

Lymphangioliomyomatosis presenting in a postmenopausal woman

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Lymphangioliomyomatosis is a rare pulmonary disorder characterised by proliferation of smooth muscle diffusely throughout the lung parenchyma. Most patients present during their reproductive years, and survival is generally less than 10 years. We report a case in a postmenopausal woman that was associated with long survival.

Case report

A 72 year old caucasian woman was admitted to hospital because of progressive respiratory failure. She had never been a smoker, and had worked as an audit supervisor for a department store. Her previous medical history included two severe episodes of pneumonia about 20 years before admission, and a hysterectomy at the age of 55. For the previous 10 years she had been investigated and treated for obstructive lung disease presumed to be due to late onset asthma. Seven years before admission she had shown appreciable gas trapping, with a total lung capacity of 110% predicted and a residual volume of 184% predicted; diffusing capacity was 33% predicted. At that time her FEV₁ was 0.5 l, forced vital capacity (FVC) 1.4 l, and forced expiratory flow (FEF₂₅₋₇₅) 0.3 l (14% predicted).

During this admission her arterial oxygen tension (Pao₂)

was 66 mm Hg (8.8 kPa), carbon dioxide tension (Paco₂) 56 mm Hg (7.5 kPa), and pH 7.38. Routine spirometry showed her FEV₁ to be 0.3 l (21% predicted) and FVC 0.7 l (33% predicted). These results were considered to be consistent with chronic airways obstruction. A chest radiograph showed hyperinflated lungs with a fine diffuse bilateral reticular pattern. The serum α₁ antitrypsin concentration was 410 (normal 220-380) mg/100 ml. She was treated with bronchodilators and oxygen, but she died of increasing respiratory failure with associated cor pulmonale.

At necropsy, the lung had a multicystic appearance with the characteristic microscopic findings of lymphangioliomyomatosis (figs 1 and 2). Similar nodules of irregular muscular proliferations were present in the hilar lymph nodes, and in a soft brown nodule present in a subdiaphragmatic position. The left kidney as well as both adrenals contained microscopic foci of smooth muscle arranged in irregular bundles with adjacent cystic spaces. The considerable right ventricular hypertrophy, with a left to right ventricular weight ratio of 1.2:1 (normal 2.3:1), correlated well with the clinical impression of cor pulmonale.

Discussion

Lymphangioliomyomatosis is an uncommon pulmonary condition. The aetiology and pathogenesis are obscure; the disease progresses, leading to death usually within 10 years.¹ It generally presents during the child bearing years, and this, together with exacerbations with menses² and during pregnancy,¹ has suggested some degree of hormonal

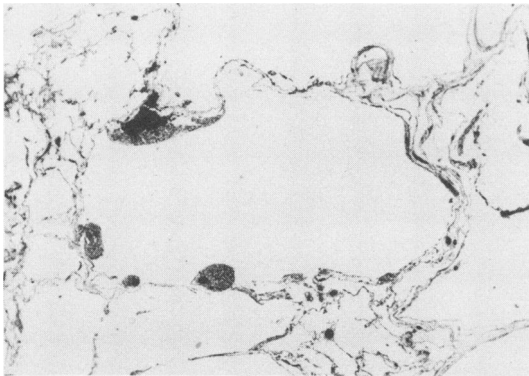


Fig 1 Typical area showing the very mild degree of muscle proliferation seen in the lungs of this patient. There are small irregular bundles of smooth muscle present in the walls of cyst like structures. (Masson's trichrome, × 12.)

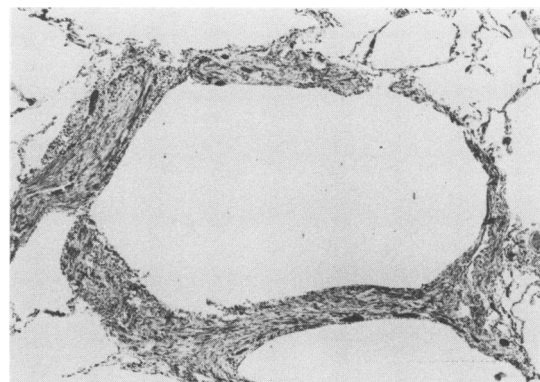


Fig 2 Higher magnification photomicrograph showing irregular bundles of smooth muscle fibres around a distorted small airway. (Masson's trichrome, × 38.)

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influence. Further evidence for hormonal effects in lymphangioliomyomatosis has recently emerged, with the documentation of steroid, oestrogen, and progesterone receptors in lung biopsy material.³ Treatment by oophorectomy⁴ or with progesterone⁵ even in receptor negative cases⁶ has resulted in clinical improvement.

Our patient is unusual in several respects: her advanced age on presentation, a previous history of hysterectomy with oophorectomy, and a prolonged course of disease. In a search of published reports we could find only four patients who were older than 55 when they developed symptoms.^{7,8} Three of these patients were alive at the time of review, surviving two, nine, and 19 years; while the fourth had died eight years after presentation. Two lung biopsies had been performed on the long term survivor⁷ with an interval of 10 years and histological examination had shown only mild progression of muscular proliferation.

Our patient had developed symptoms at the age of 60, five years after oophorectomy. We suggest that the slow progression of her disease reflects both its initiation in the perimenopausal period and the reduction of steroid dependent muscle proliferation by oophorectomy. Muscular proliferation in the lung in our patient was milder than in most cases.

Lymphangioliomyomatosis has characteristic radiological, pulmonary physiological, and pathological patterns.^{1,9} Inapparent hormonal influences may obscure the clinical findings and delay diagnosis. We believe that the pro-

longed course in this elderly postmenopausal woman provides further evidence for the role of oestrogenic steroids in this condition.

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