Total anomalous pulmonary venous drainage in an adult - a rare presentation

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Summary

We report a 25 year old man who presented with minimal cardiac symptoms and signs of an atrial septal defect on examination, who was also found to have a supracardiac type of Total Anomalous Pulmonary Venous Drainage (TAPVD).

Introduction

Total Anomalous Pulmonary Venous Drainage is an uncommon congenital heart disease[1]. Normally the four pulmonary veins open separately into the left atrium. In TAPVD, they open into the right side of the heart. There are three types, namely the supracardiac, cardiac and infracardiac depending on at which level the connection occurs[2]. The supracardiac variety of TAPVD consists of the four pulmonary veins joining to form a vertical vein which opens to the left brachiocephalic vein or superior vena cava. There is mixing of the circulations and thus a right to left shunt is necessary for survival[3]. They could be further subdivided as obstructive or non-obstructive, depending on the presence of obstruction to pulmonary blood flow. The infracardiac type is usally of the obstructive variety[4]. The majority of patients present during their first year with cyanosis and, or congestive cardiac failure[5]. If this defect is uncorrected 80 percent die within the first year [6,7].

Case Report

A 25 year old man who was in the army for the last four years, sought medical treatment as he had mild shortness of breath after running 1 mile during his physical training routine, over the past two months. He had no shortness of breath on walking or at rest and he did not have paroxysmal nocturnal dyspnoea or orthopnoea. He did not have any other symptoms referrable to the cardiovascular or respiratory systems. There was nothing of note in his past medical history and family history. He was an occasional smoker and took alcohol once a week.

On examination he was not pale, cyanosed (central or peripheral) or clubbed. His height was 5 feet 8 inches, weight 135 pounds and there were no congenital abnormalities or dysmorphic features. His pulse was 70 per minute and was of normal volume and character, blood pressure: 125/80 and his jugular venous pressure was not elevated. There was mild cardiomegaly, an ejection systolic murmur, and a fixed widely split second heart sound with a moderately loud pulmonary component in the second left intercostal space. Examination of his respiratory, abdominal and nervous systems were normal.

His electrocardiogram showed a right bundle branch block and right axis deviation. The chest X-ray showed the classical "figure of 8" or "snowman" appearance (Figure 1) with a plethoric lung field. 2 D Echocardiography showed an atrial septal defect and a supracardiac total anomalous pulmonary venous drainage, which was confirmed with a CT scan of his chest (Figure 2) and right heart catheterisation (Figure 3). The saturation of blood in the periphery was 89 percent. A first pass radionucleotide scan following injection of radionucleotide through the right cephalic vein, showed that the radionucleotide appeared in the left atrium prior to it reaching the lungs showing the presence of a left to right shunt, and that the lung recirculation time was prolonged, and there was delay in the radionuceotide reaching the aorta. At present he is awaiting surgery for correction of the anomalous venous drainage and closure of the atrial septal defect.

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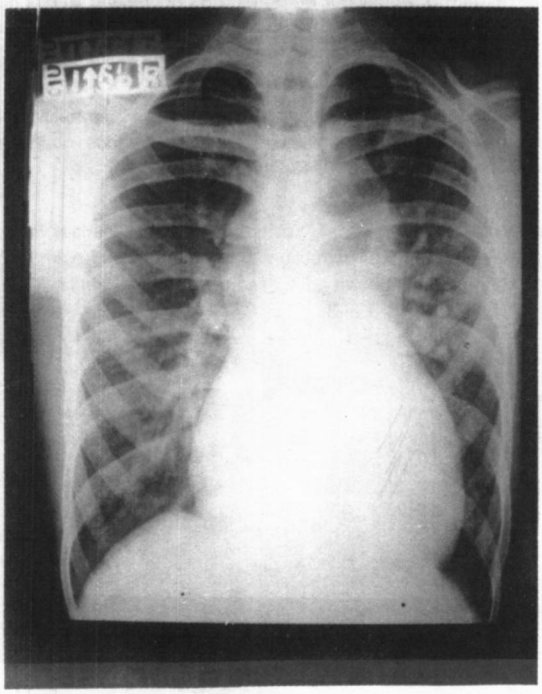

Figure 1. Chest X-ray showing the 'figure of 8', or 'snowman' appearance.

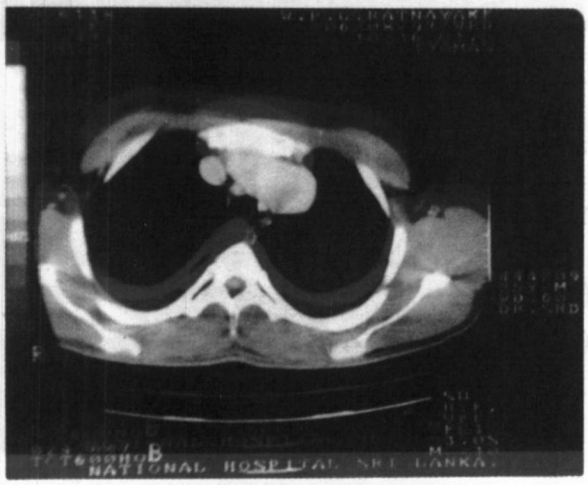

Figure 2. CT scan of thorax showing the anomalous venous connection, dilated superior venacava and vertical vein.

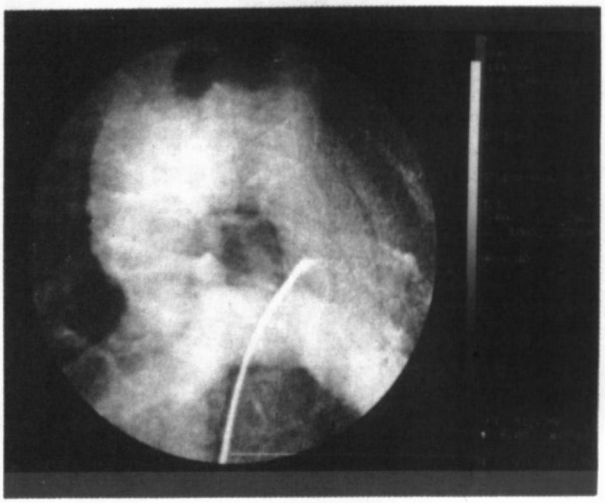

Figure 3. Right heart catheterisation showing the anomalous venous connection.

Discussion

Uncorrected TAPVD persisting to adult life in an active adult producing minimal symptoms is very rare. This has not been reported previously in Sri Lanka and there are only a few reports of it worldwide[8]. Our patient who has a supracardiac TAPVD and an atrial septal defect developed mild shortness of breath only on running 1 mile, and may have remained asymptomatic if not for exertion related to his profession. Further he had no evidence of cyanosis clinically and the peripheral arterial oxygen saturation at catheteraisation was not greatly reduced.

In a patient with a shunt defect the presence of a widened mediastinum with a possible figure of eight appearance, should prompt one to look carefully and demonstrate the opening of all four pulmonary veins. The detection of the total anomalous venous connection in our patient prior to surgery was vital as it changes the operative procedure normally employed to close an uncomplicated shunt defect.

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