International Journal of Surgery Open 13 (2018) 20–23

Contents lists available at ScienceDirect

International Journal of Surgery Open

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

Case Report

The silent killer presenting as pulmonary valve stenosis

Nicolas Nicolaou [a, *], Nigel Flint [b], Mark Andrew Nicolaou [c], Chris Conradie [d]

[a] Division of Cardiothoracic Surgery, Flora Clinic Johannesburg, South Africa
[b] Division of Radiology, Flora Clinic Johannesburg, South Africa
[c] Division of Radiology, University of the Witwatersrand, Johannesburg, South Africa
[d] Division of Cardiology, Flora Clinic Johannesburg, South Africa

ARTICLE INFO

Article history:
Received 12 April 2018
Received in revised form
25 July 2018
Accepted 29 July 2018
Available online 1 August 2018

Keywords:
Pulmonary embolus
Pulmonary hypertension
Pulmonary valve

ABSTRACT

Chronic Thrombo-Embolic Pulmonary Disease (CTEPD) is usually a silent disease representing a small group of patients who at some stage had suffered a thrombo-embolic event in which the individual may or may not have been aware of. It is estimated that this syndrome develops in approximately 3.8% of patients having suffered an acute Pulmonary Embolus (PE) thus representing a pool of patients estimated in the literature to be in the vicinity of 19,000 [1] It has further been documented that this condition is seriously underdiagnosed and is thought to represent in excess of 100,000 individuals in the United States with CTEPD which potentially could be “cured”.

1. Introduction

Chronic Thrombo Embolic Pulmonary Disease (CTEPD) is usually a silent disease representing a small group of patients who at some stage had suffered a thrombo-embolic event which the individual may or may not have been aware of. It is estimated that this syndrome develops in approximately 3.8% of clinically recognized acute PE thus representing a significant number of patients. As there are more than 500,000 survivors of acute pulmonary embolus in the United States alone this could represent a cohort group of 19,000 patients [5,8] It has also been documented that there are a group of patients who may never have been aware of any acute episode thus the total number of patients developing CTEPD are probably underestimated resulting in an a seriously underdiagnosed disease. This group of patients will ultimately develop pulmonary hypertension with all its consequences and are potentially “curable” if diagnosed and treated appropriately [1,2] The main symptom complex is non-specific but the most common presenting feature is progressive effort intolerance.

We present an interesting case of CTEPD who presented with signs of pulmonary stenosis and symptoms of progressive dyspnea.

2. Presentation of case

A 38 year old female was referred for evaluation of a cardiac murmur from a peripheral center. Her initial presenting feature was shortness of breath. She was initially diagnosed with asthma and given combination b2-steroid inhaler which did not alleviate her symptoms. As she remained symptomatic she was further evaluated and was informed that there was a prominent heart murmur which was attributed to aortic valvular stenosis. Beside Perindopril for mild hypertension and a hysterectomy 6 years previously she had suffered no other past illness that she could remember. She had never been on any hormone replacement therapy but did continue to smoke. There was no family history of any illness and her parents were well and still alive.

Clinical examination revealed a young female appearing mildly short of breath at rest with central desaturation. She was unable to walk 50 m and became significantly dyspnoeic. Further examination revealed sinus rhythm with all pulses been equal and present. Her Blood Pressure at rest was 115/85. There was mild peripheral oedema and a raised jugular venous pressure.

On auscultation a harsh crescendo-decrescendo ejection murmur was clearly audible at the left parasternal border in the second interspace that did not radiate to the base of the neck suggestive of pulmonary valve stenosis.

X-Ray Chest confirmed volume loss of the left lung and a shift of the mediastinum to the left. The left lung also demonstrates oligaemia (Fig. 1). Blood samples were performed and except for a

* Corresponding author. Division of Cardiothoracic Surgery, Flora Clinic, 2nd floor South East Block, William Nicol Drive Floracliffe, 1709, South Africa.
E-mail address: cardiocom@iafrica.com (N. Nicolaou).

https://doi.org/10.1016/j.ijso.2018.07.006

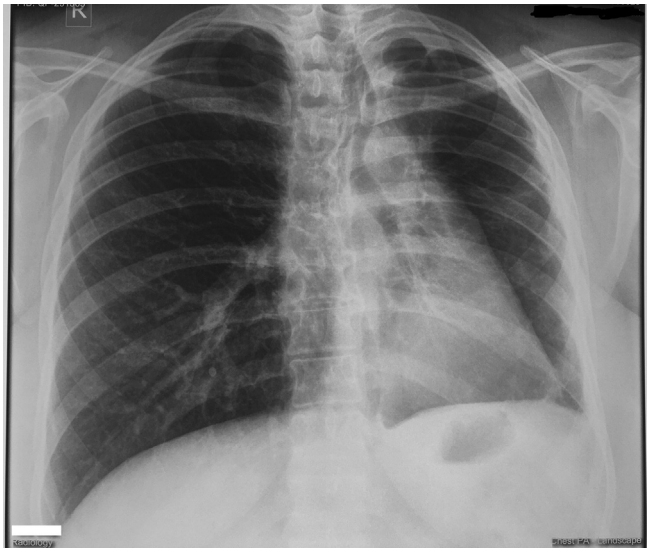

Fig. 1. X-RayChest-Volume loss of the left lung with mediastinal shift.

mildly elevated D-dimer no other anomaly was noted. Ultrasound of the limbs and pelvis did not reveal any thrombi.

A Computerized Tomogram of the chest and abdomen confirmed an extensive thrombus arising in the pulmonary trunk with an >80% occlusion of the main pulmonary artery accounting for the high gradient across the pulmonary valve. There was complete occlusion of the left pulmonary artery with no flow. Left upper lobe pulmonary fibrosis with pleural thickening explained the changes on X-Ray chest. (Fig. 2). No abnormality was noted in the pelvis.

Left and Right heart catheterization was performed. A gradient of 69 mm across the pulmonary valve was measured with a mean pulmonary artery pressure of 13 mm Hg. No flow could be demonstrated into the left pulmonary artery. The coronary arteries and left ventricular function were normal.

Transthoracic Echocardiogram post angiogram revealed a dilated dysfunctional right ventricle with mild tricuspid regurgitation. Due to the poor echocardiographic window the pulmonary valve could not be clearly visualized.

A diagnosis of advanced CTEPD was made and the patient was prepared for surgery.

A median sternotomy was performed entering the pericardium in the midline. Marked distension with a dysfunctional right ventricle was noted. The aorta and both vena cavae were cannulated in the routine fashion and cardio-pulmonary bypass instituted. Cooling was initiated and ice packs used to cool the head. Once the core temperature had reached 25° C a cross clamp was applied and ante-grade cardioplegia given to arrest the heart. Cooling continued to a target temperature of 18 °C. Both vena cavae were then snared. On palpation of the main pulmonary artery and branches a hard “mass” was felt. This extended into both pulmonary arteries. At this stage the right main pulmonary artery was dissected free under the SVC with the SVC snare helping as traction.

The main pulmonary artery was then opened and an organized hard fibrotic thrombus was found impinging upon and occluding the pulmonary valve explaining the pulmonary stenotic murmur. (Fig. 3) The thrombus was gently dissected free from the vessel wall up to the pulmonary arterial division. Incision was then extended into the right pulmonary artery and at this stage circulatory arrest was initiated. Complete removal of all clots was achieved by an

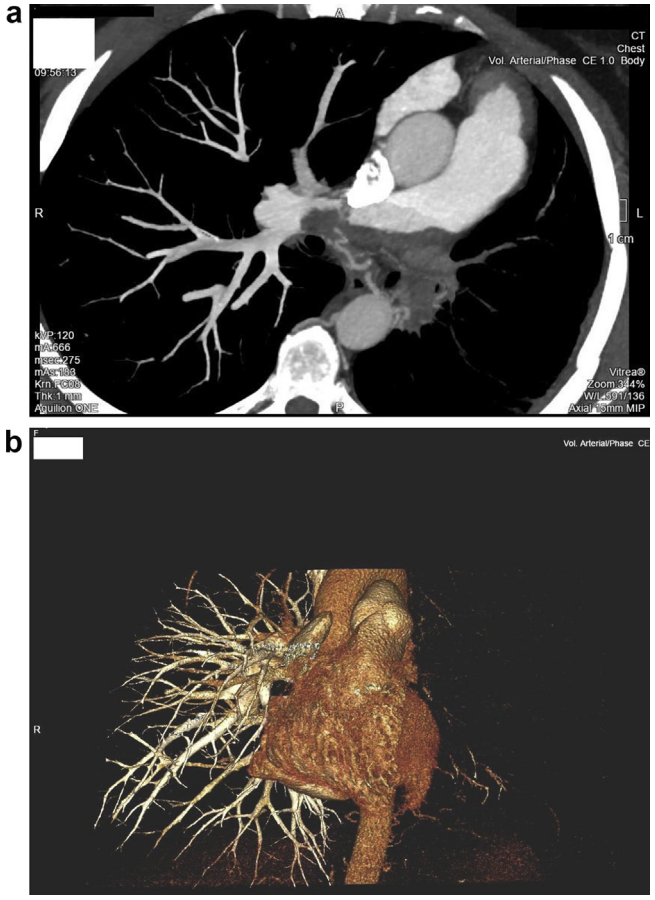

Fig. 2. a and b: Post Contrast CT Scan of Chest showing extensive thrombus with complete occlusion of the left pulmonary artery and pleural thickening in the left thorax.

endarterectomy technique. Dissection was then continued into the left pulmonary artery which was completely occluded. Both lungs were washed out with saline and gently inflated. The arteriotomy was then closed with 6/0 Prolene. The right atrium was then opened but no abnormality or PFO was noted.

After de-airing the Right side by removing the snares and the cross-clamp the patient was warmed and weaned off cardio-pulmonary bypass with low dose inotropic support. She was electively ventilated for 24 hours post operatively. After requiring an intercostal drain a week later for a residual serous effusion (Fig. 4) she was discharged home on warfarin with a markedly improved effort tolerance and a normal blood gas.

3. Discussion

The majority of patients who sustain an acute pulmonary embolism have a near total resolution with minimal sequelae and recover back to a normal pulmonary vascular flow. Acute emboli are normally lysed within a 2 week period and it is believed that if this does not occur they will attach to the arterial wall and slowly convert into an elastic fibrous material. Further emboli or thrombi will form on these areas and thus propagate to large thrombi occluding the blood flow and resulting in pulmonary hypertension. It is only in a minority of cases 3.8% [3] that will develop chronic thromboembolic pulmonary hypertension [4,5].