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Unilateral inverse moustache sign

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DESCRIPTION

A 54-year-old lady was admitted with breathlessness since 2 months (NYHA class 3). She was previously diagnosed to have rheumatic heart disease (RHD), but was not on any medications. Her chest radiograph revealed

evidence of main pulmonary artery dilatation and unilateral upper lobe venous prominence (cephalisation) on the right side (figure 1A). Echocardiogram showed partial anomalous pulmonary venous connection (PAPVC) of the left upper pulmonary vein to the left innominate vein

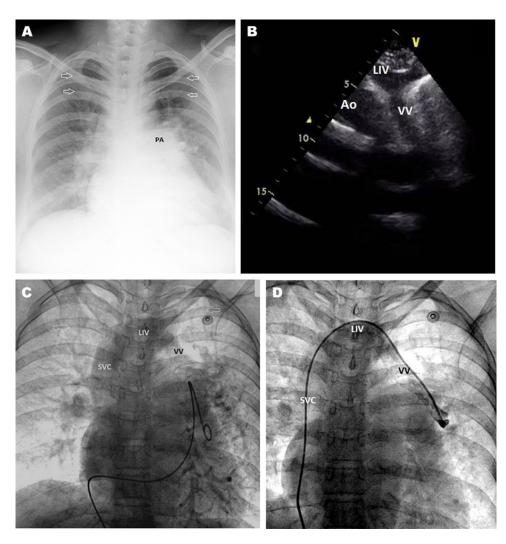


Figure 1 (A) Chest radiograph (PA-view) showing main pulmonary artery dilatation (PA) and unilateral upper lobe venous prominence (cephalisation) on the right side (arrows). (B) Echocardiogram (high left parasternal window in the transverse view) showing partial anomalous pulmonary venous connection (PAPVC) of the left upper pulmonary vein (LUPV) to the left innominate vein (LIV) via a vertical vein (VV). (C) Levophase for pulmonary venous return showing PAPVC of left upper pulmonary veins connecting to the left innominate vein with subsequent drainage to the right superior vena cava (SVC) and to the right atrium. (D) Direct injection into partial anomalous PAPVC of the LUPV to the LIV and to the right SVC via a (VV).

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via a vertical vein (figure 1B). In addition, chronic RHD with severe mitral stenosis (MS) with mitral valve area of 0.8cm² was noted. Interatrial septum was intact. The patient underwent cardiac catheterisation with selective pulmonary arteriography and the levophase for pulmonary venous return showed PAPVC of left upper pulmonary veins connecting to the left innominate vein with subsequent drainage to the right superior vena cava and to the right atrium (figure 1C). The anomalous connection was entered; direct injection of radiographic contrast dye delineated the anatomy (figure 1D). The patient subsequently underwent a successful percutaneous valvotomy for the MS. The association of PAPVC with intact atrial septum and severe rheumatic MS is rare. In this lady, the PAPVC was of such slight haemodynamic significance that the lesion was not recognised earlier, but incidentally picked up on a radiograph because of the abnormal vascular redistribution. The left atrial hypertension due to MS would result in an elevated pulmonary capillary wedge pressure in both lungs but substantially higher in the right normally draining lung; whereas the anomalous drainage into the right atrium would serve as a safety check for the left lung.2 This could explain the differential plethora detected on the radiograph which can be described as a variant of the classically described inverse moustache sign of pulmonary venous hypertension, although unilaterally.

Learning points

- Partial anomalous pulmonary venous drainage with intact interatrial septum associated with mitral stenosis is rare.
- ▶ Differential pulmonary vascular distribution (plethora) on chest radiograph is a subtle clue for the presence of anomalous venous drainage.

Competing interests None.

Patient consent Obtained.

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