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Case report

Fatal idiopathic pulmonary haemosiderosis in association with pregnancy – Medico-legal evaluation

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ABSTRACT

Idiopathic pulmonary haemosiderosis is a rare disorder characterised by repeated episodes of intra-alveolar bleeding in association with consecutive anaemia, pulmonary fibrosis, pulmonary hypertension and respiratory failure. Pregnancy may exacerbate the symptoms of idiopathic pulmonary haemosiderosis typically worsening in the third trimester. A 32-year-old female after delivery was admitted to hospital with progressive dyspnoea of about 1-month duration. Sudden circulatory collapse caused fatal complication. During the post-mortem investigation, lung haemorrhage and histologically abundant iron deposition in macrophages and interstitial fibrosis were found. Medico-legal post-mortem evaluation of fatal cases may support the clinico-pathological context of the diagnosis of this entity.

1. Introduction

Idiopathic pulmonary haemosiderosis (IPH) is a very rare disorder of unknown aetiology characterised by recurrent or chronic episodes of alveolar haemorrhage and accumulation of haemosiderin in the lung parenchyma.[1,2] The incidence of IPH is unknown; however, studies suggest that it lies between 0.24 and 1.23 cases per million live births per year.[3] Clinical manifestations of IPH include iron-deficiency anaemia without any known cause, pulmonary symptoms such as haemoptysis, progressive dyspnoea and cough and parenchyma lesions.[1] IPH over time leads to progressive secondary pulmonary fibrosis, pulmonary hypertension and cor pulmonale. It is a disease of childhood and usually occurs in children before 10 years of age; however, it could occur in young adults. Females with IPH rarely survive to childbearing age or are unable to become pregnant.[3] IPH has rarely been reported in pregnancy[4,5]; however, pregnancy may exacerbate the symptoms of IPH with the symptoms typically worsening in the third trimester.[6–8] Death may occur suddenly from acute pulmonary haemorrhage or progressive respiratory failure.[9] Post-partum complications have a great public interest and require medico-legal evaluation.

We report a case of a young woman with fatal IPH after pregnancy due to acute pulmonary haemorrhage.

2. Case report

2.1. History

A 32-year-old female was admitted to hospital with progressive dyspnoea of 1-month duration starting straight after delivery. Pregnancy was uncomplicated except for slight gynaecologic infection treated with antibiotic. Her symptoms started with chest pain and fever without any typical alteration on a chest computer tomogram. Antibiotic treatment resulted in temporary clinical remission; however, later, progressive effort dyspnoea started. By the time of her hospital admission, she had pulmonary hypertension (pulmonary arterial pressure (PAP) 67–93 mmHg) and respiratory failure with severe hypoxaemia (arterial blood gas: alveolar oxygen pressure (PaO_2): 6.84 kPa; arterial oxygen saturation (SaO_2) 89% during 3 l min^{-1} oxygen supplementation), and there was no experience of haemoptysis. A computer tomogram–pulmonary

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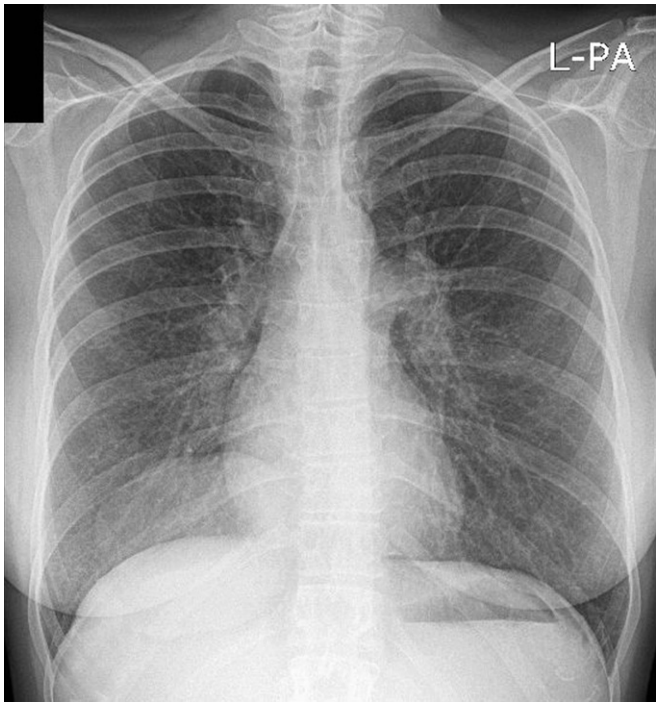

Fig. 1. Chest X-ray shows right-sided pleural effusion with sharp margin above the diaphragm. In the right lower lobe there are opacities caused by compressed parenchyma. Hilum is slightly enlarged due to dilated pulmonary artery.

angiogram demonstrated no evidence of thrombo-embolism, and confirmed bilateral pleural effusion, transitory pulmonary infiltrate and very slight parenchyma opacities; and right hilar lymphadenopathy were noted on chest X-ray (Fig. 1).

During observation, an acute episode with chest pain, haemoptysis and deterioration of her respiratory and radiology status occurred. Laboratory analysis did not support anaemia. Bronchoscopy was performed and revealed normal airways with some blood leakage from right lower lobe. Analysis of broncho-alveolar lavage fluid was not diagnostic; the patient's respiratory status did not allow for trans-bronchial lung biopsy. Immunological evaluation did not support antineutrophil cytoplasm antibodies (ANCAs) or anti-glomerular basement membrane antibodies' (anti-GBMs) positivity; however, other auto-immunity affecting lungs could not be completely excluded. Improvement of tissue oxygenation and decline of PAP (PAP 46 mmHg, PaO_2: 8.17 kPa; SaO_2 93% during 1.5 l min^{-1} oxygen supportation) was obtained, supported by high-dose methylprednisolone therapy. Lung biopsy could be indicated in case of improvement of clinical status. The second acute relapse of disease led to a sudden collapse of her circulation with unavailable resuscitation.

2.2. Autopsy findings

A complete forensic autopsy was performed. On opening the thoracic cavity, there was 1100 ml blood-tinged fluid in the right pleural cavity. Lungs weighed 2100 g, and a massive circumscribed haemorrhage – 10 cm in diameter – was observed in the right lower lobe and in the lower margin of upper lobe. The pleura was dull, opaque, and thickened. In the parenchyma, several haemorrhagic focuses 0.5–3 cm in diameter were detected around the circumscribed dark area (Fig. 2). The pulmonary vessels were dilated and could be traced to the sub-pleural surface. The heart weighed 290 g, and there was marked dilatation of the right atrium, right ventricular cavity and pulmonary trunk. Other internal organs showed hypoxic changes.

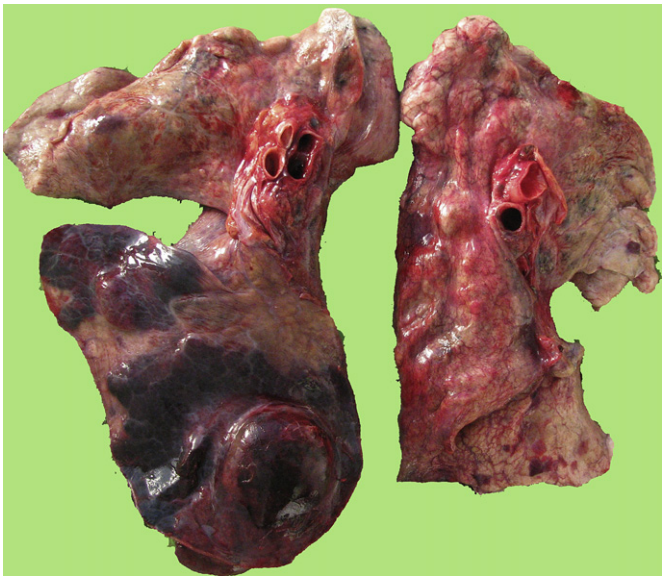

Fig. 2. Gross photograph showing a large and several smaller pulmonary hemorrhages in the right lung.

2.3. Histopathology

Tissue specimens from every internal organ were fixed in paraformaldehyde, embedded in paraffin and stained by haematoxylin–eosin (HE) and Prussian blue (haemosiderin-bound unreactive Fe^{3+}) procedures. Microscopically, thickening of pleura and alveolar walls was revealed and a severe interstitial fibrosis was observed. Haemosiderin-laden macrophages filled the compressed alveolar spaces, and in some areas a few iron-laden macrophages were detected also in thickened alveolar septa (Fig. 3(A) and (B)). In other internal organs, no iron deposition was found.

3. Discussion

In this report, we presented a case of a young woman who had post-partum pulmonary symptoms, and died suddenly by the

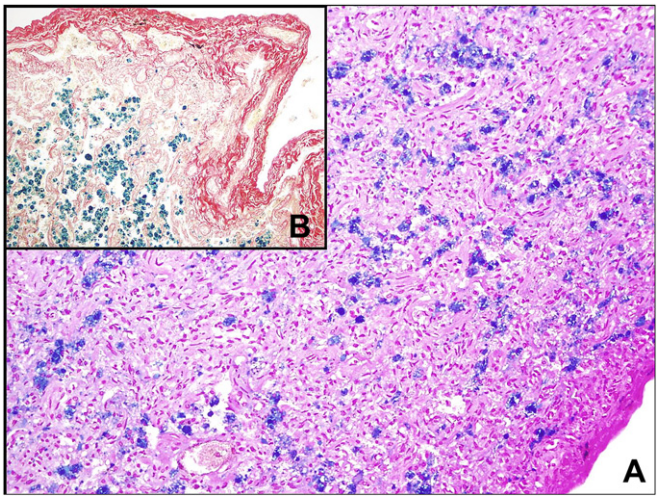

Fig. 3. A,B – Idiopathic pulmonary hemosiderosis. Fibrosis of the lung and Prussian blue positive pigment containing macrophages in alveoli (×200). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)