

Pulmonary emboli and extramedullary haematopoiesis in β -thalassaemia intermedia

Joel Dunn¹, Sameer Khan¹, Ben Ariff², Nicola Strickland²,
Adil Al-Nahas¹

¹Department of Nuclear Medicine, Hammersmith Hospital, London, United Kingdom

²Department of Imaging, Hammersmith Hospital, London, United Kingdom

[Received 14 II 2008; Accepted 27 VIII 2008]

Abstract

We present a case of a 52-year-old woman with β -thalassaemia intermedia under investigation for pulmonary hypertension. V/Q scan demonstrated bilateral pulmonary emboli. In addition there was evidence of central, posterior matched defects. CT pulmonary angiogram confirmed the presence of extramedullary haematopoiesis (EMH) in the paravertebral region. Although a few case reports of pulmonary emboli in thalassaemia intermedia have been published, to the best of our knowledge there have been no reports of matched defects caused by extramedullary haematopoiesis. This may be mistaken for enlarged pulmonary arteries, which are also seen in patients with pulmonary hypertension.

Key words: thalassaemia intermedia, extramedullary haematopoiesis, pulmonary hypertension, V/Q scan

Case report

A 52-year-old woman with β -thalassaemia intermedia was admitted to Hammersmith Hospital for investigation of pulmonary hypertension that was diagnosed on echocardiogram. An initial PA chest radiograph demonstrated a left paraspinal mass and

a large right paravertebral mass, which was initially thought to be due to gross enlargement of the pulmonary arteries. There were no significant focal lung lesions.

The patient then underwent ventilation perfusion scintigraphy using krypton-81m gas and Tc-99m MAA. The anterior and posterior views demonstrated bilateral mismatched perfusion defects (Figure 1) indicating bilateral pulmonary emboli as a probable cause for the pulmonary hypertension. In addition, central matched defects were noted. These were clearer and well defined on the posterior images, in keeping with posterior mediastinal masses. Enlarged pulmonary arteries (associated with pulmonary hypertension, which may be secondary to pulmonary emboli) are located within the middle mediastinum. Extramedullary haematopoiesis was suggested as a possible cause.

CT pulmonary angiogram was then performed which demonstrated bilateral paraspinal masses in the posterior mediastinum (more marked on the right side, corresponding to the V/Q findings) (Figure 2). The masses were well defined and of soft tissue and fat density, without calcification or cavitations in keeping with extramedullary haematopoiesis (EMH) (Figure 3).

Discussion

Extramedullary haematopoiesis is a well-described compensatory mechanism characterised by the production of red cells outside the bone marrow. The phenomenon occurs as a reactive process when red cell formation is insufficient for a long period and therefore unable to meet with red cell body demands. This may occur when red cells are rapidly removed from the blood or when their formation in the bone marrow is compromised [1]. Most cases of EMH are associated with chronic anaemia, especially myelofibrosis, thalassaemia, sickle cell anaemia and Vitamin B₁₂ and folate deficiency [2].

Patients with thalassaemia intermedia are at increased risk of developing extramedullary, mainly paraspinal, haematopoiesis when massive bone marrow proliferation fails to maintain red cell equilibrium. EMH is well recognised in the liver, spleen, adrenals, retroperitoneum, lymph nodes, breast, skin, dura matter, brain, heart and kidneys [1].

Correspondence to: Adil AL-Nahas
Department of Nuclear Medicine, Hammersmith Hospital
Du Cane Road, London W12 0HS, United Kingdom
Tel: +44 208 3834923, fax: +44 208 3831700
e-mail: adil.al-nahas@imperial.nhs.uk

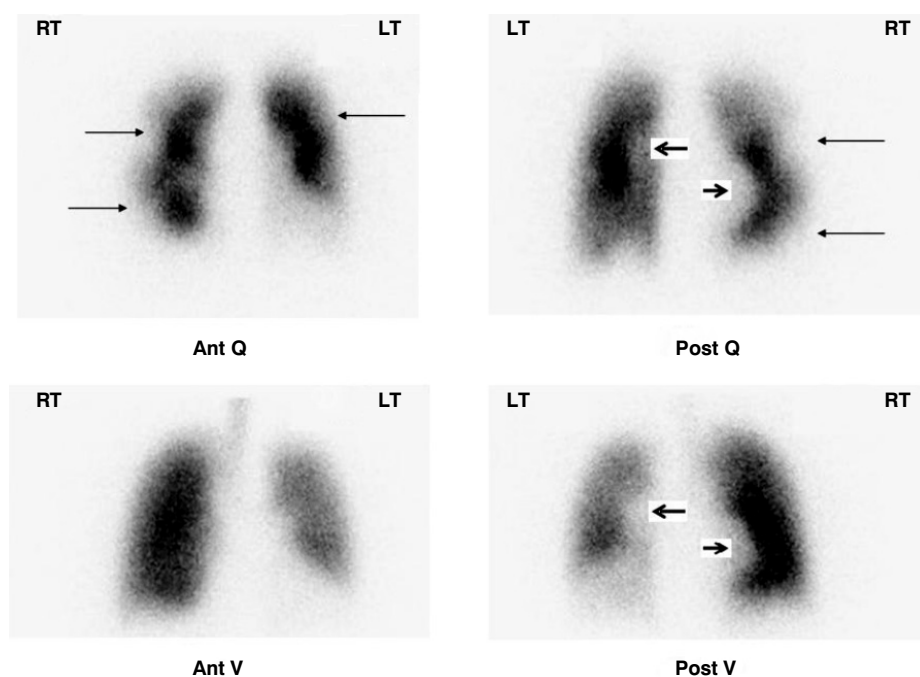


Figure 1. A 52-year-old woman with thalassaemia intermedia being investigated for pulmonary hypertension underwent V/Q scanning using krypton-81m gas and Tc-99m MAA. The anterior and posterior views demonstrate mismatched perfusion defects in keeping with bilateral pulmonary emboli (closed arrows). β -thalassaemia intermedia is a recognised risk factor for thromboembolic disease [4]. In addition, centrally matched defects were noted (open arrows). Although enlarged pulmonary arteries, which are an associated finding in pulmonary hypertension, may cause a similar appearance, these masses are much more apparent on the posterior images suggesting a paraspinal position.



Figure 2. The patient underwent a CT pulmonary angiogram. The coronal reformat demonstrates paraspinal masses in the posterior mediastinum (more marked on the right side, corresponding to the V/Q findings), in keeping with extramedullary haematopoiesis (EMH). Intrathoracic EMH most commonly occurs in the posterior mediastinum and in the lower thoracic paraspinal area [1].

The histology of the localised paraspinal lesions of EMH depends on the duration of its existence and the erythropoietic demands of the particular patient. Initially paraspinal extramedullary sites of haematopoiesis reveal immature and mature cells, mainly of the erythroid and myeloid series, and dilated sinusoids contain-

ing precursors of red cells. Later the lesion becomes inactive and reveals some fatty tissue and fibrosis or massive iron deposit [1].

Intrathoracic EMH may be visualised on plain film chest radiograph or chest CT scan as single or multiple paravertebral mass lesions. There are, however, a wide range of aetiologies with similar radiological findings, including neurogenic tumours, lymphoma and both primary and metastatic malignancy. Characteristic features seen on chest radiograph and chest CT are important in recognising intrathoracic EMH [3].

Radionuclide bone marrow scanning using ^{99m}Tc sulphur colloid has been used previously to help establish a diagnosis of EMH [2]. To the best of our knowledge, this is the first occasion EMH has been described on ventilation/perfusion scintigraphy.

In the case we present, the anterior and posterior V/Q views demonstrate bilateral pulmonary emboli (Figure 1, closed arrows), which was anticipated since thalassaemia intermedia is a recognised risk factor for thromboembolic disease [4]. In addition, central matched defects were noted (Figure 1, open arrows). Although enlarged pulmonary arteries, which are an associated finding in pulmonary hypertension, may cause a similar appearance, these masses are much more apparent on the posterior images suggesting a paraspinal position.

The presence of EMH was later confirmed by CT imaging using the characteristic features for non-invasive diagnosis of EMH [5, 6].

Conclusions

Extramedullary haematopoiesis is a rarely encountered response to bone marrow dysfunction, in which diagnosis made by non-invasive means is desirable due to the highly vascular nature

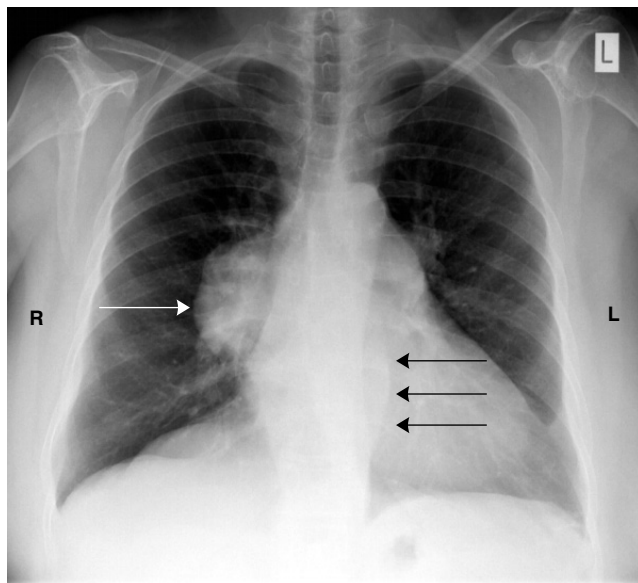


Figure 3. On the PA chest radiograph the EMH is seen as a left paraspinal mass (black arrows) disrupting the left paraspinal line and a large right paravertebral mass (white arrow), through which the normal pulmonary vessels are demonstrated, indicating the posterior position of the EMH [7].

of the masses. Characteristic features on plain film and CT imaging alone are usually sufficient in establishing a diagnosis of EMH.

We report an unusual case of centrally matched defects on ventilation perfusion scintigraphy which were highly suggestive of posterior mediastinal lesions and helped to establish the diagnosis of these lesions as EMH on CT imaging.

References

1. Tsitouridis J, Stamos S, Hassapopoulou E, Tsitouridis K, Nikolopoulos P. Extramedullary paraspinal hematopoiesis in thalassemia: CT and MRI evaluation. *Eur J Radiol* 1999; 30: 33–38.
2. Bolaman Z, Polati M, Cildag O, Kadikoylu G, Culhaci N. Intrathoracic extramedullary hematopoiesis resembling posterior mediastinal tumor. *Am J Med* 2002; 112: 739–41.
3. Alam R, Padmanabhan K, Rao H. Paravertebral mass in a patient with Thalassemia Intermedia. *Chest* 1997; 112: 265–266.
4. Shmuel G, Cappellini MD, Goldfarb A et al. Pulmonary thromboembolism in thalassemia intermedia patients. *Haematologica* 1999; 84: 959–960.
5. Gumbs R, Ford EAH, Tea JS et al. Intrathoracic extramedullary hematopoiesis in sickle cell disease. *AJR* 1987; 149: 889–893.
6. Fielding JR, Owens M, Naimark A. Intrathoracic extramedullary haematopoiesis secondary to B12 and folate deficiency: CT appearance. *J Comput Assist Tomogr* 1991; 15: 308–310.
7. Whitten CR, Khan S, Munneke G, Grubnic S. A diagnostic approach to mediastinal abnormalities. *Radiographics* 2007; 27: 657–671.