Cardiology in the Young (2010), **20:** 89–90
doi:10.1017/S1047951109991168

Brief Report

Regression of Left Ventricular Dilation After Percutaneous Closure of a Large Intralobar Pulmonary Sequestration

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Abstract We describe an infant of 8 months who presented with left ventricular dilation due to an extensive intralobar sequestration of the right lung. The pulmonary sequestration was associated with a patent arterial duct and a right aortic arch. Percutaneous closure of the anomalous aberrant artery feeding the sequestrated lung resulted in prompt regression of the left ventricular enlargement.

Keywords: Heart failure; aberrant systemic artery; percutaneous intervention

Received: 30 March 2009; Accepted: 17 July 2009; First published online 30 September 2009

Pulmonary sequestration is a rare congenital disorder, usually presenting during childhood, and said to involve from 0.15 to 1.7% of population[1,2]. It consists of an aberrant systemic artery feeding isolated pulmonary parenchyma that lacks any communication with the airways. Patients with such pulmonary sequestration may remain asymptomatic for years, only to be diagnosed during routine chest radiography. Symptoms, when they exist, are often non-specific, and include chest pain, dyspneoa, and recurrent pulmonary infections. Open surgical excision is the usual treatment, albeit that other strategies, such as thorascopic intervention and transcatheter embolization have been reported[3]. Rarely, the lesion may become manifest in small infants and children with left ventricular volume overload, and symptoms of congestive cardiac failure.[4] In these rare cases, diagnosis is not easily achievable, specially when set against the more frequent causes of left venricular overload. Because of this, the optimal therapeutic strategy has yet to be determined.[5,6]

Case Report

A neonate aged 1 month, and weighing 3.8 kg, was referred to our department because of poor gain of weight after birth. A systolic murmur graded at 3 from 6 was audible over the precordium. The echocardiographic examination revealed the presence of a patent arterial duct, which was causing left-to-right shunting, with a mean gradient across the duct of 30 mmHg, along with significant left ventricular dilation, albeit with normal ventricular function. The end-diastolic diameter was measured at 27 mm, corresponding to a z score of 6, with end-systolic diameter of 13 mm. The end-diastolic volume was calculated at 122 ml/m^2, giving a z score of 15. The aortic arch was right-sided. The chest-X-ray was normal for in terms of the parenchymal appearances and the bronchial, but as anticipated, there was a big cardiac shadow. Anticongestive diuretic therapy was started. During close follow-up, the infant continued to show evidence of the left ventricular dilation, now accompanied by signs of cardiac failure, which initially we attributed to the left-to-right shunting across the arterial duct. We decided, therefore, to undertake cardiac catheterization so as to close percutaneously the arterial duct, the patient being aged 8 months and weighing 6 kg. Angiography confirmed the presence of a patent but restrictive arterial duct, with the ratio of pulmonary to systemic flow being unity. It revealed, however, the presence of a large anomalous artery, having a diameter of 6 mm, which arose from the descending thoracic aorta and fed a sequestrated postero-inferior lobe of the right lung (Figs 1a and 2a). The venous return from the

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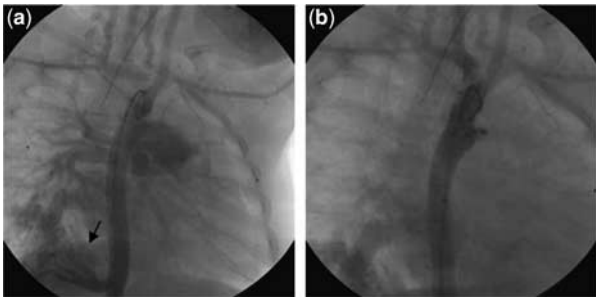

Figure 1.
*Aortography (**a**) performed to evaluate the anatomy of a patent arterial duct unmasked a large artery arising from descending aorta (arrow), which fed the right inferior lobe. The duct was closed percutaneously (**b**) using an Amplatzer device.*

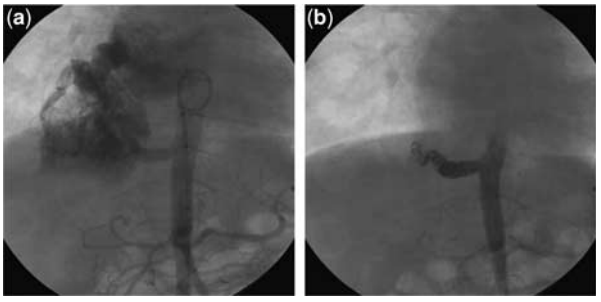

Figure 2.
*Angiography of the descending aorta (**a**) revealed the big intralobar pulmonary sequestration, with a large anomalous systemic artery feeding the postero-inferior segment of the right lung, which drained via the inferior pulmonary vein to the left atrium. The aberrant artery was closed successfully (**b**) by implantation of multiple coils.*

sequestrated part of the lung was normal, passing via the inferior right pulmonary vein into the left atrium. The pulmonary arteries were normally distributed to the remainder of the lungs, but the bronchial tree was not connected with the anomalous pulmonary mass, which therefore represented a classic intralobar pulmonary sequestration. We deduced that flow through the anomalous artery was responsible for the left ventricular volume overload and chamber dilation. We decided, therefore, to close both defects percutaneously. Using venous access, we closed the duct with an Amplatzer device of size 8/6 mm (Fig. 1b). The anomalous artery was successfully closed using multiple coils (Fig. 2b). Within 1 week of the procedure, the child was in good health, and echocardiographic examination confirmed the prompt regression of the left ventricular dilation, the z score for end-diastolic diameter having returned to 1, with the score for the end-diastolic volume, at 56 ml/m^2, being 3.5.

Discussion

Pulmonary sequestration is a rare congenital disorder, accounting for around one-twentieth of all congenital pulmonary malformations[1,2]. It exists when non-functioning lung tissue receives blood from anomalous systemic artery, lacking any communication with the tracheobronchial tree. The venous drainage usually is normal, but anomalies can sometimes occur, with drainage either directly to the right atrium, or to one of the major systemic veins of the thorax or abdomen, this latter feature being seen in extralobar pulmonary sequestrations and the scimitar syndrome. Children with pulmonary sequestration may be asymptomatic for years, albeit that signs of recurrent infection, such as fever and productive cough, are more common. The condition can be diagnosed accidently during a routine chest radiograph, when the sequestrated area can be seen as a homogeneous consolidation. Rarely, it can appear as a hyperlucent area, albeit not easily detected. Pulmonary sequestration can also, as in our patient, be a rare cause of significant left-to-right shunting, with the subsequent left ventricular volume overload producing signs of congestive cardiac failure[3]. Surgical or percutaneous treatment is recognised as restoring the good health of these small patients[4–6]. To the best of our knowledge, our patient is the first to show the association of an extensive intralobar pulmonary sequestration with a right aortic arch and patency of the arterial duct. The sequestration was detected in the catheterization laboratory during angiography for definition of the ductal anatomy, and was the true cause of the significant left ventricular dilation. Percutaneous closure of the aberrant artery produced prompt reversal of the left ventricular enlargement.

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