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Histiocytoma of the Lung: Report of a Case

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Summary

Abdurrahman, M. B. and Fasawe, T. O. (1979). *Nigerian Journal of Paediatrics*, **6(2). 44. Histiocytoma of the Lung: Report of a case.** A case of histiocytoma of the lung in a four-year old boy is reported. He presented with cardiac failure and features of chronic pulmonary disease and died during operation. Macroscopically, the lesion appeared benign, but histological appearances revealed low grade malignancy. There is controversy about the nature of histiocytic tumours.

Primary tumours of the respiratory tract are rare in children, and histiocytomas are even rarer. Histiocytomas are soft tissue tumours with variable and polymorphic histologic appearance, consisting of fibroblasts, histiocytes and foam cells with a high lipid content which gives the tumours their xanthomatous appearance. Kauffman and Stout (1961) presented a detailed review of histiocytic tumours in children, indicating that they occur most commonly in skin, tendons, joints, and soft tissues, but rarely in the lungs. About 50 cases of intrathoracic histiocytoma have been reported in world literature, and less than 20 of these were in children (Pearl, 1972; Armstrong, Elston, and Sanderson, 1975).

The following is a case of histiocytoma of the lung in a four-year old Nigerian. It is reported to illustrate some diagnostic difficulties as well as to serve as a reminder that opacities in the lung are not always synonymous with pneumonia.

Case Report

T.M., a four-year old boy, was admitted to the Ahmadu Bello University Teaching Hospital, Kaduna, complaining of cough and dyspnoea for two weeks, fever and swelling of the feet for three days prior to admission. The cough was said to be productive of mucoid, blood-stained sputum. Initially, the dyspnoea was on exertion, but this progressed to orthopnoea after a week. Past medical history was unremarkable. In particular, there was no history of, or contact with, any case of chronic cough.

Physical examination showed an ill-looking, orthopnoeic child with mild pallor and a temperature of 38^{0}C. There was moderate degree of pitting pedal and sacral oedema, and moderate digital clubbing. There was diminished movement, stony dullness, and diminished air entry over the whole of the right side of the chest. The

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trachea was deviated to the right. There was tachycardia (heart rate, 150-160/minute); the apex beat was located in the sixth left intercostal space in the anterior axillary line; there was no cardiac murmur. Jugular venous pressure was raised to near the angle of the jaw. There was ascites, as well as hepatosplenomegaly of 8 cm and 9.5 cm respectively.

The provisional diagnoses were chronic pulmonary disease, cor pulmonale and right-sided cardiac failure. The possible causes considered included tuberculosis, infected congenital lung cyst, bronchiectasis and schistosomiasis.

Investigations revealed haemoglobin 8.0G/dl; leucocytes, 12.2x 10^9/L (neutrophils, 68 per cent, mainly unsegmented; lymphocytes, 24 per cent and monocytes, 8 per cent); reticulocyte, 4.6 per cent; platelet 115.0 x 10^9/L; haemoglobin genotype AA; slightly elevated serum urea of 12 mM/L (normal: 3.5–10); 1+ albuminuria and normal urine microscopy. Serum electrolytes were normal. Stool specimens examined on several occasions showed no ova or parasites, and rectal snip contained no schistosome ova or tubercles. Sputum and gastric washings on three different occasions and cultures revealed no acid-fast bacilli. Diagnostic pleural aspiration carried out on two occasions yielded no fluid. Chest radiograph showed uniform opacity occupying most of the right hemithorax, with a sharply-defined lower border; there was also a small pleural effusion. The Mantoux tuberculin (5 T.U.) reaction was 7 mm.

Initial treatment consisted of digitalization and administration of frusemide, and intramuscular penicillin and cloxacillin. Although signs of cardiac failure disappeared within 72 hours of starting treatment, the respiratory signs and hepatosplenomegaly persisted. After ten days, the penicillin and cloxacillin were discontinued, and the patient was started on triple anti-tuberculous therapy. Because of deterioration in his respiratory symptoms and signs, it was decided that the patient should undergo thoracotomy. At the time

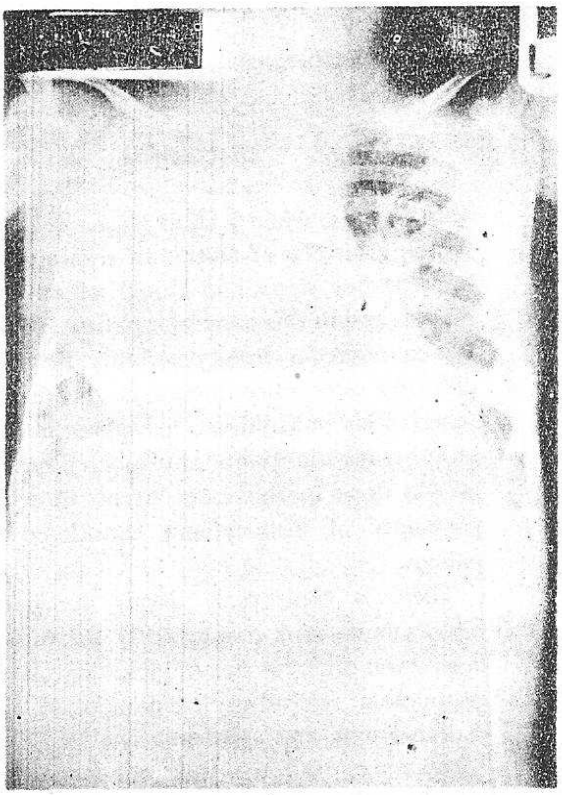

Fig. Chest radiograph of a patient with histiocytoma, showing uniform opacity occupying most of the right hemithorax, and a small pleural effusion.

of surgery, the patient was still on anti-tuberculous therapy and maintenance dose of digoxin.

Twenty days after admission, the patient underwent thoracotomy. The finding was an encapsulated mass in the posterior mediastinum arising from the upper lobe of the right lung. There were no adhesions and no visible infiltrations of the surrounding tissues. The mass was removed without difficulty, but immediately after removal, the patient had cardiac arrest and died.

The tumour consisted of a yellow soft tissue, measuring 10 x 10 x 5 cm. in size, and weighing 250 gm. The cut surface had a pale yellow appearance, with areas of necrosis. The histology revealed a tumour composed partly of elongated cells with pale cytoplasm and pale, plump oval nucleus, and partly of larger polyhedral cells with pale granular cytoplasm and vesicular nucleus

without prominent nucleolus. A number of bizarre multinucleated giant cells as well as mitotic figures were also present. These findings were considered to be compatible with histiocytoma of low grade malignancy.

Discussion

In Nigeria, when a child presents with a long-standing history of fever and respiratory symptoms and/or signs, one thinks of an infectious disease, and in this particular patient, tuberculosis was considered a strong possibility. Because of the difficulty often encountered in the diagnosis of tuberculosis in children, one sometimes resorts to a therapeutic trial of anti-tuberculous therapy as was done in this case. At no time was the possibility of histiocytoma considered in our patient.

There is controversy about the nature of histiocytoma, and consequently the tumour has been given different names including plasma cell granuloma, sclerosing haemangioma, vascular endothelioma, and xanthomatous pseudotumour (Armstrong, Elston, and Sanderson, 1975). Histologically, there are three basic patterns namely: predominant fibrous tissue (fibrome), predominant vascular element with variable degree of reactive fibrous obliteration of the vessels (sclerosing angioma), and predominant histiocytic element (histiocytoma). We believe that all these are tumours of histiocytes, with the histiocyte exhibiting its pluripotential nature to varying degrees in various lesions.

Histiocytoma appears to be a true neoplasm. Many of the reported cases are benign in behaviour (Dubilier, Bryant, and Danielson, 1968), but some are definitely malignant (Kyriakos and Kempson, 1976; Blitzer, Lawson, and Biller, 1977). O'Brien and Stout (1964) have estimated that about 1 per cent of this tumour are malignant. It is only on long-term follow-up that the true nature of the lesion becomes evident, because assessment of its malignancy on the basis of histological appearance alone can be difficult. For example, Hakimi *et al* (1975) have reported a case which was histologically malignant but clinically benign after over two years follow-up.

Respiratory tract histiocytoma may present as early as in the neonatal period (Searer *et al.*, 1973), or as late as in the sixth decade of life (Dubilier, Bryant, and Danielson, 1968). It may be an incidental radiologic finding in an asymptomatic patient (Dubilier, Bryant, and Danielson, 1968; Grossman *et al.*, 1973), or may present with respiratory distress (Shearer *et al.*, 1973), cough with wheezing and haemoptysis (Pearl, 1972; Hakim *et al.*, 1975; Armstrong, Elston. and Sanderson, 1975).

Our patient presented with right-sided cardiac failure and features of chronic pulmonary disease associated with haemoptysis. At surgery, the lesion appeared benign, but on histology there was evidence of low grade malignancy. On the basis of available data on intrathoracic histiocytoma, the treatment of choice is wide local excision and long term follow-up.

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