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CASE REPORT

Case Report: Ostium secundum atrial septal defect with unilateral lung hypervascularity revealing associated right pulmonary artery stenosis [version 1; peer review: 2 approved with reservations]

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Abstract

Background: Atrial septal defect (ASD) is often an isolated disease, but its association with other abnormalities can make diagnosis challenging. Careful analysis of simple complementary exams can help precise anatomical diagnosis ensuring suitable treatment. The aim of this article is to report, from a case report and literature review, diagnostic challenges and the contribution of simple complementary exams, such as chest X-ray, for the diagnostic orientation of an ASD associated with peripheral pulmonary artery stenosis, as well as therapeutic particularities.

Case report: We report the case of a girl born in 2007, with history of dyspnoea and recurrent bronchitis in whom a loud systolic murmur was detected fortuitously at the age of 2 years. Her clinical examination was otherwise normal. The electrocardiogram recorded sinus rhythm, incomplete right bundle branch block, and right ventricular hypertrophy. Chest X-ray showed moderate cardiomegaly and hypervascularity of the left lung field contrasting with reduced blood flow to the right lung. Doppler echocardiography revealed a wide ostium secundum ASD, right chamber volume overload and right pulmonary artery stenosis. The latter was confirmed by CT angiography and right cardiac catheterization. The patient underwent percutaneous right pulmonary artery dilation with stent placement. Control chest X-ray noted bilateral hypervascularity of the lung. The ASD was closed percutaneously one year later. The outcome was uneventful.

Conclusion: The combination of ASD with pulmonary artery stenosis limits pulmonary hyperflow. In our case, this stenosis was tight and sat on the right branch of the pulmonary artery reducing significantly blood flow to the ipsilateral lung. Careful chest X-ray analysis may suggest diagnosis, which can be confirmed by ultrasounds and if necessary, by further examination, allowing treatment adaptation. To

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Approval Status

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1. **P. Syamasundar Rao** , University of Texas-Houston McGovern Medical School, Children Memorial Hermann Hospital, Houston, USA
2. **Kartik Patel** , U. N. Mehta Institute of Cardiology and Research Center, Ahmedabad, India

Any reports and responses or comments on the article can be found at the end of the article.

our knowledge, this association is very rare and no similar case has been reported.

Keywords

Atrial septal defect, pulmonary stenosis, echocardiography, angioplasty, cardiac catheterization.

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Author roles: Slim M: Conceptualization, Software, Writing – Original Draft Preparation; Bodian M: Conceptualization, Writing – Original Draft Preparation; Neffati E: Supervision, Validation, Writing – Review & Editing; Boughzela E: Supervision, Writing – Review & Editing

Competing interests: No competing interests were disclosed.

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Introduction

Atrial septal defect (ASD) is a very common congenital heart disease^{1,2}. It can be associated with other cardiovascular abnormalities; the most common is pulmonary stenosis³. The latter usually concerns the valve or the right outflow tract but rarely pulmonary artery branches. This unusual association can be suspected by careful analysis of complementary exams. Currently, advances in interventional treatment make possible reliable and effective treatment of ASD even when associated with other lesions, in particular pulmonary stenosis³.

The aim of this article is to report, from a case report and literature review, diagnostic challenges and the contribution of simple complementary exams, such as chest X-ray, for the diagnostic orientation of an ASD associated with peripheral pulmonary artery stenosis, as well as therapeutic particularities.

Case report

We report the case of a girl born in 2007, with history of dyspnoea and recurrent bronchitis in whom a systolic murmur was detected in our outpatient office at the age of two years. The physical examination at this age noted a non-dysmorphic child with normal growth and psychomotor development. Auscultation of the pulmonary area noted loud 3/6 systolic ejection-type murmur, splitted second heart sound with a marked pulmonary component. An electrocardiogram recorded sinus rhythm, incomplete right bundle branch block and right ventricular hypertrophy.

Chest X-ray showed moderate cardiomegaly with cardio-thoracic ratio of 0.53, convex mid-left arch, and above all marked hypervascularity of the left lung contrasting with reduced blood flow to the right lung (Figure 1).

Doppler echocardiography noted a 20 mm diameter ostium secundum ASD with right chamber volume overload associated with right pulmonary artery (RPA) stenosis. Diagnostic confirmation of peripheral pulmonary branch stenosis was made by CT scan and right heart catheterization (Figure 2).

A two-step percutaneous treatment for these lesions was decided. RPA stenosis was treated firstly with 8 mm × 2 cm balloon in 2012 with poor initial result. A novel attempt in the same year with a balloon and stent placement (Express™ Vascular LD 10*37mm) was successful (Figure 3).

This result was optimized 6 months later through a 18 mm × 20 mm balloon leaving mild residual gradient of 10 mm Hg between pulmonary trunk and RPA. The ASD was closed successfully 1 year later in July 2013 with a 24 mm Figulla Flex II prosthesis. The procedure was uneventful and fluoroscopic control at the end noted ASD prosthesis in place and stent at RPA level (Figure 4). Control chest X-ray showed symmetrical bilateral vascularisation of the two lungs (Figure 5). Outcome was favourable. Control echocardiography performed at 4 years of regular follow-up noted mild residual pulmonary stenosis (maximal residual gradient of 15 mmHg), no stent restenosis and a well-sealed ASD prosthesis (Figure 6). Systolic right ventricle function indices were normal.

Figure 1. Chest X-ray of the patient. Cardiomegaly, convex middle arch and unilateral left lung hypervascularity contrasting with reduced blood flow to the right lung is shown.

Figure 2. Chest CT scan of the patient. (A) Tight stenosis at the origin of the right pulmonary artery (red arrow); (B) small right pulmonary artery (15 mm) contrasting with (C) dilated left artery (27 mm).

Figure 3. Right pulmonary artery angioplasty. (A) Selective angiography of the right pulmonary artery and **(B)** stent deployment.

Figure 4. Fluoroscopic image after right pulmonary stenosis angioplasty and percutaneous atrial septal defect closure. Atrial septal defect prosthesis (red arrow) and the right pulmonary artery stent (green arrow).

Figure 6. Last control echocardiography showing well-expanded right pulmonary artery stent.

Figure 5. Chest X-ray after right pulmonary angioplasty and percutaneous atrial septal defect closure. Symmetrical pulmonary vasculature and right pulmonary artery stent shown by red arrow.

Discussion

Our case emphasizes perfectly the importance of careful basic semiology analysis in the diagnosis process of congenital heart disease. In fact, heart murmur characteristics and asymmetric pulmonary vasculature in chest X-ray oriented the diagnosis of ASD associated with pulmonary branch stenosis⁴. Confirmation was made by appropriate investigations, particularly Doppler echocardiography, thoracic CT angiography and finally cardiac catheterization with selective angiograms. Chest roentgenogram still retains great value for the diagnostic process in cardiology. Thus in our patient, the finding of an unbalanced pulmonary vasculature especially when associated with an intense pulmonary murmur oriented the diagnosis of pulmonary artery branch stenosis. RPA stenosis caused pulmonary flow deviation mainly to the healthy side resulting in increased

vascularisation of the left lung contrasting with a hypovascularisation of the right one. This unilateral hyper-flow was quite marked because of its association to relevant left to right shunt related to wide-associated ASD. This suspicion of RPA stenosis was easily confirmed by echocardiography, CT scan, and right-heart catheterization with measure of pressure in right heart chambers, pulmonary branches and finally selective pulmonary artery branches angiographies.

Conventionally, treatment of this condition was surgical with ASD closure and pulmonary artery branch plasty. Currently, balloon dilatation with stent placement has revolutionized management of pulmonary stenosis especially those involving branches^{5,6}. Pulmonary artery stenosis can complicate the course of many congenital heart diseases. Percutaneous treatment can be performed as a surrogate or adjunct to surgery and it is considered as standard of care for proximal stenosis. For distal stenosis, it allows treatment of lesions inaccessible to the surgeon, often in addition to repair surgery of right ventricular outflow tract³. Angioplasty of the pulmonary arteries has evolved considerably since its introduction in the early 1980s. High pressure balloons usually are 2 to 4 times larger than the diameter of the stenosis are used. Stents used are still currently most often not pre-mounted and have the advantage of being expandable to a diameter sufficiently close to vessel size in adulthood⁷. This type of stent was successfully used in our patient and allowed restoration of pulmonary vasculature by removing the peripheral pulmonary stenosis. The success of pulmonary dilatation authorized percutaneous closure of the ASD, which was performed successfully one year later by prosthesis. Percutaneous closure is currently the standard treatment for ostium secundum ASD with adequate rims and diameter less than 38 mm with a success rate close to 100% and lower morbidity compared to surgery.

Our case is very rare and to our knowledge, no similar cases have been reported. It proves feasibility and reliability of

percutaneous treatment for such a case. The sequence of the lesion treatment is dictated by lesions complexity whose failure can shift the case to surgery. This is the reason why we waited obtaining a satisfactory and stable result on pulmonary artery stenosis before treating ASD.

Conclusion

Pulmonary stenosis can be associated with ASD limiting pulmonary hyper-flow. In our case, this stenosis was tight and sat on the origin of the right branch, which resulted in reducing significantly blood flow to the ipsilateral lung. Therefore, ASD-related pulmonary hyperflow was directed to the left lung field, explaining the radiological aspect particularly unilateral hypervascularity. Careful chest X-ray analysis can allow suspicion of pulmonary artery branch stenosis. Confirmation can be made by Doppler echocardiography and, if necessary, by further examination allowing treatment adaptation. Management of this association benefited from interventional techniques progress allowing successfully treatment with stable long-term outcome. Indeed, with a follow-up of four years, the atrial septum was tight and there was no residual pulmonary stenosis with a normalized RV function.

Consent

Written informed consent for publication of the clinical details and images was obtained from the patient's father.

Data availability

All data underlying the results are available as part of the article and no additional source data are required.

Grant information

The author(s) declared that no grants were involved in supporting this work.

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Kartik Patel

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I congratulate authors for the successful management of this case. This case report is well written still I have some comments for the same.

1. It requires editing by someone who is proficient in English.
2. Can authors describe what would be the probable reason for narrowing of right pulmonary artery ostia? what was the position of the arch of aorta, right or left?
3. Looking at follow up image the stent seems to be protruding into the main pulmonary artery. Does there are any signs of hemolysis?
4. Can authors explain about the approach to the patients with ostial right pulmonary artery stenosis and advantage vs disadvantages of surgery vs stent?
5. I also have doubt of considering the lesion as peripheral branch stenosis of pulmonary artery. I think better word would be right pulmonary artery stenosis or RPA ostial stenosis.
6. Why authors have decided to do two stage approach? why not single stage approach?

Is the background of the case's history and progression described in sufficient detail?

Yes

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Yes

Is the case presented with sufficient detail to be useful for other practitioners?

Yes

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: Pediatric and Adult Cardiac Surgery

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Reviewer Report 10 December 2018

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P. Syamasundar Rao

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The paper reports a rare co-occurrence of atrial septal defect and branch pulmonary artery stenosis and is worthy of eventual indexing.

The expression and syntax should be improved by editing, by someone who is proficient in English.

The assertion that "no similar case has been reported" should be removed. In the past, the association of atrial septal defect and branch pulmonary artery stenosis was reported (Lloyd *et al.*, 1994¹ and Rao *et al.*, 1995²), although the subject of these papers did not allow for detailed description.

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Is the background of the case's history and progression described in sufficient detail?

Yes

Are enough details provided of any physical examination and diagnostic tests, treatment

given and outcomes?

Yes

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Yes

Is the case presented with sufficient detail to be useful for other practitioners?

Yes

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: Congenital heart disease and Interventional Pediatric cardiology

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

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