

Case report

Primary choriocarcinoma of the pulmonary artery mimicking pulmonary embolism

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Abstract. Due to their rarity, primary tumours of the pulmonary arteries are often incorrectly diagnosed as more common diseases such as pulmonary thromboembolism and are seldom diagnosed during a patient's lifetime. Surgery or potentially curative chemotherapy may therefore be withheld. We report a patient with a primary choriocarcinoma of the left pulmonary artery, which was first suspected on a CT scan. The neoplastic nature of the obstruction of the pulmonary arteries was confirmed by positron emission tomography.

Case report

A 33-year-old female with a 5 month history of recurrent episodes of exertional dyspnoea, pleuritic chest pain and an urge to cough was referred to hospital with the tentative diagnosis of pulmonary embolism. She had had an uncomplicated delivery 3 years previously and an abortion 10 months prior to admission. Following this abortion, there was an elevated level of β -human chorionic gonadotropin (β -hCG) at 600 IU l^{-1} . No trophoblastic tissue was found at laparoscopy but the patient refused further investigations. On admission, she was cyanotic and tachypnoeic at rest. She complained of left-sided pleuritic chest pain. On examination, reduced breath sounds in the left upper chest and tachycardia (110 bpm) were noted. The initial anteroposterior supine chest radiograph showed peripheral wedge-shaped opacities in the left upper and the right lower lobes, bilateral pleural effusions and signs of pulmonary arterial hypertension (Figure 1). The initial mean pressure in the pulmonary artery was elevated to 34 mmHg (normal below 15 mmHg). The electrocardiogram revealed no sign of right heart strain. At presentation, the serum β -hCG concentration was elevated to 129.500 U l^{-1} (normal up to 5 U l^{-1}). Ultrasonography, MRI and laparoscopy excluded an extrauterine pregnancy. Subsequent contrast enhanced CT of the hilar region demonstrated an intraluminal contrast filling defect in the left pulmonary artery (Figure 2) and multiple peripheral well defined opacities in both lungs. Compared with the pre-contrast CT scan, the intraluminal mass showed heterogeneous enhancement of up to 48 Hounsfield units. Pulmonary angiography

demonstrated complete occlusion of the left pulmonary artery and occlusion of peripheral branches of the right inferior pulmonary artery. MRI of the chest could not be performed due to the poor state of health. Positron emission tomography with 2-[fluorine-18]-fluoro-2-deoxy-D-glucose (FDG-PET) confirmed the neoplastic origin of the intraluminal mass with increased metabolism overlying the left pulmonary artery (Figure 3). Peripheral opacities in the left upper and right lower lobe with slightly increased metabolic activity solely in their periphery were suggestive of pulmonary abscess formation due to infarction after tumour

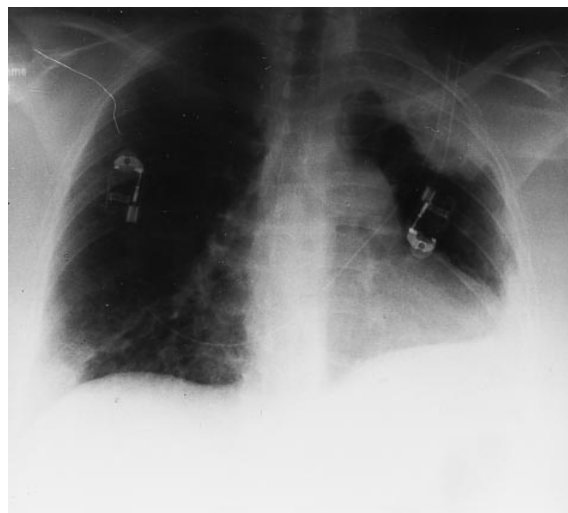


Figure 1. Posteroanterior chest radiograph showing peripheral wedge-shaped opacities in the left upper and right lower lobes. Left-sided pleural effusion and pulmonary arterial hypertension with fullness of the main pulmonary artery and an abrupt decrease of the caliber of peripheral branches.

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Figure 2. Contrast enhanced CT scan of the hilar region showing heterogeneous enhancement of an intraluminal mass in the left pulmonary artery (arrow). A well defined opacity in the left upper lobe with incomplete rim enhancement (arrowhead) was an abscess.

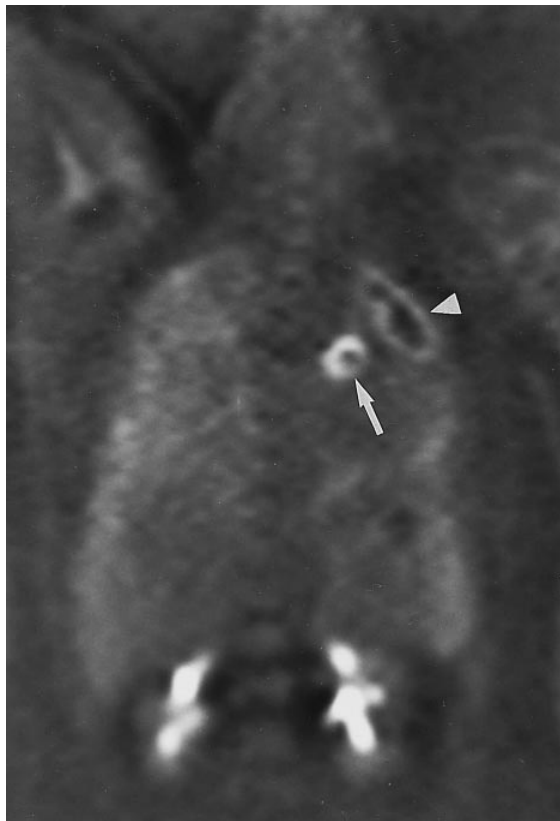


Figure 3. Coronal FDG-PET depicts two regions of increased metabolism: one in the left hilar region corresponding to the primary choriocarcinoma of the left pulmonary artery (arrow); the other region in the left upper lobe with slightly increased metabolism in its periphery representing an abscess formation (arrowhead).

embolism. The patient died of acute cardiopulmonary insufficiency 1 day after administration of methotrexate (0.4 mg kg^{-1}). At autopsy, a tumour was found occluding the left pulmonary artery extending into the right atrium. Multiple infarcts

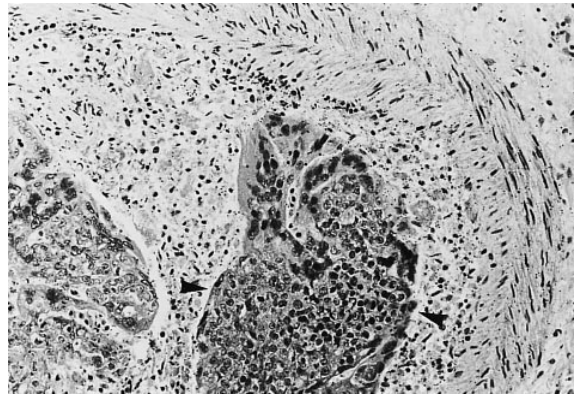


Figure 4. Histology of the tumour shows polymorphic tumour cells with hyperchromatic nuclei (arrowheads), surrounded by extensive areas of necrosis, representing a choriocarcinoma.

in both lungs were caused by disseminated tumour cell emboli. Histological evaluation showed a primary choriocarcinoma (Figure 4). The autopsy disclosed no evidence of a primary tumour in the abdomen or mediastinum.

Discussion

Primary malignant tumours arising from the pulmonary arteries are extremely rare. Bagshaw and Brooks were the first to suggest that pulmonary embolism and pulmonary hypertension could be due to a choriocarcinoma of the pulmonary arteries [1]. To our knowledge, only four cases of primary choriocarcinoma of the pulmonary arteries diagnosed during the patients' lifetime have been reported [2, 3].

It is very important to be aware of this clinical presentation since primary choriocarcinoma may be cured by chemotherapy. Non-specific symptoms that may lead to the diagnosis include chest pain, dyspnoea, cyanosis, non-productive cough and haemoptysis. A high serum or urinary β -hCG in fertile women in combination with these symptoms should alert the physician [4]. Since active trophoblastic tissue always produces β -hCG, this blood test can either confirm or exclude the possibility of choriocarcinoma [5]. Nevertheless, the diagnosis is difficult to establish because of the tendency of these tumours to masquerade as more common diseases. The most likely differential diagnosis is pulmonary thromboembolism, followed by other primary neoplasms of the pulmonary arteries such as sarcoma or leiomyosarcoma which are more common than choriocarcinoma [6, 7]. Chest radiography may show no specific signs [2], but can reveal pulmonary embolism associated with pulmonary artery hypertension as in our case. Radiological signs of peripheral pulmonary hypoperfusion and fullness of the pulmonary trunk have also been reported in patients with primary

sarcoma of the pulmonary arteries [4, 6]. In the majority of cases, pulmonary scintigraphy shows hypoperfusion that varies with the extent and localization of the tumour [7]. Angiography does not usually distinguish between intraluminal tumour and thromboembolus of the pulmonary arteries. The non-specific angiographic finding consists of deficient filling of one or both pulmonary arteries with abrupt interruption to flow of contrast medium. Pulmonary angiography can be fatal because of fragmentation or dislocation of the tumour by the catheter tip [2, 3]. CT and MRI of the chest may disclose more detailed anatomy of the neoplasm. Contrast enhanced CT may allow differentiation between vascular tumour showing enhancement and non-enhancing intraluminal blood clot. In our patient we observed heterogeneous enhancement of the intraluminal mass up to 48 Hounsfield units. The diagnostic potential of FDG-PET in detecting malignant tumours has been reported previously [8] but there are no published cases of FDG-PET in patients with primary choriocarcinoma of the pulmonary artery. In the present case, FDG-PET confirmed the neoplastic origin of the intraluminal mass by an increase in glucose metabolism which would be unlikely to occur in an ordinary thromboembolus.

Once the diagnosis is suspected, high dose chemotherapy (methotrexate $0.4 \text{ mg kg}^{-1} \text{ day}^{-1}$) promises an excellent outcome [2, 3] and should be initiated immediately. The disease course can

be monitored clinically by serial β -hCG measurements and contrast enhanced CT. Definitive tissue diagnosis is not essential before therapy, since β -hCG is a reliable tumour marker for choriocarcinoma.

References

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