Respiratory Medicine Case Reports 22 (2017) 238–242

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: www.elsevier.com/locate/rmcr

Case report

Unilateral absent pulmonary artery in an adult - A diagnostic and therapeutic challenge

Faheem Seedat [a, *], Ismail S. Kalla [b], Charles Feldman [b]

[a] Department of Internal Medicine, Faculty of Health Sciences, University of Witwatersrand, Johannesburg, South Africa
[b] Division of Pulmonology, Department of Internal Medicine, Charlotte Maxeke Johannesburg Academic Hospital and Faculty of Health Sciences, University of Witwatersrand, Johannesburg, South Africa

ARTICLE INFO

Article history:
Received 21 June 2017
Received in revised form
8 September 2017
Accepted 9 September 2017

Keywords:
Pulmonary hypertension
Unilateral absent pulmonary artery
Diagnosis
Therapy

ABSTRACT

Unilateral absent pulmonary artery (UAPA) is a congenital abnormality rarely diagnosed in adults. UAPA has a myriad of clinical presentations and pulmonary hypertension is present in a quarter of all cases. Isolated UAPA commonly affects the right pulmonary artery and occurs as a result of abnormal development of the sixth aortic arch segment. Due to its rarity, it remains a diagnostic and therapeutic challenge. We describe a case of UAPA in an adult presenting with severe pulmonary hypertension. We describe the appropriate diagnostic approach to a patient with pulmonary hypertension and illustrate the importance of a detailed evaluation to determine the underlying aetiology, particularly in rare causes. Furthermore, we review the clinical presentation, diagnosis and management challenges of UAPA in adults.

1. Introduction

Unilateral absent pulmonary artery (UAPA) is a rare congenital abnormality due to pulmonary artery agenesis, most commonly of the right pulmonary artery, and may present as a cause of pulmonary hypertension in adults [1]. No consensus exists regarding therapy and it is a diagnostic challenge [2]. We highlight this condition presenting with pulmonary hypertension, review therapeutic principles for UAPA in adulthood and demonstrate that to ensure an accurate diagnosis of rare causes of pulmonary hypertension a high index of clinical suspicion and close scrutiny of investigations is necessary.

2. Case report

A 53-year-old woman, with well-controlled hypertension and hypothyroidism, presented to the emergency department with sudden onset of dyspnoea associated with palpitations. On examination, her blood pressure was 60/40 mmHg with a pulse rate of 160 beats per minute and she was noticeably cyanosed. Her respiratory examination was normal but on cardiovascular examination she was tachycardic and on auscultation a loud pulmonary component of the second heart sound and a murmur of tricuspid regurgitation were present.

An electrocardiogram (ECG) showed a supraventricular tachycardia (SVT), which was terminated following synchronized electrical cardioversion, using intravenous midazolam titrated to effect for sedation under the direction of an emergency medicine physician (Fig. 1). The subsequent ECG demonstrated features of right ventricular hypertrophy with pressure overload: a right axis; an associated S1T3 pattern; a qR pattern in lead V1; T wave inversion in leads V2–V4 and p - pulmonale (Fig. 2). Subsequently, the patient was started on therapeutic low molecular weight heparin along with intravenous amiodarone, to maintain sinus rhythm and prevent the possible recurrence of the hemodynamically unstable supraventricular tachycardia, and admitted to the Intensive Care Unit for monitoring.

Transthoracic echocardiography showed a dilated right ventricle with severe tricuspid regurgitation and a pulmonary artery pressure of 120 mmHg. The left atrium and ventricle were normal with an ejection fraction of 73%. The pulmonary function

Abbreviations: UAPA, Unilateral absent pulmonary artery; CTEPH, Chronic thromboembolic pulmonary hypertension; SVT, Supraventricular tachycardia; ECG, Electrocardiogram; CTPA, Computed tomography pulmonary angiogram; MRI, Magnetic resonance imaging.

* Corresponding author. P.O. Box 96630, Brixton, 2019, South Africa.
E-mail address: faheem@global.co.za (F. Seedat).

http://dx.doi.org/10.1016/j.rmcr.2017.09.004

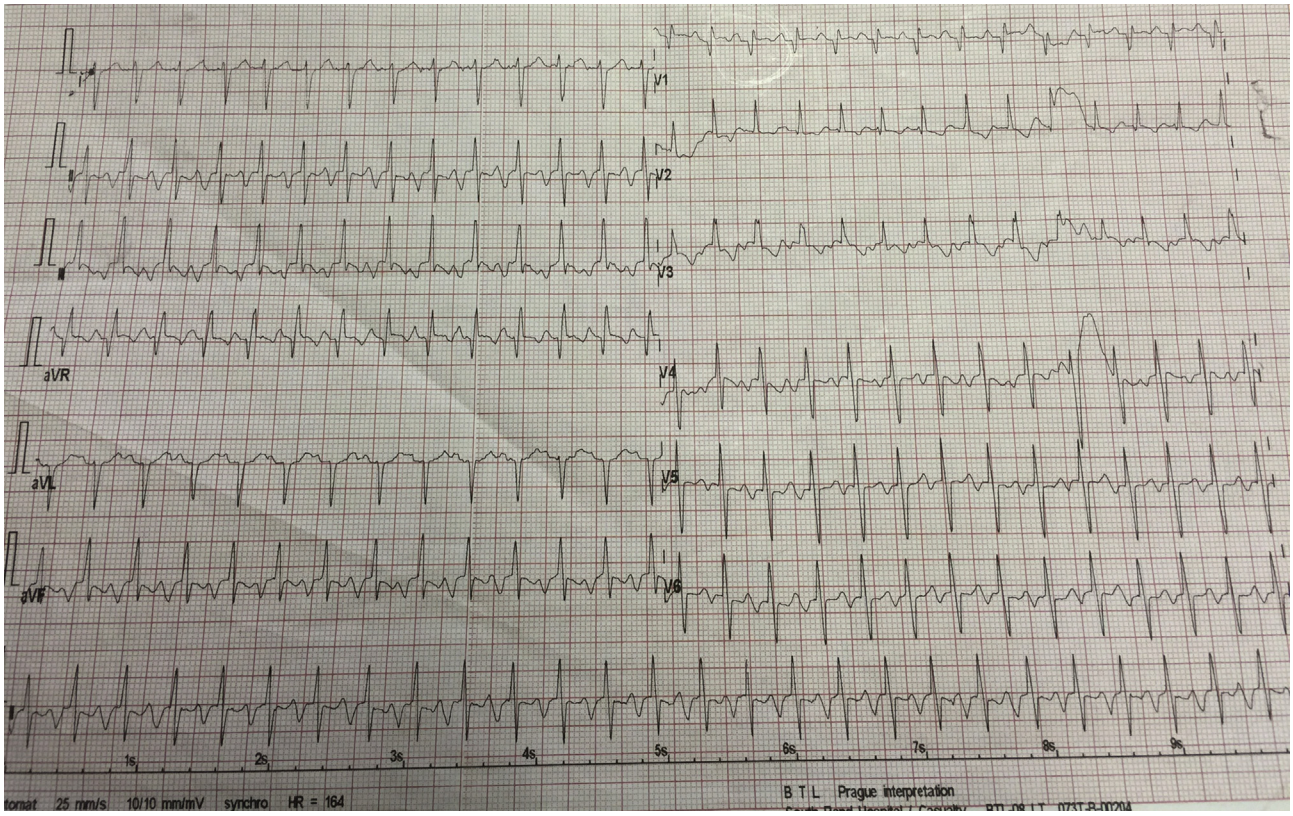

Fig. 1. Supraventricular tachycardia with features of right ventricular strain.

test showed a low diffusion capacity of 45% with a normal FEV1, FVC and FEV1/FVC ratio. Due to her initial SVT and marked pulmonary hypertension a diagnosis of chronic thromboembolic pulmonary hypertension (CTEPH) was initially suspected. On chest radiograph, a small oligaemic right lung field and empty right pulmonary bay (hilum) was noted with hyperinflation and plethora of the left lung field and a prominent left pulmonary artery (Fig. 3). A radionuclide ventilation/perfusion scan showed a barely visible right lung field on perfusion images but was well visualized on ventilation images. The computed tomography pulmonary angiogram (CTPA), following close review, demonstrated an absent right pulmonary artery with abrupt cutoff and atresia after its origin and no collateral circulation to the right lung was noted (Figs. 4 and 5). A massive right pulmonary artery clot was excluded and the atretic right pulmonary artery was confirmed on cardiac magnetic resonance imaging (MRI), which also showed an increased right ventricular mass and end-systolic volume with reduced systolic function (Fig. 6). A right heart catheterization confirmed high pulmonary pressures measuring 130/38 mmHg; a mean pulmonary artery pressure of 76 mmHg and 23.4 Woods units. A nitric oxide vasoreactivity test was negative with pulmonary pressures measuring 121/41 mmHg, a mean pulmonary artery pressure of 76 mmHg and 18.02 Woods units post the administration of inhaled nitric oxide. This was performed with a senior anaesthetist and cardiologist present, as great care should be taken in the sedation of patients with severe pulmonary hypertension.

The patient remained stable throughout the admission but due to her age surgical re-vascularization was deemed not possible. She was started on supplemental home oxygen therapy, sildenafil for her pulmonary hypertension, oral furosemide due to clinically evident right heart failure and an antiarrhythmic and has remained stable on therapy.

3. Discussion

Unilateral absent pulmonary artery (UAPA), first described by Frentzel in 1868 [3], is a rare congenital malformation and occurs in 1 in 200,000 adults with a median age of presentation of 14 years [1,4,5]. Agenesis of the pulmonary artery occurs due to altered development of the sixth aortic arch segment that does not connect to the pulmonary trunk and in two thirds of cases affects the right pulmonary artery, the side opposite to the aortic arch [4–6]. Often it occurs in association with other cardiac defects, such as tetralogy of Fallot or septal defects, but it can occur in isolation, as in our patient [1,2,4]. The distal vessels of the affected lung remain intact and are vascularized by bronchial vessels and abnormal collaterals from bronchial, subclavian, intercostal and sub-diaphragmatic arteries [4,7,8].

In UAPA the pulmonary hypertension occurs on the basis of pulmonary arterial vascular remodeling where increase pulmonary blood flow to one lung, through the single patent pulmonary artery, results in increase shear stress with subsequent endothelial intimal injury. This increased pulmonary blood flow stimulates the release of endothelin, which acts as a potent vasoconstrictor and stimulates vascular smooth muscle proliferation with resultant intimal hyperplasia, medial hypertrophy and, ultimately, collagenous replacement of the intima in late disease. A similar phenomenon occurs to some patients who have undergone a pneumonectomy, particularly of the right lung, who develop pulmonary hypertension [9–12].

The clinical presentation of patients with UAPA is varied and some patients may be asymptomatic. A review of 108 cases in 2002 showed that 40% of patients presented with poor effort tolerance and dyspnea; 37% presented with a combination of recurrent chest infections, pleural effusion and chest pain and 12% with high