a sodium load. Experimental studies support the view that an increased oncotic pressure in the peritubular capillaries, consequent on polycythemia, facilitates increased sodium reabsorption (Schrier and Earley, 1970).

The decreased glomerular filtration rate in patients with cyanotic congenital heart disease and an increasing awareness of the limited homeostatic capabilities and decreased renal function in infants emphasize the importance of careful monitoring of renal function in these patients, especially in the early postoperative period.

Summary

Children with cyanotic congenital heart disease had a decreased glomerular filtration rate (71.8 ± 18.9 ml/min per 1.73 m^2) measured by endogenous creatinine clearances, compared with children who had had complete corrective surgery, children with noncyanotic heart disease, and normal children. There was a significant correlation between low glomerular filtration rate and haematocrit values above 50%. Daily urinary sodium excretion was reduced in the cyanotic patients.

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Pulmonary eosinophilic granuloma in a child

While the various forms of histiocytosis X are not particularly uncommon in childhood, it appears to be excessively rare for the disease to be localized to the lungs, most patients with isolated lung disease being adults (Lewis, 1964). We report a case of isolated pulmonary eosinophilic granuloma in 3-year-old child, who presented in the characteristic fashion and who failed to respond to radiotherapy.

Case report

A female born in 1971 of Asian parents had been a healthy child, though small, until the age of 3 years 5 months when she became unwell with nausea and abdominal pain. The pain later shifted to her chest, and by next morning was more severe, associated with shortness of breath. She was referred to hospital where a right pneumothorax was found. A chest drain was inserted with continuous underwater suction. The lung re-expanded but the drain continued to bubble air for a further 15 days. It became apparent on chest x-ray that she had a honeycomb abnormality of both lungs, and she was transferred to Hammersmith Hospital for further investigation.

On examination she appeared unwell, with a chest drain *in situ*. She was small, height being 91 cm (3–10th centile) and weight 8.3 kg (3rd centile). Respiratory rate was 24/min, with no cyanosis, but crepitations were audible at the right lung base and there was clubbing of fingers and toes. There was no significant lymphadenopathy, and no enlargement of liver or spleen. All other systems were normal.

Chest x-ray showed a honeycomb appearance throughout both lungs. Her arterial Po_2 in air was 84 mmHg, pH 7.39, and Pco_2 31 mmHg. An adequate sample in oxygen was not obtained. Blood count showed Hb 12.7 g/dl, WBC $12\,800/\text{mm}^3$ ($12.8 \times 10^9/\text{l}$) (neutrophils 92%, lymphocytes 2%, monocytes 6%), erythrocyte sedimentation rate 5 mm/h, Mantoux 1:1000 was negative. A gamma scan of the lungs using radioactive N_2 (Ronchetti *et al.*, 1975) showed poor ventilation of both lungs, right worse than left. Sweat electrolytes were normal and there were no clinical features to suggest epiloia. There was no radiological evidence of bony granulomata as judged by her skeletal survey and skull x-rays, which were normal. There was no clinical or laboratory evidence of diabetes insipidus (urine osmolality 784 Osm/kg after overnight starvation).

The pleural drain was removed, but 2 days later the pneumothorax recurred. The opportunity was taken to treat this surgically, and at the same time to take a lung biopsy. At thoracotomy (Mr. M. P. Singh) the cystic abnormality of the lungs was confirmed. Biopsies were taken from lung and rib and 50% dextrose solution instilled to induce pleural adhesion. She made a satisfactory recovery and the pneumothorax did not recur.

FIG. 1.—A low power micrograph of the lung showing the characteristic subpleural, perivascular, and peribronchiolar distribution of the infiltration. Rupture of the pleura is seen top centre. (Haematoxylin and eosin. $\times 27$.)

FIG. 2.—Electron micrograph of part of a histiocyte demonstrating rod-shaped X-bodies. ($\times 39\,000$.)

The lung biopsy (Fig. 1) showed characteristic, widespread, subpleural, perivascular, and peribronchiolar granulomata. These were composed predominantly of histiocytes, some lymphocytes, plasma cells, polymorphs, and many eosinophils. In at least one area the infiltrate had penetrated the pleura and had invaded and destroyed the wall of a bronchiole. Medium-sized and small cysts were present in the lung parenchyma and fibrosis had occurred in some areas. This established the diagnosis as eosinophilic granuloma. Rod-shaped

structures (X bodies) were detected by electron microscopy in some histiocytes (Fig. 2). These structures were more obvious in histiocytes of lung parenchyma with established fibrosis. They were lined externally by a double membrane with a linear central density and a few of them showed the terminal tennis-racket-shaped dilatation. Rib biopsy and marrow aspirate were normal, with no evidence of histiocytic infiltration.

Her respiratory rate was persistently raised (40–60/min) and arterial PO_2 in air fell to 60 mmHg. Her lung disease appeared to be progressing and she was unwell. 2 weeks after operation she underwent a course of radiotherapy to the whole right lung, receiving a total of 900 rads over 10 doses. During this period her general well-being improved and her respiratory rate fell to 30/min. However, a repeat of the gamma scan of her lungs showed no change in the ventilation of the lungs, nor any radiological improvement. She then had a similar course of radiotherapy to the left lung. Her general condition continued to improve though she did not gain weight despite an improved appetite. She completed treatment a month later and went home. Her respiratory rate was 24–28/min.

Four weeks later she was cheerful, but had a poor appetite and had not gained weight. The finger clubbing was still present, and there were râles audible over the right lung. Further studies of lung function and T lymphocytes were planned, but before readmission she went on a visit to India where she collapsed suddenly and died before medical assistance could be obtained. Necropsy was not performed.

Discussion

We know of only 2 other children reported with pulmonary eosinophilic granuloma. One was a 3½-year-old Caucasian girl who appeared to have isolated lung disease, which proved fatal. Necropsy showed involvement of thymus, thyroid, bone-marrow, and liver in addition to the lungs (Weber, Margolin, and Nielsen, 1969). The other was a female aged 9 months whose disease was also fatal at the age of 1 year, despite treatment. Necropsy established that the disease was confined to the lungs (Aftimos, Nassar, and Najjar, 1974). Most other patients in published reports with isolated lung disease are young adults, but a wide age range may be affected (Lewis, 1964). Our patient had a typical mode of presentation (pneumothorax) as first noted by Roland, Merdinger, and Froeb (1964). Clubbing has not been found as a general rule, though 3 of the 75 cases reviewed by Lewis (1964) did show this feature.

The tubular particles seen in electron micrographs of histiocytes have been reported in cases of adult pulmonary and extrapulmonary eosinophilic granuloma as well as other members of the histiocytosis-X group (Basset and Turiaf, 1965; Basset and Nezelof, 1966; Brody *et al.*, 1974). A cyto-

plasmic membranous complex was reported in 2-5% of the histiocytes characteristic of all the clinical variants of histiocytosis-X (Bassett, Escaig, and Le Crom, 1972). We were not able to show this complex in spite of an exhaustive search.

The natural history and the prognosis of pulmonary eosinophilic granuloma is very variable and unpredictable. Some patients recover, yet others may die within months. One patient reported by Roland *et al.* (1964) developed successively mandibular granulomata, diabetes insipidus, and multiple skeletal lesions, all starting 2 years after discovery of the lung disease. Spontaneous remissions have been common, and response to treatment is capricious and unproven (Hoffman, Cohn, and Gaensler, 1962). Steroids, ACTH, and radiotherapy have all been used, but there is no definite evidence of consistent or reliable response. It seems reasonable to use these therapies when the natural course appears to be one of deterioration, as in the child reported here. There was clinical evidence of a response to radiotherapy in our patient, but in retrospect this seems to have been little more than a temporary improvement since there was no objective evidence of improvement, as judged by chest x-rays and gamma scan. It is probable that she died of a recurrence of pneumothorax.

Summary

The occurrence of pulmonary eosinophilic granuloma in a 3-year-old child is described. She presented with a pneumothorax and typical radiological changes and the diagnosis was confirmed by lung biopsy. There was no objective evidence of improvement after radiotherapy when lung function was assessed by gamma scans. She died suddenly while abroad.

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