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CASE REPORT

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Acute thrombosis of ductus arteriosus aneurysm causing bilateral pulmonary artery occlusion in a neonate

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Abstract

A 9-day-old male neonate was found to have a systolic murmur during a routine follow-up for skin jaundice. Imaging revealed a large mass at the bifurcation of the main pulmonary artery, causing significant bilateral stenosis. The patient underwent emergency surgery due to critically compromised pulmonary blood flow. Intraoperative exploration demonstrated a large ductus arteriosus aneurysm (DAA) with extensive thrombosis. The DAA was completely resected under deep hypothermic circulatory arrest, followed by reconstruction of the descending aorta and repair of the pulmonary artery bifurcation defect. The patient was discharged on the 20th postoperative day and remained asymptomatic at the 3-month follow-up, exhibiting normal growth and cardiac function.

Keywords Ductus arteriosus, Aneurysm, Thrombosis, Neonate, Diagnosis

Background

Ductus arteriosus aneurysm (DAA) can be classified as congenital or acquired and is characterized by a localized saccular or fusiform dilation of the ductus arteriosus. Congenital DAA is most commonly observed in late pregnancy and the neonatal period, with an incidence ranging from 0.8–8.8% [1, 2]. Most congenital DAAs are benign, with or without thrombosis and fibrosis, and may spontaneously regress or shrink [2]. However, approximately 30% of cases can be associated with severe complications such as thromboembolism, rupture, erosion, infection, and compression of adjacent structures(3, 4). This article presents a case involving a newborn with an incidental finding of a mass in the main pulmonary artery (MPA), which led to occlusion of both pulmonary arteries. The patient underwent emergency surgical repair and made a successful recovery, ultimately being discharged and showing significant improvement over the following three months.

Case presentation

A 9-day-old male neonate was transferred to our hospital because of an incidentally detected systolic murmur in his regular visit for skin jaundice at a district clinic. Echocardiography showed a large irregular mass located at the bifurcation of the main pulmonary artery (MPA), protruding into the lumen, causing bilateral stenosis and increased flow velocity, with left PA of 3 m/s and right PA of 3.2 m/s, respectively (Fig. 1 Panel A). Cardiac computed tomography (CCT) revealed that the mass exhibited homogenous density without contrast enhancement, making it difficult to differentiate from the surrounding

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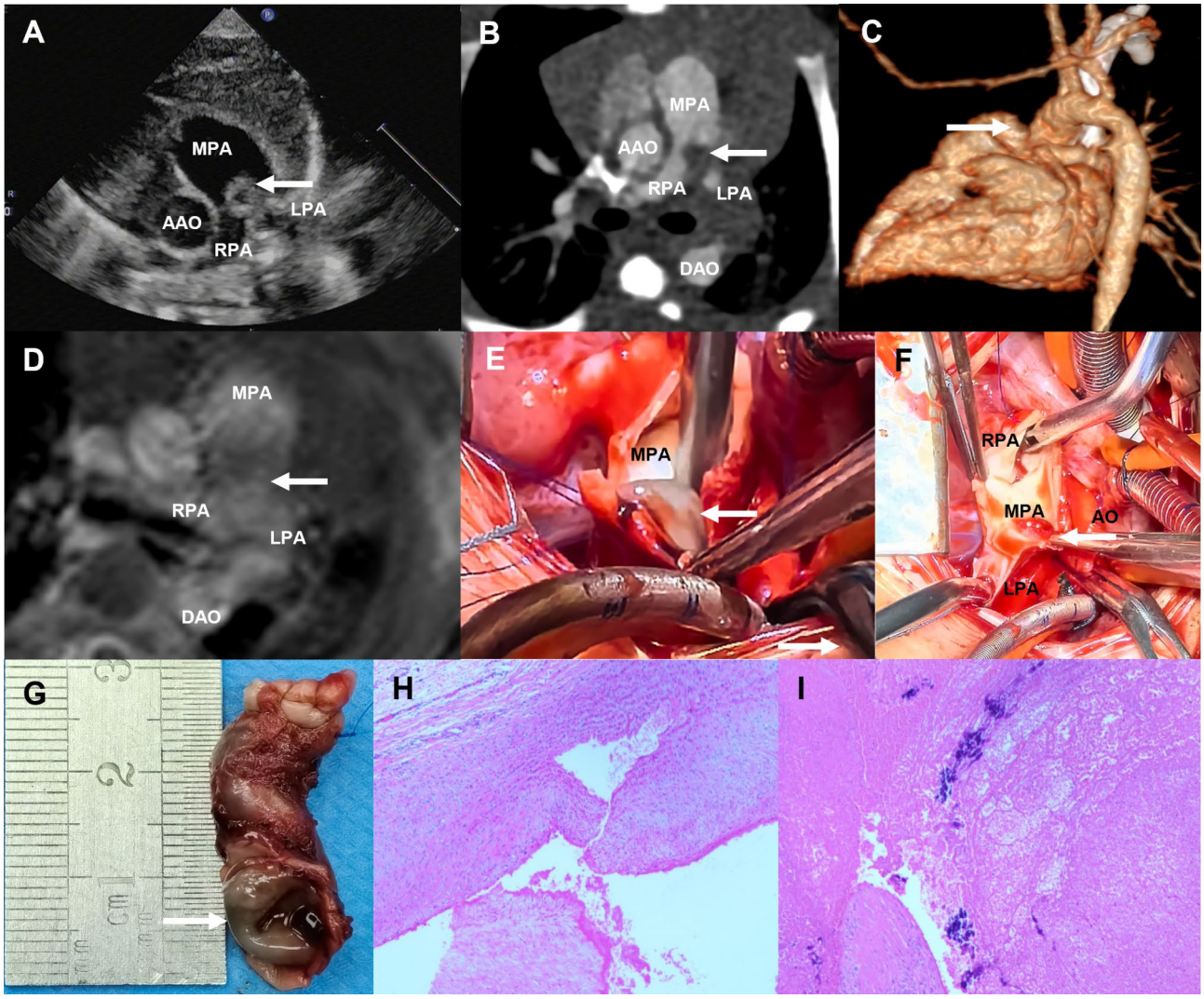

Fig. 1 Imaging findings reveal a large mass with indistinct boundaries situated at the bifurcation of the main pulmonary artery, causing bilateral stenosis: Echocardiography (Panel **A**), Cardiac CT (Panel **B**), a sagittal 3D reconstruction by CT (Panel **C**), and cardiac MR (Panel **D**); Intra-operative views (Panel **E**, **F** and **G**); Histopathology confirming ductus arteriosus aneurysm with thrombosis (**H**&**E** 5×, Panel **H** and **I**). All white arrows indicate the lesion
Abbreviations: CT, computed tomography; 3D, 3-dimensional; MR, magnetic resonance.

thymus tissue (Fig. 1 Panel B and C). For further evaluation, cardiac magnetic resonance (CMR) was performed, demonstrating a well-defined lesion (approximately 20 mm × 9 mm) with a filling defect situated between the MPA and the descending aorta (DAO), raising suspicion that the lesion originated from the ductus arteriosus (Fig. 1 Panel D).

The patient underwent emergency surgery for critically compromise of bilateral pulmonary arterial flow. Cardiopulmonary bypass (CPB) was established through aortic and bicaval cannulation. Following extensive and meticulous dissection, a large fusiform DAA was identified, with a waistline measuring 15 mm, an aortic end of 12 mm, and a pulmonary end of 10 mm in diameter. Deep hypothermic circulatory arrest (DHCA) was employed to facilitate radical resection of the aneurysm at 20 °C. The aortic end of the DAA and surrounding unhealthy ductal tissue were carefully excised. The distal aortic arch is transected and ligated. The DAO was mobilized, brought up to the undersurface of the proximal arch, and anastomosed in an end-to-side fashion. After the anastomosis was completed, CPB was resumed. The pulmonary end of the DAA was subsequently resected, and the defect at the bifurcation of the MPA was repaired using an autologous pericardial patch. (Fig. 1 Panel E, F and G, white arrows). The total durations for CPB, aortic cross-clamping, and DHCA were 147 min, 65 min, and 22 min, respectively. Postoperative histopathology confirmed the diagnosis of DAA (Fig. 1 Panel H and I).

The patient was extubated on postoperative day (POD) 2 and discharged on POD 20 without complications. At the 3-month follow-up, the patient exhibited no

symptoms of respiratory or heart failure, demonstrating normal growth, intelligence, and motor abilities.

Discussion

Ductus arteriosus aneurysm (DAA) is often asymptomatic and insidious, complicating the accurate determination of its incidence. DAA is identified in approximately 0.8% of cases in autopsy studies [3], and 1.5 ~ 2.2% in late pregnancy [1, 4]. Given the challenges in prenatal diagnosis, the true incidence may be underestimated. Some studies indicate that ductus arteriosus aneurysmal dilation could be a normal variant or part of the natural closure process, which may explain the spontaneous resolution observed in many asymptomatic newborns and children [5, 6]. However, about one-third of DAA cases can develop complications, with thrombosis and compression of surrounding structures being the most common issues. Serious complications, such as spontaneous rupture, erosion of nearby organs (like bronchi and esophagus), thromboembolism, and infective endocarditis, pose significant risks [1, 5, 7–12].

Although the precise pathogenesis of ductus arteriosus aneurysm (DAA) is not fully understood, several theories suggest connections to the ductus arteriosus closure mechanism, anatomical features, hemodynamics, and genetic factors. One proposed mechanism is that delayed closure at the aortic end of the ductus may lead to wall weakness and dysregulated changes [13]; however, this does not entirely explain the prenatal developmental processes. Observations of necrosis and myxomatous degeneration in the ductus media indicate wall weakness [1, 8, 14, 15], although similar phenomena can occur in a normal ductus. In patients with connective tissue disorders, abnormal elastin may predispose the ductus wall to congenital weakness [12, 16]. Another theory posits that ductal stenosis near the pulmonary artery end during the fetal period may contribute to dilation of the ductus arteriosus after birth [17, 18].

In this case, normal development of the ductal intima was noted, but localized rupture of the media's smooth muscle was observed, aligning more closely with the first theoretical explanation after ruling out a family history of connective tissue disease. Research indicates that infants born to mothers with diabetes or those born later in gestation may have a higher incidence of DAA [2]. However, in this case, the mother had no history of diabetes and delivered at term. Based on clinical features, imaging characteristics and histopathological findings of this particular case, we speculate that the DAA formed shortly after birth. Due to exposure to systemic circulation pressure, the intimal cushion had not fully developed, leading to disruption of the smooth muscle in the media and insufficient fibrin formation, which hindered the development of the intimal cushion [12]. Consequently, physiological closure of the ductus was prevented and the weak vessel wall ultimately ruptured under systemic pressure, resembling an aortic dissection. Additionally, failure or delayed closure of the ductus can produce physiological stenosis at its pulmonary end, leading to turbulence or damage to the endothelium, which may trigger thrombosis. The thrombus formed in the ductus could subsequently progress under systemic pressure to the main pulmonary artery, obstructing its branches.

Although approximately 70% of thrombotic DAA cases may resolve spontaneously without intervention or can be managed with anticoagulation (e.g., aspirin or low molecular weight heparin) [1, 3, 19–21], some cases require surgical intervention. Surgical indications include: [1] persistent patency of the ductus in the neonatal period [2], coexisting connective tissue disorders [22], thrombus progression to adjacent vessels or embolic events, and [14] significant compression of neighboring structures(1, 4). In this case, cardiac magnetic resonance imaging (CMR) revealed substantial growth of the DAA compared to earlier assessments, resulting in nearly total occlusion of the left pulmonary artery and necessitating emergency surgical intervention.

When the DAO is involved, complete excision of the DAA and the affected arterial wall is crucial to prevent thrombus recurrence. Deep hypothermic circulatory arrest is recommended for precise repair of the arterial wall defect. Direct closure or pericardial patching are often associated with high tension at the anastomotic site, which can lead to long-term stenosis. Adequate mobilization of the distal DAO and end-to-side anastomosis with the proximal aortic arch may be optimal alternatives. In confirmed DAA cases, direct ligation of the ductus is not recommended due to the risks of rupture, bleeding, and embolic events.

Conclusion

In summary, this report presents a rare case of DAA in a neonate, detailing its diagnosis and treatment while offering valuable insights for clinical practice and future research. Treatment strategies for DAA should be tailored to the individual circumstances of each patient, with close follow-up and early intervention recommended for those with connective tissue disorders. Additionally, prenatal examinations and neonatal echocardiography should be utilized to enhance early detection and management of this condition.

Abbreviations

DAA	Ductus Arteriosus Aneurysm
MPA	Main Pulmonary Artery
PA	Pulmonary Artery
CCT	Cardiac Computed Tomography
CMR	Cardiac Magnetic Resonance
CPB	Cardiopulmonary Bypass
DAO	Descending Aorta

POD Postoperative Day

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Not applicable.

Author contributions
XZ and HC designed the study and performed the surgery. WX and ZC also designed the study, wrote the first draft of the manuscript, and completed the revision. JC and JZ were in charge of data collection and video recording. JZ, HC, and JC provided professional suggestions and discussion on the surgery and supervised the research. All authors contributed to editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work to take public responsibility for appropriate portions of the content and agreed to be accountable for all aspects of the work in ensuring that questions related to its accuracy or integrity.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
The study was approved by Guangdong Provincial People's Hospital Ethics Committee (No. GDREC2019338H(R2)) on 17th September 2019. The written informed consent was obtained from the patient's parents.

Competing interests
The authors declare no competing interests.

Consent
Written consent was obtained from the patient.

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