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Clinical vignette

Congenital abdominal aortic aneurysm in a term neonate: a case report

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Background: Congenital abdominal aortic aneurysm (AAA) is a rare condition in neonates. To our knowledge, the natural course of the disease in a Thai neonate has not yet been reported.
Objectives: To report the characteristics and clinical course of congenital AAA in a Thai neonate.
Methods: A female Thai infant was born spontaneously at term (3,990 g) having a large, pulsatile, abdominal mass. Computed tomographic angiography (CTA) of the abdominal aorta showed a large infrarenal AAA, and a fusiform aneurysm at the left common iliac artery. Two small right renal arterial aneurysms were also noted. The large aneurysm was partially resected and a Gore-Tex vascular graft was placed at 15 days old. Histopathology of the aneurysmal wall revealed no specific etiology. Ultrasonography revealed thrombosis of the graft on the 13th day after surgery.
Results: Repeated CTA of the abdominal aorta at age 13 mo showed complete thrombosis of the graft with reconstitution of collateral circulation. The infrarenal AAA and left common iliac aneurysm and 2 small right renal artery aneurysms were completely thrombosed. The patient grew and developed normally to the most recent follow up at age 36 mo.
Conclusions: Congenital AAA with failure of graft replacement may regress spontaneously.

Keywords: Abdominal aortic aneurysm, neonate, aortic aneurysm, idiopathic abdominal aortic aneurysm, neonatal abdominal aortic aneurysm, pulsatile abdominal mass

Congenital abdominal aortic aneurysm (AAA), a rare condition in neonates, is usually associated with umbilical artery catheterization [1, 2] or other conditions such as congenital heart or aortic malformation, infection (mycotic aneurysm) [3, 4], connective tissue diseases (Marfan syndrome, Ehlers–Danlos syndrome, Loeys–Dietz syndrome) [5, 6], or vasculitis (Takayasu's disease, Kawasaki syndrome) [7]. An idiopathic congenital AAA is extremely rare. We report a case of idiopathic congenital AAA consisting of multiple lesions in a Thai neonate born at term, who survived after a partial surgical correction. To our knowledge, this case is the first idiopathic abdominal aortic aneurysm in a Thai neonate reported.

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Case report

A female infant was born spontaneously at term (3,990 g) to a 19-year-old Thai mother with Apgar scores of 9 and 9 at 1 and 5 minutes, respectively. The mother, known to have a hemoglobin E trait, had an uneventful pregnancy, and received regular antenatal care. The Thai father of the neonate had suffered from hemophilia A disease. There was no reported family history of consanguinity.

On admission to the nursery, marked abdominal distension with a large pulsatile mass approximately 10 cm in diameter was noted at midabdomen. Faint bruit was heard over the mass area. Blood pressures measured on the arm and leg were 77/39 mmHg and 67/41 mmHg, respectively. Heart rate was 132 beats per min and regular. Both femoral pulses were weak. Other physical findings were unremarkable. Initial laboratory results showed hemoglobin 14.6 g/dL, white blood cell (WBC) count 8,888 cells/mm^3 (neutrophils 53%, lymphocytes 39%, monocytes 6%, and eosinophils 2%), nucleated red blood cells 7/100 WBC,

platelet count 229,000/μL, prothrombin time 14.8 s, activated partial thromboplastin time 39.6 s, international normalized ratio 1.33, blood urea nitrogen 7 mg/dL, and serum creatinine 0.7 mg/dL. Blood culture result showed no growth after 48 h.

Bedside ultrasonography of abdomen found a large cystic mass with flow inside the mass, a suspected aneurysm. Hydronephrosis of her left kidney was also noted. Computed tomographic angiography of the abdominal aorta showed a large infrarenal AAA located approximately 3.6 mm below the right renal arterial origin down to aortic bifurcation measuring 5.4 cm × 9.1 cm × 4.4 cm. Another lesion was an 8.2 cm × 7.1 cm × 6.0 cm fusiform aneurysm at the left common iliac artery just distal to the aortic bifurcation. Two small right renal arterial aneurysms were also noted. Other findings included a 1 cm patent ductus arteriosus, severe left hydronephrosis, and moderate right hydronephrosis. Cardiac catheterization revealed a good left ventricle contraction with no evidence of coarctation of the aorta. The right femoral artery arose from the abdominal aneurysm (**Figures 1 and 2**).

When the patient was 15 days old, the largest aneurysm was partially resected and a Gore-Tex vascular graft was placed. The left common iliac aneurysm was not removed because of difficulty in identifying its distal part. Immediately after surgery, both lower extremities became cold to touch with weak femoral pulses. Doppler ultrasonography at 2 weeks after surgery revealed thrombosis of the graft. Blood flow in the right femoral artery was noted, which was believed to result from the collateral arteries. Left femoral blood flow could not be assessed. The right renal artery blood flow appeared normal. Daily heparin infusion was attempted to dissolve the thrombus for 1 week without success. Other postoperative complications included acute renal injury, renovascular hypertension, and a neurogenic bladder. All of these complications had resolved within 3 weeks. The infant's condition remained stable and she was discharged from hospital at age 56 days with only antibiotic prophylaxis for a urinary tract infection.

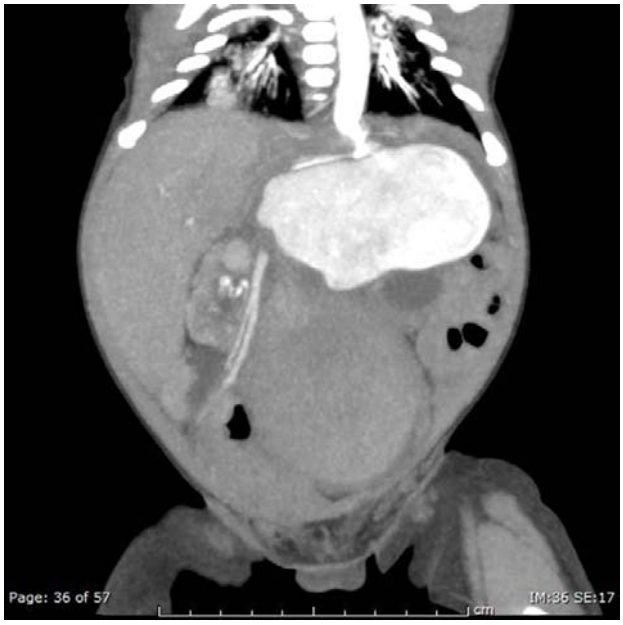

Figure 1. 2D image of the aorta in frontal projection during arterial phase reveals opacification of the more proximal abdominal aortic aneurysm. The contrast media has been diluted in this large aneurysm and has not yet passed to the distal vessels or another aneurysm.

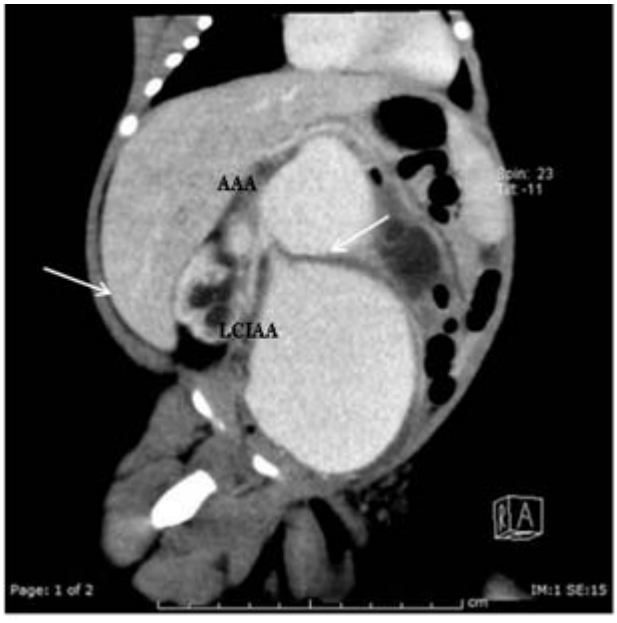

Figure 2. 2D coronal oblique plane of the abdomen in venous phase reveals an abdominal aortic aneurysm (AAA) and left common iliac artery aneurysm (LCIAA). Bilateral hydronephroses are noted (arrows).

The histopathological examination of the aneurysm wall revealed a marked reduction in elastic fiber in the tunica media with disorganization of the fibrocollagenous stroma. These findings were accompanied by attenuated medial muscle. The intimal surface was covered with fibrin strands with circumferential area of dystrophic calcification. There was no evidence of suppurative inflammation and necrosis. No features of mycotic aneurysm were noted. Elastin staining showed marked reduction and fragmentation of elastic fibers of the aneurysm wall (**Figure 3**). Left ureter was embedded to the removed aneurysm.

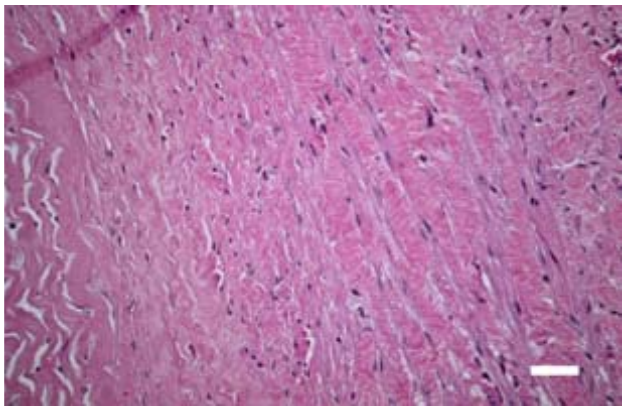

Figure 3. Histopathology of the aortic wall reveals marked reduction in elastic fiber and smooth muscle cells in the tunica media with disorganization of the fibrocollagenous stroma (scale bar 100 μm, original magnification 200×).

A follow-up CTA of the abdominal aorta when the patient was aged 13 mo showed complete thrombosis of the aortic graft at infrarenal abdominal aorta down to both common iliac, internal iliac and external iliac arteries, with reconstitution at bilateral distal external arteries from multiple collateral branches. There was a complete thrombosis of the left common iliac aneurysm, and two small right renal artery aneurysms. Right hydronephrosis was completely resolved. The left kidney was not clearly demonstrated. Physical examination at age 36 mo showed weak femoral pulses. The left leg was 1 cm shorter than the right leg. Mid-leg circumference of left leg was 1 cm smaller than right leg. Muscle tone and sensory of both legs were normal. Her development was appropriate for age.

This case report was assessed by Institutional Review Board of the Faculty of Medicine at Chulalongkorn University and exempted from full review (Certificate of Exemption No. 022/2016, IRB No. 490/59). Written informed consent was obtained from the mother of the patient to publish this case report.

Discussion

Idiopathic congenital AAA is extremely rare and the cause remains unknown. Common histological findings include intimal fibroplasia, which reflects a secondary process rather than an etiological factor. To date, only 26 cases of congenital AAA have been reported in the English literature to our knowledge [7]. Fourteen cases were diagnosed while the patient was a neonate; 7 cases were diagnosed after birth [7-13]; and 7 cases were diagnosed prenatally [14-20]. Of these 14 patients, 13 had isolated presentation of AAA, while 1 patient also had porencephaly [11]. The most common clinical manifestation was pulsatile abdominal mass. Most reported cases were infrarenal AAAs.

In the presently presented case, we could not identify the definite cause of her AAA. The infant had no signs of infection or systemic diseases such as joint hyperlaxity or vasculitis. The histopathological examination revealed vascular aneurysm with disorganization and reduction of elastic fibers. No evidence of mycotic aneurysm was found. Therefore, the pathology in this patient may be classified as idiopathic congenital AAA.

AAA has been described in Loeys–Dietz syndrome, which is an autosomal dominant aortic aneurysm syndrome with widespread systemic involvement. The syndrome is associated with heterozygous mutations in the genes encoding transforming growth factor-receptors 1 and 2 (*TGFBR1* and *TGFBR2*) [6] and characterized by the triad of arterial tortuosity and aneurysms, hypertelorism, and bifid uvula or cleft palate. Our patient did not have such findings.

Adult patients with hemophilia may develop AAA with histological findings indistinguishable from those of the AAA of nonhemophiliacs. Currently, the association between hemophilia and AAA remains unknown [21-23]. Our patient's pathology is unlikely related to the underlying hemophilia A disease in her father.

Management of AAA in neonates is challenging and involves both surgical and nonsurgical management. To date, there is no definite criteria for

surgical repair in neonates with AAA [6]. In surgical candidates, special considerations must be made for the timing and type of operation, and type of vascular graft. Successful surgical repair has been described in 5 cases; 2 with allograft [17, 19]; 2 with synthetic vascular graft [8, 15], and 1 with native vessels [9]. Our patient was managed with a synthetic vascular graft, which developed a complete thrombosis afterwards. Nonsurgical management for AAA in neonates has been reported in 7 patients with less success. Four of them died [7, 12, 13, 16], the aneurysm in one of them ruptured [12], and the 3 others died from renal and cardiac dysfunctions [7, 13, 16]. Transient renovascular hypertension was often found in patients with AAA. Most cases were successfully controlled with antihypertensive agents, which can be discontinued before discharge. Transient hypertension in our patient occurred immediately post partial resection of one of the abdominal aortic aneurysm. The hypertension responded to amlodipine. Her blood pressure returned to normal before discharge.

Among the 26 reported cases, there was 1 child with considerably healthy condition at age 39 mo [9], which was the longest follow up reported. At 27 mo old, our patient was still in fairly good health. No medication was needed. Although idiopathic congenital AAA is extremely rare among neonates and infants, it is associated with high morbidity and mortality. Like our patient, some can survive without serious sequelae. We consider that the thrombotic aortic graft might play a role in the resolution process of the aneurysm.

All patients with AAA with or without surgical repair need a long-term follow up. Postoperatively, patients should be monitored for high blood pressure, ischemic signs in the extremities, abnormal renal functions, and recurrence of AAAs. Until now, no studies have evaluated the long-term prognosis of idiopathic congenital AAA after surgical correction [6]. Those with conservative management also need a close monitoring of the aneurysm size, blood pressure, and for signs and symptoms of rupture of the aneurysm. Surgical repair might be required in patients who have failed conservative therapy. Our patient survived with minimal sequelae (shortness of one leg) despite the formation of complete thrombosis of the aortic graft. This demonstrates that the idiopathic congenital AAA of the neonate can spontaneously regress by 13 mo of age, with failure of graft placement. The development of multiple collateral circulation allowed the infant survive with a fairly normal life, at least on the last follow up at age 36 mo. However, a long-term close follow up of the present patient is required.

Author contributions

AS, SP, and PT conceived and designed of this report. AS, PT, and JN acquired the data. All authors contributed to data analysis and interpretation. AS, SP, and PT drafted the report, and all authors critically revised it, approved the final submitted version, and take responsibility for the statements made in the report.

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Conflict of interest statement

The authors declare that there is no conflict of interest regarding this case report.

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