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ENDGAMES

CASE REVIEW

Patent ductus arteriosus illuminating an old eponym

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A 60 year old woman was sent from primary care for assessment of acute left sided pleuritic chest pain and dyspnoea without sputum production. She had been experiencing malaise, intermittent fever, and rigors for four weeks. A patent ductus arteriosus (PDA) had been identified in late adulthood 10 years earlier, after a murmur was detected on routine medical examination. This was confirmed by transthoracic echocardiography; there were no other signs of haemodynamic importance.

At presentation she had a fever (39.0°C) with left sided pleural rub. Precordial examination identified a continuous "machinery" murmur. Electrocardiographic findings were within normal limits and laboratory investigations confirmed normocytic anaemia, leucocytosis, and preserved renal function. Group B streptococcus was isolated from all three separate blood cultures. Her pleural rub corresponded to an abnormality on chest radiography (fig 1).

Questions

1. What abnormality on chest radiography corresponds to the pleural rub?
2. Given the clinical presentation and history of PDA, what is the likely cause of this abnormality?
3. What investigation would confirm the cause of this abnormality?
4. What is the initial cardiac imaging technique of choice?

Answers

1.

What abnormality on chest radiography corresponds to the pleural rub?

Short answer

The wedge shaped peripherally based opacity in the left mid zone.

Discussion

The triangular (wedge) shaped opacity in the left mid zone (fig 2) corresponds to the clinical history of left sided pleuritic pain and examination finding of a pleural rub. Such a wedge shaped opacity is eponymously termed "Hampton's hump"—an old but important plain radiological sign of pulmonary embolism. It was first described in 1940 by the radiologist Aubrey Otis Hampton.[1] On close inspection of the enlarged radiograph, smaller scattered opacities that project over the right lung field can be seen, as well as unconnected adhesive electrocardiographic monitoring connectors.

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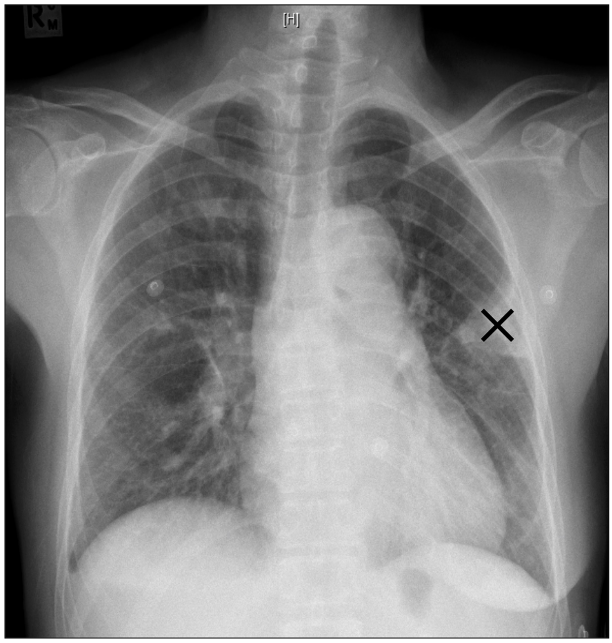

Fig 2 Chest radiograph showing a peripheral wedge shaped opacity in the left mid zone (Hampton's hump; X)

2.

Given the clinical presentation and history of PDA, what is the likely cause of this abnormality?

Short answer

Pulmonary embolism and infarction owing to septic pulmonary emboli from the infected PDA.

Discussion

Infective endocarditis is an important but rare complication to be considered in any adult with fever and congenital heart disease. The pleuritic chest pain and chest radiographic changes could be consistent with pneumonia or pulmonary embolism (with pulmonary infarction). Ockham's razor favours a single unifying diagnosis, and given our patient's predisposing condition of PDA, infective endocarditis with septic pulmonary emboli in the lung is the most likely cause of the presentation and explains the abnormal radiograph. The diagnosis of infective endocarditis is supported by group B streptococcal septicaemia, symptoms and signs of disseminated infection, and vascular and immunological phenomena. She also had a purpuric rash, which, in conjunction with high fever and rigors, is suggestive of leucocytoclastic vasculitis. This can be seen in severe bacterial infections and is not uncommon in infective endocarditis.[2] PDA is a vascular connection between the roof of the pulmonary trunk near the origin of the left pulmonary artery and the proximal descending aorta. In the fetal circulation it allows the right ventricle to bypass the high resistance pulmonary circulation by pumping into the descending aorta. However, its persistence after the first few weeks of life is abnormal—it has a prevalence of one in 500 and it accounts for up to 10% of all forms of congenital heart disease.[3] Although a very small PDA may not be apparent on clinical examination, our patient had the classic sign of a PDA—a continuous systolic and diastolic "machinery" murmur over the upper left sternal edge. Importantly, infective vegetations in patients with PDA usually occur at the pulmonary artery end of the ductus (probably because of turbulent left to right flow), with embolic events occurring in the lung rather than the systemic circulation.[4]

The current incidence of PDA endarteritis in the United Kingdom is thought to be lower than the 0.45-1% per year before routine surgical closure and antibiotics became widely available in the mid-20th century.[5]

3.

What investigation would confirm the cause of this abnormality?

Short answer

Computed tomographic pulmonary angiography (CTPA) would confirm left sided pulmonary embolism leading to distal pulmonary infarction.

Discussion

In patients with suspected pulmonary embolism a clinical scoring system such as the modified Well's score can help to determine the clinical probability and need for further diagnostic imaging.[6] D-dimer testing can reduce the need for additional testing of low risk patients but is not helpful here owing to the high pre-test probability and suspected non-thrombotic pulmonary embolism. Although ventilation/perfusion (V/Q) scans are more sensitive for the detection of chronic thromboembolic pulmonary emboli, in this case a computed tomographic pulmonary angiogram (CTPA) was more useful because it can help delineate other chest pathology and confirm the acute left sided pulmonary embolism with pulmonary infarction. CTPA is a quick, accurate, and sensitive first line investigation for diagnosing acute pulmonary embolism in patients at intermediate or high risk.[7] The coronal CTPA image (fig 3) demonstrates the filling defect caused by septic pulmonary emboli within a branch of the left pulmonary artery (red arrow) with distal lung infarction (yellow arrow), as seen in the chest radiograph. The calcified PDA (purple arrow) can be seen between the distal aortic arch and the roof of the pulmonary artery below.

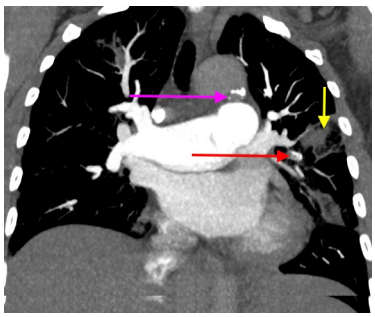

Fig 3 Coronal computed tomographic pulmonary angiogram showing septic pulmonary emboli (red arrow) with resultant pulmonary infarction (yellow arrow). The calcified patent ductus arteriosus (purple arrow) that connects the non-contrast enhanced aortic arch with the contrast enhanced main pulmonary artery below can also be seen

4.

What is the initial cardiac imaging technique of choice?

Short answer

Echocardiography (initially with a transthoracic study) is the initial imaging technique of choice for investigating suspected endocarditis.

Discussion

The unifying diagnosis of PDA associated streptococcal endocarditis and endarteritis would be further supported by echocardiographic evidence of a vegetation or endocardial involvement. The diagnosis of infective endocarditis is generally based on clinical, microbiological, and echocardiographic findings. The modified Dukes's criteria (box 1) have a sensitivity and specificity of more than 80% for the clinical diagnosis of infective endocarditis and are the reference criteria for diagnosis.[8] Specifically, two major criteria, one major and three minor criteria, or five minor criteria need to be met.

Transthoracic echocardiography is the initial technique of choice and is often preferred for assessing right sided valves and the severity of associated valvular dysfunction.[9 10] In this case transthoracic echocardiography allowed visualisation of the right sided heart structures, particularly the PDA vegetation as it protrudes into the pulmonary artery (fig 4).

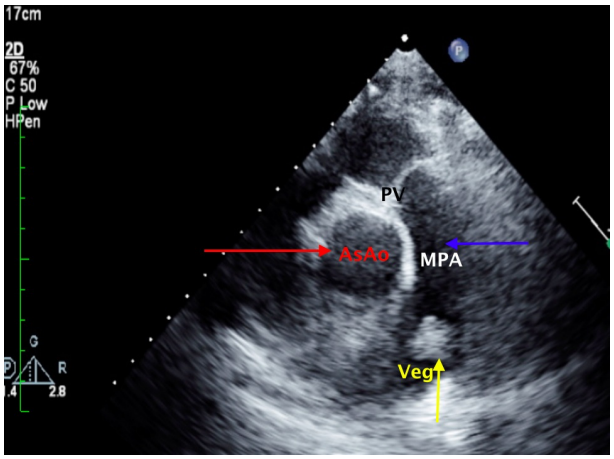

Fig 4 Parasternal short axis transthoracic echocardiogram at the level of the pulmonary valve showing the vegetation (yellow arrow; Veg) within the main pulmonary artery (blue arrow; MPA) beside the ascending aorta (red arrow; AsAo)

This clip can be viewed online with a video showing the PDA flow on colour Doppler jet reflecting blood flowing back into the pulmonary artery owing to the overlying connection from the aortic arch (fig 5).

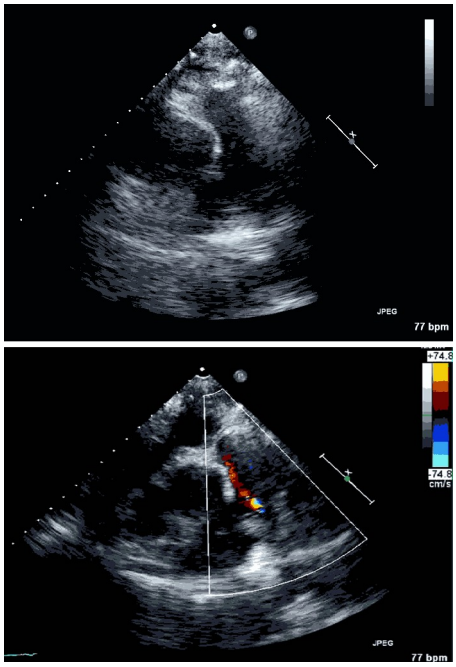

Fig 5 Video showing the patent ductus arteriosus flow on colour Doppler jet reflecting blood flowing back into the pulmonary artery owing to the overlying connection from the aortic arch

Patient outcome

CTPA confirmed that the Hampton's hump seen on chest radiography was caused by a septic pulmonary embolism with infarction in the left mid zone. Open cardiothoracic surgery was performed with patch closure of the PDA and vegetectomy. Her course was complicated by disseminated infection, including septic arthritis of the knee (requiring joint washout) and cervical discitis. Six weeks of intravenous antibiotics were needed before she fully recovered.

Competing interests: We have read and understood BMJ policy on declaration of interests and declare the following interests: none.

Provenance and peer review: Not commissioned; externally peer reviewed.

Patient consent obtained.

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Box 1: Modified Dukes's criteria for the diagnosis of infective endocarditis[7]

Major

- Microbiology—Blood culture positive for infective endocarditis (typical organism from two separate blood cultures or consistent organism from at least two blood cultures drawn >12 h apart)
- Echocardiography—Evidence of endocardial involvement (vegetation or new valvular regurgitation)

Minor

- Predisposition to infective endocarditis (such as injecting drug use)
- Fever (>38°C)
- Vascular phenomena (such as major arterial emboli, septic pulmonary infarcts)
- Immunological phenomena (such as glomerulonephritis)
- Microbiological evidence (positive blood culture not meeting major clinical criterion)