Phaeochromocytoma associated with adult respiratory distress syndrome

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The presence of a phaeochromocytoma has been associated with many clinical presentations. We report a patient who presented with adult respiratory distress syndrome, an association that we believe has not been described before.

Case report

A 19 year old man was admitted as an emergency to the surgical department with a four month history of episodic abdominal pain and vomiting. On the night of admission he had been vomiting "continuously" for four hours. A recent barium meal had been reported as normal. On examination he was ill and tender in the epigastrium; widespread crackles were audible in both lung fields. A chest radiograph showed a few rounded shadows in both lungs suggestive of bronchopneumonia. His full blood count showed a leucocytosis of $30 \times 10^9/l$ but was otherwise normal. Intravenous antibiotic treatment with cefuroxime was started.

The next day he had deteriorated. He was appreciably tachypnoeic, with a respiratory rate of 25/min, and was clinically centrally cyanosed. The chest radiograph showed extensive homogeneous shadowing throughout both lung fields and when he was breathing spontaneously wearing a high concentration oxygen mask (MC mask at $10l \text{ min}^{-1}$) his arterial oxygen tension was 51 mm Hg (6.8 kPa), carbon dioxide tension 38.7 mm Hg (5.16 kPa), and pH 7.24. He was transferred to the intensive care unit and intermittent positive pressure ventilation was started. Despite positive end expiratory pressure of 10 cm H_2O there was no improvement (fig 1). Pulmonary artery catheterisation indicated a pulmonary capillary wedge pressure of 5 mm Hg and a mean pulmonary artery pressure of 35 mm Hg. A diagnosis of adult respiratory distress syndrome was made.

The aetiology of this remained obscure. All microbiological cultures and viral antibody titres were negative and plasma lipase and amylase concentrations normal, and there was no suggestion of aspiration while he had been vomiting. A notable feature during ventilation was the considerable lability of his blood pressure, which varied from 220 mm Hg systolic to 90 mm Hg systolic over 10-15 minute intervals. Because of this the 24 hour urinary metanephrine output was measured. This was raised at 30.7 (normal range <5.5) $\mu\text{mol}/24$ hours; but, in view of his overall stressed state, no firm conclusion that this might represent a phaeochromocytoma could be reached.

Two days after admission disseminated intravascular coagulation developed, with peripheral gangrene and

Fig 1 Chest radiograph on the second day of hospital admission, showing diffuse homogeneous shadowing throughout both lung fields.

Fig 2 Section of the adrenal tumour showing large cells with abundant granular cytoplasm and pleomorphic nuclei typical of phaeochromocytoma. (Haematoxylin and eosin.)

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thrombocytopenia. The platelet count was 14 (normal > 150) $\times 10^9/l$, fibrin degradation products were 60 (normal < 10) mg/ml, and the prothrombin time was 17 (normal ≤ 14) seconds. Later, because of the preceding history of epigastric pain, an ultrasound examination was performed to exclude an underlying abdominal lesion. This suggested the presence of a mass 3×4 cm in diameter related to the tail of the pancreas containing fluid. We considered that this could have been a pancreatic pseudocyst but, because of his overall condition, after appropriate consultation surgical intervention was rejected. Despite supportive treatment he died one week later from multiorgan failure.

At necropsy the lungs were heavy, deeply congested, and airless. Histological examination showed severe diffuse alveolar damage with extensive hyaline membrane formation and intravascular fibrin thrombi. Early bronchopneumonia was also present. There was a moderate degree of left ventricular hypertrophy (total heart weight 405 g, left ventricular weight 221 g) with thrombus in the right atrium. The liver and kidneys showed evidence of severe disseminated intravascular coagulation. The left adrenal gland contained a haemorrhagic encapsulated nodule, 4.5 cm in diameter, which on histological examination proved to be a pheochromocytoma that had undergone almost total haemorrhagic necrosis (fig 2).

Discussion

We have described a young man with a pheochromocytoma who developed adult respiratory distress syndrome. Despite the history of four months' symptoms, we believe that the development of adult respiratory distress syndrome was causally related to the pheochromocytoma and suspect that tumour necrosis triggered the response. Possibly this was an incidental finding, and certainly the association of adult respiratory distress syndrome, "viral infection," and disseminated intravascular coagulation has been described. Although initially the "working diagnosis" for this patient was pneumonia, microbiological investigation failed to find any evidence of a viral aetiology. Furthermore, we think it unlikely that the disseminated intravascular coagulation could be entirely responsible for the total necrosis of the abnormal adrenal gland and suggest that this came first.

There is other circumstantial evidence to suggest that pheochromocytoma and adult respiratory distress syndrome may be causally related. Pheochromocytomas have been reported to cause cardiogenic pulmonary oedema and sudden death. Melicow,¹ however, in a review of 100 cases of pheochromocytoma (1926–1976) described four patients who died with "bronchopneumonia" shortly after admission. Without ventilation and intensive supportive measures our patient would have presented a similar picture. More recently Jones and Durning² described a patient who presented with acute abdominal pain, a left sided abdominal mass, and hypertension. Soon after admission the patient died from "pulmonary oedema" and necropsy revealed a 22 cm diameter haemorrhagic pheochromocytoma. Commenting on this, Bullimore and Miloszewski³ suggest that the severe systemic vasoconstriction produces a fluid shift to the pulmonary circulation. A similar mechanism can be seen in pulmonary oedema associated with severe head injuries, where massive α adrenergic discharge is thought to be the mediator^{4,5}; and this effect has been blocked experimentally in animals by adrenergic blocking agents.⁶ We would therefore suggest that pheochromocytoma should be added to the list of conditions associated with the adult respiratory distress syndrome and that the role of the adrenal gland in the aetiology of this condition needs further evaluation.

References

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