Short reports

Intravascular bronchioloalveolar tumour

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The intravascular bronchioloalveolar tumour, first described by Farinacci,1 is a rare neoplasm of unknown aetiology, thought by Dail and Liebow to behave as a malignant lesion.2 Only 28 cases have been published and the details of these are sparse.1,3 We report another case of intravascular bronchioloalveolar tumour and review the published reports.

Case report

A 42-year-old man presented with a two-week history of increasing right upper quadrant pain. He was admitted to hospital and found to have mildly raised bilirubin and alkaline phosphatase concentrations. The results of the oral cholecystogram, barium enema, and intravenous pyelogram were normal, but a barium-meal examination showed scarring of the duodenal bulb. The chest radiograph showed fibrous infiltrates in the left lower zone. A chest film taken previously, in June 1978, was clear; but retrospective review of a chest film of June 1979 showed a single linear shadow in the peripheral left lower zone. The remainder of the laboratory examination indicated only mild polyclonal increase in gammaglobulins with no indirect antinuclear antibody reactivity. The findings of oesophagogastroduodenoscopy and endoscopic retrograde pancreaticholangiography were normal. The tuberculin test gave negative results. Abdominal computed tomography showed three lesions in the left lower lung zone and no intra-abdominal pathological features. Bone scan, liver scan, and sonography of the liver gave negative results, and pulmonary function tests and arterial blood gas measurements gave normal values. The patient underwent diagnostic thoracotomy, at which three lesions were found. Two of these were firm and nodular, similar to the "popcorn" texture of a hamartoma. One lesion of the left upper lobe was removed by wedge resection and one lesion deep within the left lower lobe was enucleated. The histological features of these tumours were identical to the neoplasms described as intravascular bronchioloalveolar tumours.4 The third lesion was found to be a granuloma. Because of the known multicentric origin of the former tumours the procedure was terminated, and the patient made an excellent postoperative recovery. During his convalescence further diagnostic studies were performed. Computed tomography of the right lung showed no evidence of disease. Laparoscopy performed on the sixth postoperative day showed fibrinous adhesions between the right lobe of the liver and the abdominal wall. These were lyed laparoscopically and liver biopsy showed no evidence of tumour. The patient was discharged on the seventh postoperative day. Four months later, because of recurrent progressive right upper quadrant pain, the patient was admitted to another institution, where liver biopsy showed metastatic intravascular bronchioloalveolar tumour.

Discussion

The intravascular bronchioloalveolar tumour at present remains a pathophysiological curiosity. The progenitor cell is controversial. Although Dail and Liebow thought the tumour to be of bronchioloalveolar origin,2 recent electron-microscopic studies indicate a possible vascular origin from precursor mesenchymal vasoformative reserve cells.4 Symptoms caused by these lesions are minimal and many are found unexpectedly in the course of routine physical examination or in the investigation of an unrelated condition.1,3 Two-thirds of the cases have occurred in women; and although the age range is large, 14–71 years, 40% have been found in patients under 30 years of age (also paper presented by MS Segal, with notes from D Dail, Pulmonary Pathology Conference, La Jolla, California, 1978). The tumour is of multicentric origin and radiographically both lungs may contain multiple small, rounded, non-calcified opacities often mistaken for metastatic lesions or fibronodular infiltrates.2,4 No primary tumour has ever been found outside the lung4 and a search for environmental and aetiological factors has been unsuccessful.2 Diagnosis is made by exploratory thoracotomy, and at first sight the lesions have often been regarded as chondrosarcoma, chondromyxosarcoma, or multiple pulmonary hamartomas5 (and Segal, 1978, as above). These tumours are characterised histologically by microepithelial masses that fill alveoli and occasionally bronchioles and invade both arteries and veins, filling their lumina. While the periphery of the tumour appears epithelial, there are also chondrocyte-like elements, simulating the histology of a pleomorphic adenoma of the salivary gland. Centrally there is necrosis with residual masses of acellular material present within lacunar spaces (figs 1–2). The central necrosis, peripheral extension into pulmonary bronchioloalveolar vascular spaces, and multicentricity are most valuable in the recognition of this neoplasm. The tumour may have previously been regarded as a multicentric cartilaginous hamartoma, and apart from the unique features described above a cartilaginous origin is a reasonable suggestion. It has been reported that the central necrotic zones may calcify.5 The clinical course is one of slow progression until lung and pleura are extensively affected. Isolated metastatic
lesions have occurred in the liver and clavicle and regional lymphogenous spread has been reported. Those patients known to have died from this disease survived two, eight, and 12 years after diagnosis; while Dail and Liebow have been following a further patient six years after the diagnosis was established. The terminal course is characterised by rapid development of respiratory insufficiency. There is no known chemotherapy and radiotherapy is not known to be effective.

Our patient has developed two tumours within six months. As mortality is related to pulmonary disease and not distant metastases, until further information about the therapeutic sensitivity of this tumour is forthcoming we plan to treat new pulmonary nodules with local resection, preserving as much lung tissue as possible.

In conclusion, the intravascular bronchioloalveolar tumour is a rare multicentric neoplasm with a penchant for predominantly local and regional invasion. Although the diagnosis is usually not suspected before excision of the nodules, clues to the nature of these lesions at the time of exploration are their gritty, hamartomatous texture and multicentric occurrence. The course is prolonged before death from respiratory insufficiency occurs. We believe that in the present state of knowledge local wide excision is the only acceptable treatment.

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References


