

CASE STUDY

Fatal haemorrhage from mesenchymal cystic hamartoma of the lung

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Fatal haemorrhage from mesenchymal cystic hamartoma of the lung. S.L. Chadwick, B. Corrin, D.M. Hansell, D.M. Geddes. ©ERS Journals Ltd 1995.

ABSTRACT: A 34 year old woman was admitted to the hospital with a 9 year history of intermittent haemoptysis associated with increasing breathlessness. A working diagnosis of lymphangioleiomyomatosis was made, based on clinical, radiological and histological findings.

Three years later, the patient was admitted to hospital with worsening haemoptysis, which rapidly progressed and resulted in death from massive pulmonary haemorrhage. Postmortem examination and histology revealed findings consistent with multiple mesenchymal cystic hamartomas of the lungs.

This is a rare condition which has previously been described as having a good prognosis. This case is the first fatality resulting directly from the disease to be reported.

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Mesenchymal cystic hamartoma of the lung, first described in 1986 [1], is characterized by nodules of immature mesenchymal cells that gradually increase in size and then become cystic. The diagnosis is made on the basis of its characteristic histology. There have been eight previous case reports, and in all of these the disease has followed a relatively benign course [1–4]. Although haemorrhage into the cysts has previously been described, this is the first fatality from pulmonary haemorrhage to be reported.

Case report

A 34 year old woman complained of intermittent haemoptysis occurring approximately once every 2 months for 10 yrs. These episodes were unrelated to chest infections, exercise or menses. Chest radiographs 10 yrs previously had been reported as showing widespread nodular opacities measuring 0.5–3 cm in diameter, but bronchoscopy and needle biopsy had failed to identify a cause for the haemoptysis. One year later, the radiographic appearances had progressed and multiple cystic lesions were reported.

On admission, the patient was severely breathless but in good general health. Examination was unremarkable and laboratory investigations revealed only a microcytic anaemia (haemoglobin 84 g·L⁻¹). Chest radiography showed multiple fluid-filled, thin-walled cysts ranging 2–10 cm in diameter, and a right pneumothorax (fig. 1). In addition to the cysts, computed tomography (CT) revealed a few noncavitating soft tissue pulmonary nodules, 0.4–1 cm in diameter (fig. 2).

An open lung biopsy was performed, and it was noted that the pleural space was partially obliterated by

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Fig. 1. – Chest radiograph showing multiple large thin-walled cysts containing fluid levels and a right pneumothorax.

adhesions and that the lung contained nodules and many plum-coloured cysts. Lymphangioleiomyomatosis was diagnosed and the patient was treated with depot injections of medroxyprogesterone acetate, without any evident beneficial effect apart from a decrease in her previously troublesome menorrhagia and an increase in her haemoglobin to 143 g·L⁻¹.

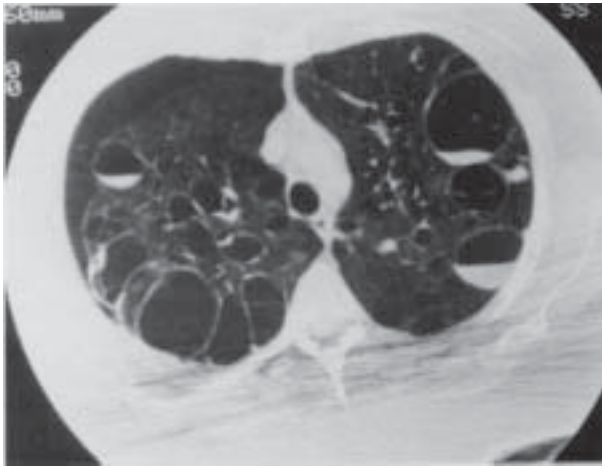


Fig. 2. – Computerized tomography showing fluid in some of the cysts and at least one nodule in the left upper lobe.

Three years later, the patient was admitted to her local hospital with increasingly severe haemoptysis. Chest radiographs showed fluid filling the left side of the chest, presumed to be haemorrhagic. Her condition deteriorated, with progressive respiratory failure. Artificial ventilation was instituted but was unsuccessful, and she died from a combination of bleeding and respiratory failure.

Pathology

The open lung biopsy showed cystic change and marked haemosiderosis, consistent with previous haemorrhage. Elastic fibres in the walls of alveoli and small blood vessels were refractile, basophilic or brown due to calcium and iron encrustation, fragmented and attended by foreign body giant cells. The walls of the cysts showed proliferation of stellate or spindle cells that resembled the immature smooth muscle of lymphangiomyomatosis, but failed to stain for smooth muscle actin. Similar cells occluded small veins, and this postcapillary block was thought to account for the pulmonary haemorrhage.

At autopsy, both lungs were largely replaced by blood-filled cysts, measuring up to 8 cm in diameter and separated by areas of normal tissue. Other systems were unremarkable. Microscopy showed that the cysts were lined by ciliated epithelium, beneath which was a prominent layer of immature mesenchymal cells up to 2 mm thick, corresponding to the cambium layer described in mesenchymal cystic malformation of the lung [1].

Discussion

In the eight previously reported cases of mesenchymal cystic hamartoma of the lung, the disease has followed a relatively benign course. One of the five patients reported by MARK [1] subsequently died of a neoplasm and the original diagnosis may have been wrong [5]. It has been brought to our attention that Mark's first case has

also been reported as one of pulmonary metastases of endometrial stromal sarcoma [6]. It is, therefore, worth emphasizing that at postmortem (performed by LC. Ilesley, Kent and Canterbury Hospital) our patient showed no extrapulmonary abnormalities whatsoever. The uterus contained a contraceptive device but the endometrium and myometrium were normal, both macroscopically and microscopically, as were the ovaries. The lung lesions in our case failed to stain for either desmin or smooth muscle actin, further evidence that our patient was not suffering from metastatic endometrial stroma sarcoma.

In one of the cases reported by MARK [1], brisk pulmonary haemorrhage requiring thoracotomy occurred, but ours is the first report of death from this condition. The disease appears to follow a variable course, with some patients having only small numbers of cysts and scattered noncystic nodules, and some patients, such as ours, in whom over a period of years the nodules become cysts which virtually destroy the normal architecture of the lung. The clinical features of the disease are correspondingly highly variable. Some patients may be asymptomatic [3] whilst others experience dyspnoea, haemoptysis, pneumothorax or haemothorax, or any combination of these symptoms.

Our initial working diagnosis was pulmonary lymphangiomyomatosis, of which our patient had the following characteristics: female gender, age in the reproductive years, pulmonary cysts, pneumothorax, and pulmonary haemorrhage attributable to a mechanism (postcapillary block) previously understood to account for the bleeding of lymphangiomyomatosis [7]. Against this diagnosis was the unusually large size of the cysts, their irregularity and the presence of solid nodules. In lymphangiomyomatosis the cysts are thin-walled and of fairly uniform size, no greater than 3 cm in diameter, and there is no nodular component [8, 9]. Our inability to stain for smooth muscle actin was also contrary to the diagnosis of lymphangiomyomatosis, but only postmortem histology showed the cambium layer of immature mesenchymal cells, beneath normal ciliated epithelium that is pathognomonic of mesenchymal cystic hamartoma.

Mesenchymal cystic hamartoma of the lung cannot always be considered to carry a good prognosis. It comprises a broad spectrum of disease ranging from non-progressive nodular disease and isolated cysts to gradually but relentlessly expanding cysts, which disrupt the normal architecture of the lung. Uncontrollable pulmonary haemorrhage is, as in this case, a potentially fatal complication.

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