

Case Report

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Neuroblastoma — An Unusual Presentation

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SUMMARY

A case of Neuroblastoma with an unusual presentation with persistent haemothorax has been reported and discussed.

INTRODUCTION

Neuroblastoma is relatively a common malignant solid tumour in childhood. Half of the cases occur in children two years of age and younger and three quarters before 4 years of age. Neuroblastoma develops from neural crest tissue. The first clinical manifestation is often due to metastatic lesions

which mimic other conditions so well that the diagnosis may be difficult.

CASE REPORT

M.A., 2½ years female, Asian girl was admitted to the University Teaching Hospital on 22nd January 1979 with a history of cough 5 days prior to admission. Two weeks previously she had a limp and pain in the right leg with fever which subsided with cotrimoxazole. X-ray of the right hip was normal. However, she continued to have low grade fever. There was a distant family history of tuberculosis but the child had not been in contact with the affected member.

On examination, she was dyspnoeic, ill-looking and pale with a respiratory rate of 58 per minute. Chest move-

ments and air entry were diminished on the right side of the chest and the percussion note on the right side was stony dull. No abnormality was detected on cardiovascular and abdominal examination. The X-ray of chest showed a homogenous opacity filling the whole of the right side of the chest (Fig. 1). A diagnostic pleural-tap showed uniformly haemorrhagic fluid. A provisional diagnosis of malignancy of pleura (Mesothelioma) was therefore made and she was investigated on those lines.

On admission, haemoglobin was 8.6gm%, total leucocytic count 6,400/cu.mm with lymphocytosis of 72% and normal platelet count and E.S.R. Prothombin Time (PT) and Partial Thromboplastine Time (PTT) were normal. The pleural fluid proteins were 6.2mg% with concentration of albumin 3.2gm%. Microscopic examination revealed 110 leucocytes/cu.mm with numerous red cells and there was no growth on culture. No definite malignant cells could be seen on cytological examination and there were no A.F.B. or amoebae in the pleural fluid. The pleural biopsy did not show any specific pathology while the bone marrow was normal. Tuberculin test was negative. She was given antibiotics and a blood transfusion. But repeated pleural-tap showed haemorrhagic fluid, filling-up quickly after each aspiration (Fig. 2). Since there was no improvement and the pleural biopsy did not suggest malignancy, antitubercular drugs were given on the basis of the family history of tuberculosis.

A week later hepatomegaly of 4cms was noted, increasing up to 12cms. She developed maculopapular itchy rash on the right side of abdomen extending to the right side of chest. Toxicity of Thiacetazone was suspected and Thiazina was stopped but Isoniazid was substituted in its place. Antihistamines were also given. The rash subsided within 3 days. The liver function tests and blood electrolytes were normal. The repeat PT, PTT and blood counts were normal. Liver scan showed a downward displacement of the liver with no filling defect. The repeat X-ray chest showed again the homogenous opacity on the right side with no erosion of ribs.

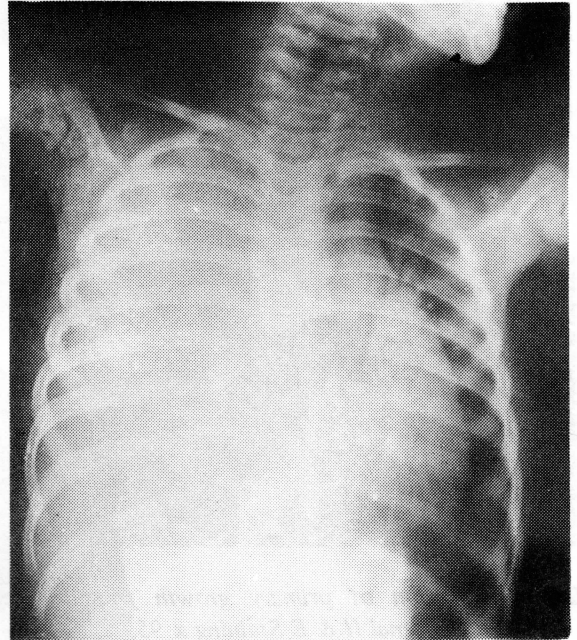
The reduced air entry and dull percussion note on the right side of the chest remained unchanged. On 15th February, she developed fine petechial rash on the lower limbs with an haemangioma on the right sole and behind left ear. Hess's fragility test was positive. Her general condition deteriorated and she died on 17th February, 1979.

At autopsy, a whitish, firm, nodular mass, measuring about 6 x 6 x 4cm was found involving right adrenal gland, adherent to the right kidney. It was ill-defined and was extending superiorly into the right paravertebral region of the posterior mediastinum invading the posterior border of right lung, and inferiorly into the pelvic cavity. On microscopic examination, the growth proved to be neuroblastoma (Fig. 3 & 4), with a diffuse infiltration in the liver (Fig. 5).

DISCUSSION

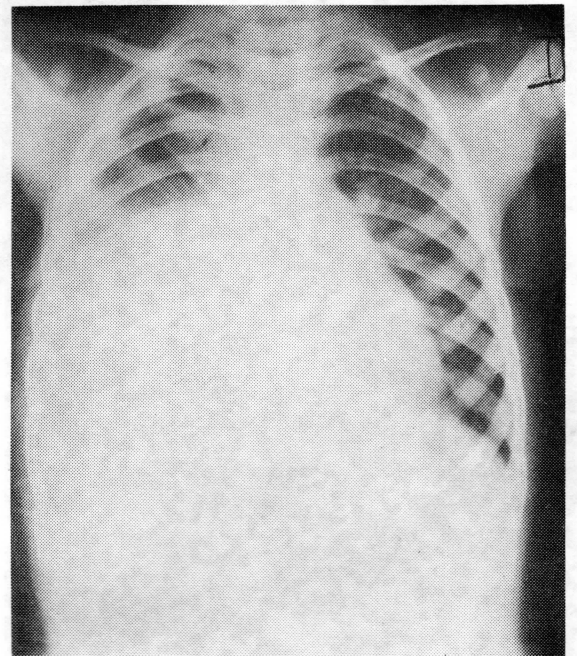
Metastasis is often the first sign of neuroblastoma. In Allen's (1977) series of 213 cases, the diagnosis was made through symptoms produced by metastases. About two third of neuroblastoma arise from the adrenal or the upper abdominal ganglia and present, initially, as a painless mass in the flank. The

Fig. I



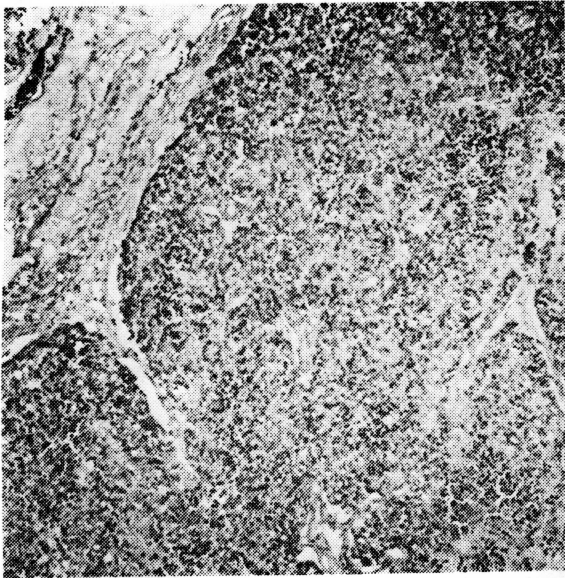
Case II: Skiagram showing homogenous opacity on right side of the chest before pleural aspiration.

Fig. II



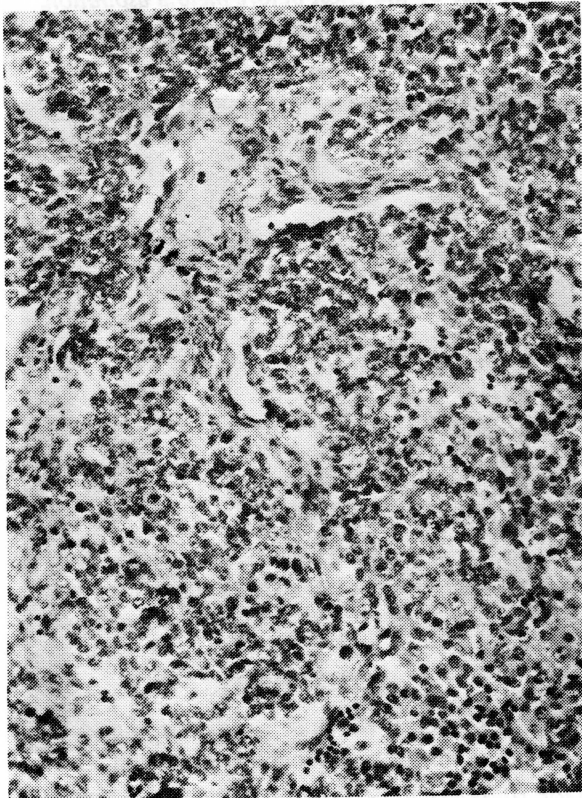
Case II: Skiagram persistence of opacity on right side of the chest after complete pleural aspiration on right side.

Fig. III



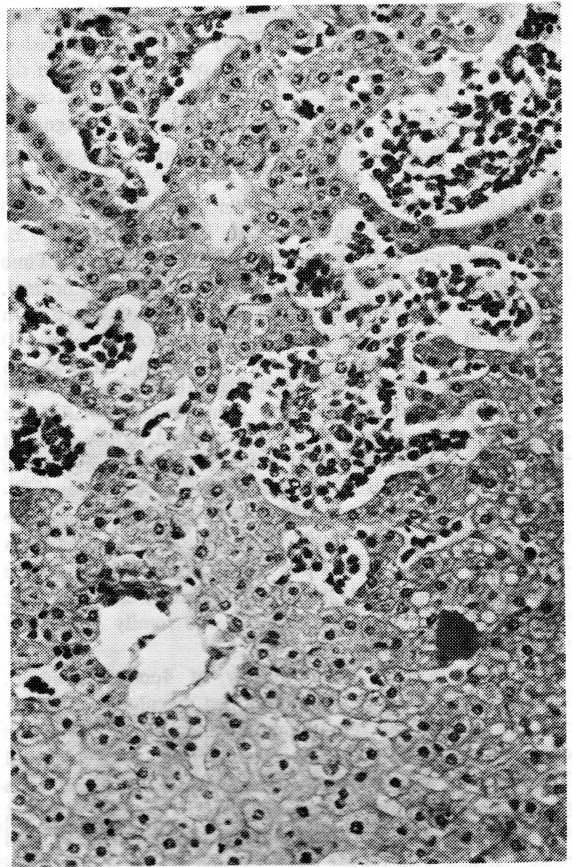
Case II: Section of primary growth from right adrenal H & E Staining x 95.

Fig. IV



Case II: High-power view of field shown in Fig. V with rosette formation. H & E Staining x 240.

Fig. V



Case II: Section of liver showing diffuse metastasis of neuroblastoma. H & E Staining x 240.

present case had no palpable abdominal mass inspite of involvement of right adrenal seen on necropsy (Fig. 3 & 4). The tumour also commonly occurs in the posterior mediastinum.

Mediastinal compression is typical in type I neuroblastoma, where a large rapidly growing necrotic semifluid undifferentiated tumour often extending as a mass into one pleural cavity and attached on its medial aspect to the mediastinum. Progressive dyspnoea or tachypnoea, a brassy cough, dysphagia and congestion of face and neck are the common features of such tumour (Peter and Peter 1976). In spite of pleural metastasis, intrapulmonary metastasis are extremely rare (William 1972; Peter and Peter 1976). Hemothorax as a feature of thoracic neuroblastoma has not been described. The present reported case had hemothorax with malignant metastasis in the right pleura invading the pulmonary parenchyma but throughout the patient's life, neuroblastoma was not considered in the differential

diagnosis of hemothorax. The literature, however reveals only one case of pleural effusion, with extensive mediastinal involvement and erosion of the ribs due to a mediastinal neuroblastoma (Joseph 1968).

Hepatic metastasis is one of the common features of neuroblastoma arising from right adrenal, but it occurs in patients less than two years of age and takes one of the two different forms (i) spherical or irregular masses causing irregular knobby enlargement of liver (ii) diffuse infiltration (Peter and Peter 1976). The present case had hepatomegaly of 4cms in size which went up to 12cms but it was a smooth, firm, liver. Primary malignancy of liver was also suspected but the liver scan did not show any abnormality and liver biopsy was refused by the parents. The microscopic examination of the necropsy specimen showed diffused metastasis of liver (Fig. 5) which is uncommon at this age. Diffuse metastases of the liver, which is also called Pepper's syndrome, is found in infancy (Williams 1972; John 1973; Peter and Peter 1976). Hepatic metastases that occur in older age group tend to be nodular (Williams 1972).

Along with hepatomegaly the present case also had unilateral rashes of the right side of the body involving chest and abdomen which was thought to be due to the Thiacetazone toxicity. Later on, when the literature was reviewed, William (1972) has described that right sided thoracic neuroblastoma may present with purpuric unilateral rashes over the chest and abdomen due to the effect of sympathetic

nervous system disturbance. However skin and subcutaneous nodular deposits along with hepatomegaly during infancy is one of the commonest features observed by all (Williams 1972; John 1973; Peter and Peter 1976) and according to Evans (1971) classification it belongs to stage IV and has good prognosis.

ACKNOWLEDGEMENTS

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REFERENCES

1. Allen, D.S. (1977), *Med Clin. North Am.* 61, 1053.
2. Evans, A.E. and D'Angio, G.T. (1977) *Cancer*. 27, 374.
3. Joseph, F. (1968) *Ann. Surg.* 167, 132.
4. John, O. Grodden (1973) *Cancer in children - Holt, Rinehart and Winsten of Canada, Limited, Toronto, Canada*, P. 17.
5. Peter, G.J. and Peter, E.C. (1976) *Tumour of infancy and childhood Blackwell Scientific publication*, P. 46¼.
6. Williams, I.G. (1972) *Tumour of childhood. A clinical treatise - William Heinemann Med. Books Limited, London*, P. 60.