimensional surface rendering computed tomography (CT), and spontaneous regression after four weeks of follow-up. **(Korean J Pediatr 2006;49:1363-1366)**

Key Words : Patent ductus arteriosus, Aneurysm

Introduction

Aneurysms of the thoracic aorta have been reported in infants and children in association with the Marfan syndrome, coarctation of the aorta, valvular aortic stenosis, arteritis, Turner syndrome, and tuberous sclerosis[1]. Ductus arteriosus aneurysm (DAA) is a rare lesion that can be associated with severe complications including thromboembolism, rupture and death. Dyamenahalli et al[2] suggested an incidence of congenital DAA of 1.5% by reviewing 200 consecutive third trimester fetal ultrasounds. Jan et al[3] reported 48 (8.8%) patients with congenital DAA detected by echocardiography in 548 fullterm infants. The mechanism of ductus aneurismal formation remains uncertain; an altered circulation and weakening of the wall of the ductus arteriosus may lead to aneurismal formation. Transthoracic echocardiography is still an important tool for the diagnosis and follow-up of DAA. Other imaging techniques such as magnetic resonance imaging or computed topographic scanning are helpful for the diagnosis of DAA, and adjacent vascular structure and complications. In Jan's study[3], follow-up echocardiograms detected spontaneous closure of all patients with DAA in one month of life, with the exception of one patient. However, it may be appropriate to consider early intervention for the larger DAA in patients with connective tissue diseases and in the setting of significant symptoms or complications associated with the DAA.

We report a case of congenital ductus arteriosus aneurysm confirmed by 3-dimensional reconstructed CT, which was spontaneously resolved after a four weeks of life.

Case Report

A male neonate with a gestational age of 42 weeks and a birth weight of 3140 g was admitted to our hospital because of cyanosis, tachypnea and meconium-stained amniotic fluid.

Physical examinations revealed a moderate chest retraction and a coarse breathing sound, but no cardiac murmurs were detected. Laboratory studies failed to reveal any abnormality, except a mild metabolic acidosis. Chest radiography demonstrated infiltrations and consolidations on both lung fields and a mass shadow in the aortopulmonary window(Fig. 1). He was diagnosed as meconium aspiration pneumonitis. Oxygen therapy was then started and the patient was treated with empirical antibiotics. Transthoracic echocardiography was performed after the second day of life, showed a moderate secundum atrial septal defect with a left-to-right shunt. The patent ductus arteriosus was

접수 : 2006년 7월 18일, 승인 : 2006년 9월 15일
책임저자 : 이영아, 동아대학교 의과대학 소아과학교실
Correspondence : Young Ah Lee, M.D.
Tel : 051)240-5124 Fax : 051)242-2765
E-mail : yalee@dau.ac.kr

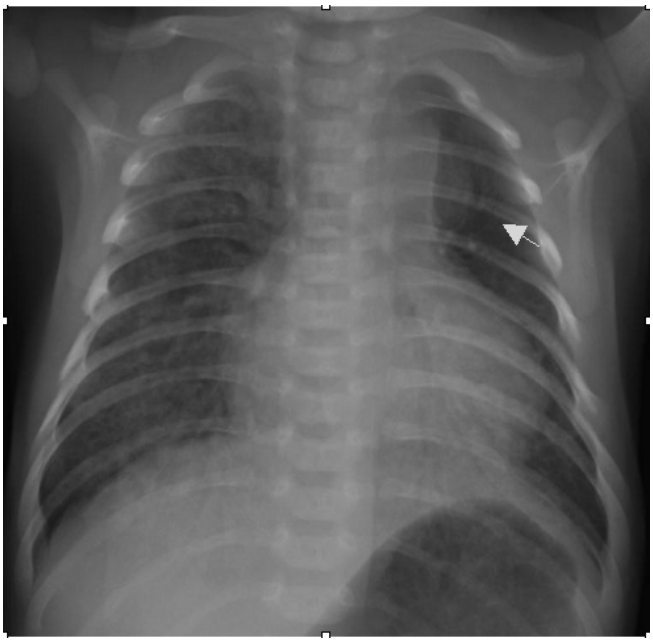

Fig. 1. Chest radiography obtained 1st day. Diffuse hazziness of both lung fields with air bronchogram, diffuse pneumonia and mass shadow (arrow) in the aortopulmonary window.

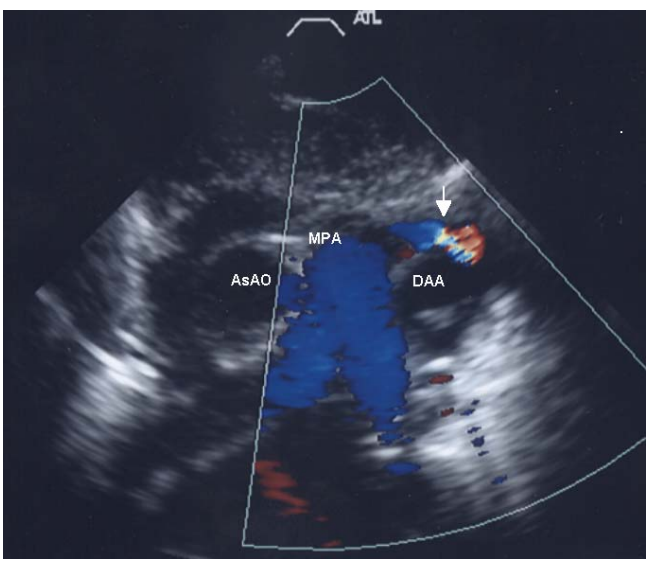

Fig. 2. Transthoracic echocardiography was performed after second day of his life. Color Doppler mapping shows a ductal shunt jet (arrows) from saccular dilation of ductus arteriosus to the main pulmonary artery in a parasternal short-axis view. Abbreviation : DAA, ductus arteriosus aneurysm; AsAO, ascending aorta; MPA, main pulmonary artery.

interpreted as a dilated left pulmonary artery (15×16 mm) (Fig. 2).

A computed tomography (CT)-scan of the chest with contrast was done on the fourth day after birth. Axial section CT images revealed a vascular shadow at the

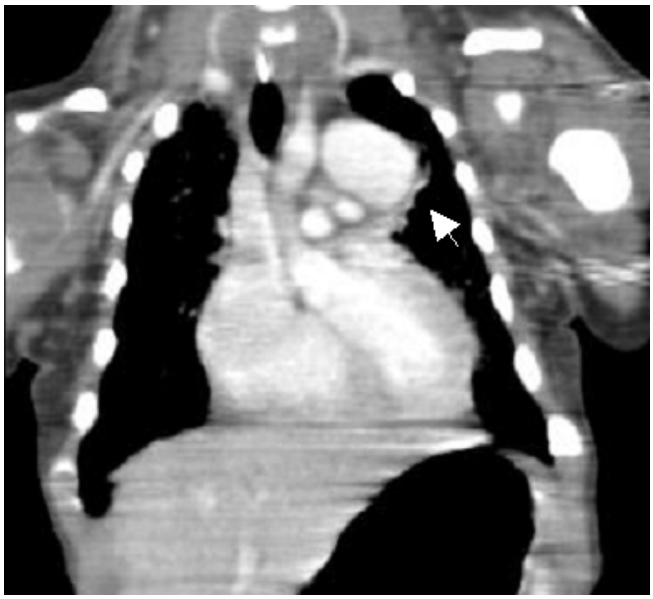

Fig. 3. Contrast enhanced CT in axial obtained four days after birth. A vascular shadow (arrow) at the arotopulmonary window.

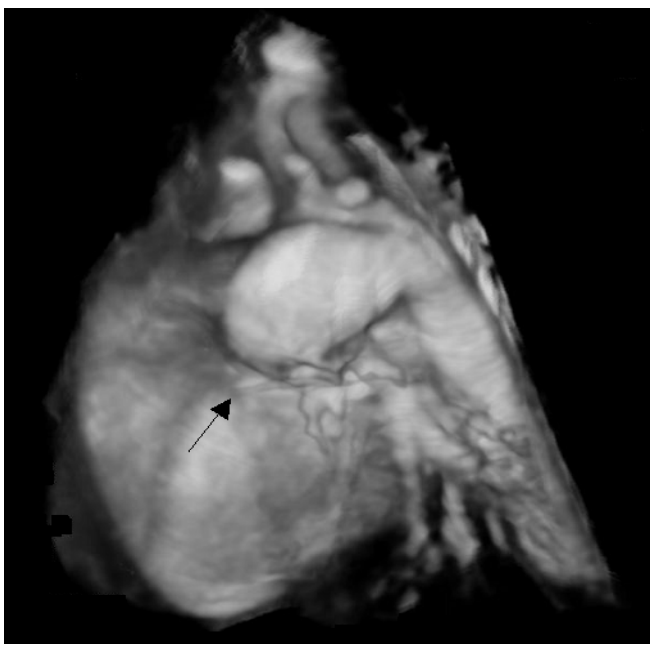

Fig. 4. 3D reconstructed CT obtained four days after birth. A large aneurysm (arrow) is seen.

aortopulmonary window (Fig. 3). Trans section CT images revealed a diffuse patchy consolidation with air bronchogram on both lung fields. Three-dimensional reconstructed CT images showed an aneurysm, connecting between the aorta and the left pulmonary artery (Fig. 4). Follow up transthoracic chocardiography was performed on day 9, showing the patent ductus arteriosus was closed, and the

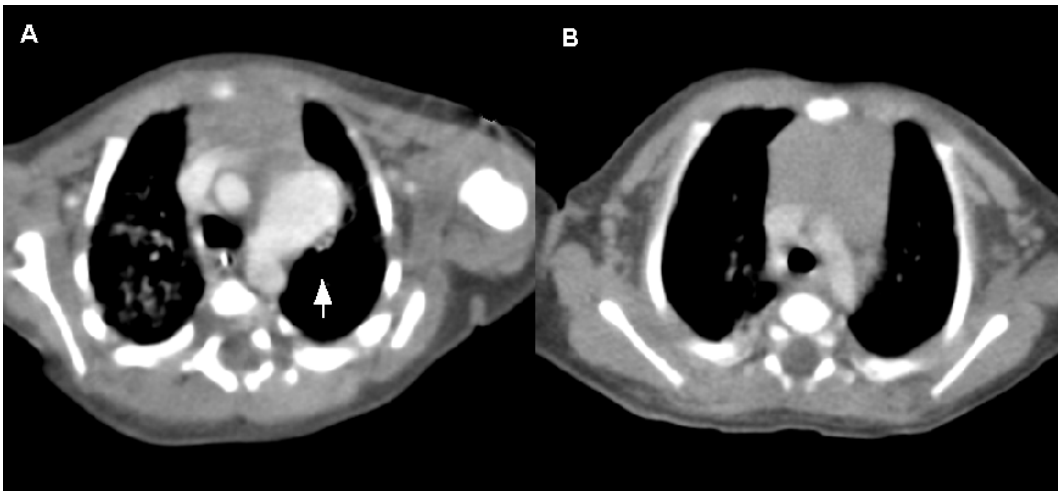

Fig. 5. Contrast-enhanced CT in trans section obtained 4 days after birth. A vascular shadow (arrow) at the aortopulmonary window is seen (A). Follow up contrast-enhanced CT in trans section obtained 28 days after birth, revealed rapid regression of the aneurysm (B).

aneurysmal dilatation was decreased in size (12×13 mm).

On day 10, the antibiotics were stopped, but a mild chest retraction still remained. After 2 weeks of life, tachypnea and the chest retraction disappeared, and oxygen therapy was discontinued. Follow-up CT-scans obtained at four weeks after his birth, revealed regression of the aneurysm, and nearly total resolution of bilateral pneumonia (Fig. 5).

Discussion

An aneurysm of the ductus arteriosus (DAA) results from a local dilatation of the vessel, or from an enlargement of the remaining ductal tissue[4]. The incidence of infantile aneurysm of ductus arteriosus is not clear, but Dyameneahalli et al[2] and Jan et al[3] suggested 1.5-8.8%.

The mechanism of ductal aneurysmal formation remains uncertain, and there are several theories about its pathogenesis. One theory is that delayed closure at the aortic end of the ductus arteriosus, exposes the ductal wall to systemic pressure, which may lead to aneurysm formation[4]. However, this mechanism is not likely in fetal or early neonatal cases when pressure in the pulmonary artery is higher or similar to that in the aorta. A second theory states that the congenital wall weakness may result from necrosis and mucoid degeneration of the media in the ductal wall[4], but the same changes can be found in normal-closing ductal tissue[5, 6]. A third theory is that the increased flow through the ductus arteriosus in the uterus may lead to aneurysmal formation[3]. The fourth theory is that the intrauterine ductal constriction may cause post-stenotic dilatation of the ductus arteriosus[5]. Finally, it may be the result of an abnormal intimal cushion formation or a defective elastin in the ductus aeteriosus[2]. However, the definitive pathogenesis of DAA needs further investigation.

Dyameneahalli et al[2] reported a prenatal diagnosis of nine patients with DAA. All of them were asymptomatic, and Jan et al[3] reported 48 patients with DAA, the same as Dyameneahalli et al, and all of them were asymptomatic. But, Lund et al[7] reviewed 61 cases of DAA and revealed that up to 30% of DAA was associated with lethal complications, including thromboembolism, rupture, dissection, and infection[7]. A few reports describe compression of adjacent structures such as recurrent laryngeal or phrenic nerve and left main bronchus[8].

Radiological diagnosis of a DAA begins with the chest radiograph which shows a soft tissue, mediastinal mass-like shadow in the aortopulmonary window, also known as "ductal bump". Persistence or enlargement of a mass in the aortopulmonary window beyond the 3 days of age suggests the possibility of a ductal aneurysm[8]. With increasing utilization of echocardiography, more cases can be discovered based on findings on echocardiograms[9]. Conventional chest CT shows that the mass is well enhanced and connected to both the aorta and the pulmonary artery. However, the exact relationship of a ductal aneurysm with the aorta and pulmonary artery can only be inferred from axial section CT images. After reconstruction with 3D rendering, we can easily identify the spatial location of the aneurysm[10].

The use of oral or intravenous indomethacin has led to successful closure of PDA in premature infants[11], but it is ineffective in full-term infants. Although one case report demonstrated the regression of ductal aneurysm after indomethacin treatment[10], the definitive effect of indomethacin on the ductal aneurysm is uncertain. Lund et al[7] suggested surgical resection if the aneurysm does not regress within a few days because of a high incidence if complications. Usually sugical resection of DAA was suggested if any of the following conditions existed: 1) a PDA and DAA beyond the newborn period, 2) an associated connective tissue disorder, 3) a thrombus within the DAA with extension to adjacent vessels, 4) evidence of thromboembolism, or 5) functional compromise of adjacent structures due to pressure effects[2].

Jan et al[3] suggested spontaneous closure of DAA may be a normal process in full-term infants. An unnecessary operation should be avoided if regression is confirmed. Surgical intervention may be delayed until the ductal aneurysm persists beyond the neonatal period or early complication develop. Continued echocardiograpic follow-up of affected infants is suggested.

한 글 요 약

Congenital ductus arteriosus aneurysm 1례

동아대학교 의과대학 소아과학교실

왕승문 · 김지은 · 이영석 · 이영아

신생아에서 흉강내 대동맥류는 드물며, 말판 증후군, 대동맥 축착, 대동맥 판막 협착, 동맥염, 터너 증후군, 결절성 경화증 등과 연관되어 보고된 바 있다.

신생아에서 발견되는 ductus arteriosus aneurysm은 매우 드문 질환이나, 대동맥 파열이 유발 될 수 있는 기형이다. 병인으로 대동맥 쪽에 있는 동맥관의 폐쇄 지연, 선천적으로 동맥관 벽이 약한 경우, 자궁내에서 동맥관으로 혈류 증가, 자궁내에서 동맥관의 협착 등이 있으나 명확하지 않은 상태이다.

Ductus arteriosus aneurysm은 흉부 방사선, 심초음파, 3차원 전산화단층촬영, 자기 공명영상 검사 등으로 진단할 수 있다. 치료는 인도메사신의 투여, 수술적 절재 등의 방법이 있으며, 자연 소실되는 경우도 있다.

저자들은 태변흡인증후군 환아에서 우연히 발견된 ductus arteriosus aneurysm이 생후 4주 뒤 자연 소실되는 1례를 경험하였기에 보고하는 바이다.

References

1) Chien YH, Sung TC. Aneurysm of the ascending aorta in a neonate. Acta Paediatr Taiwan 1999;40:121-3.
2) Dyamenahalli U, Smallhorn JF, Geva T, Fouron JC, Cairns P, Jutras L, et al. Isolated ductus arteriosus aneurysm in the fetus and infant: a multi-institutional experience. J Am Coll Cardiol 2000;36:262-9.
3) Jan SL, Hwang B, Fu YC, Chai JW, Chi CS. Isolated neonatal ductus arteriosus aneurysm. J Am Coll Cardiol 2002;39:342-7.
4) Lund JT, Jensen MB, Hjelms E. Aneurysm of the ductus arteriosus. A review of the literature and the surgical implications. Eur J Cardiothorac Surg 1991;5:566-70.
5) Acherman RJ, Siassi B, Wells W, Goodwin M, DeVore G, Sardesai S, et al. Aneurysm of the ductus arteriosus: a congenital lesion. Am J Perinatol 1998;15:653-9.
6) Silver MM, Freedom RM, Silver MD, Olley PM. The morphology of the human newborn ductus arteriosus: a reappraisal of its structure and closure with special reference to prostaglandin E1 therapy. Hum Pathol 1981;12:1123-36.
7) Lund JT, Hansen D, Brocks V, Jensen MB, Jacobsen JR. Aneurysm of the ductus arteriosus in the neonate: three case reports with a review of the literature. Pediatr Cardiol 1992;13:222-6.
8) Das BB, Solowiejczyk DE, Kamath V, Sharma J. Congenital ductus arteriosus aneurysm presenting with stridor in a newborn. J Perinatol 2002;22:179-80.
9) Malone PS, Cooper SG, Elliott M, Kiely EM, Spitz L. Aneurysm of the ductus arteriosus. Arch Dis Child 1989;64:1386-8.
10) Lee WJ, Chen SJ, Wu MH, Li YW. Regression of ductus arteriosus aneurysm in a neonate demonstrated by three-dimensional computed tomography. Int J Cardiol 1999;68:231-4.
11) Gersony WM, Peckham GJ, Ellison RC, Miettinen OS, Nadas AS. Effects of indomethacin in premature infants with patent ductus arteriosus: results of a national collaborative study. J Pediatr 1983;102:895-906.