

ADULT NEUROBLASTOMA: A CASE REPORT

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Summary

Neuroblastoma is an uncommon malignancy in adult life as compared with childhood. A case of Stage IV neuroblastoma in a 24-year-old Chinese man seen at the University Hospital, Kuala Lumpur is reported. The diagnosis was based on characteristic histological features seen in an excised lymph node with metastatic involvement and an elevated urinary metanephrine level. The finding of a suprarenal mass on computerised axial tomographic scan supported an adrenal origin. Other diagnostic aids which help to distinguish this tumour, especially the less differentiated forms, from other "small, dark, round cell tumours" are discussed.

Keywords: Neuroblastoma, **small** cell tumour.

INTRODUCTION

Although neuroblastoma is the most common extracranial solid malignancy in childhood, it is rarely seen in adults. We report its occurrence in a 24-year-old Chinese male to highlight the importance of considering this tumour in the differential diagnosis of "small, dark, round cell tumours" in non-paediatric patients.

CASE REPORT

A 24-year-old male Chinese was admitted to the University Hospital, Kuala Lumpur in 1986 with complaints of low grade fever and bone pains in the left chest and right thigh for a duration of one month prior to admission. On physical examination he was found to be pale. An epigastric mass, about 8 x 6 cm, was palpable. He also had generalised lymphadenopathy with lymph nodes ranging from 0.5 to 3 cm in size. No hepatosplenomegaly or localised bone tenderness was detected.

Investigations revealed a haemoglobin level of 9.8 g/dl. There was evidence of thrombocytopenia (89×10^9) but the leucocyte count was within normal range (4.9×10^9). A leucoerythroblastic picture was seen on examination of the peripheral blood film. Liver function tests demonstrated raised serum enzyme levels (aspartate transaminase of 159 IU/L; alanine transaminase of 62 IU/l and alkaline phosphatase of 227 IU/l). The serum bilirubin was 12 mmol/l while the serum albumin was 37 g/l. Blood urea, serum electrolytes, calcium, phosphate and uric acid levels were normal. A computerised axial tomographic examination of the abdomen

showed a large, left, suprarenal, retroperitoneal mass which measured 10 x 10 cm. This mass was medial to the spleen and extended to the inferior border of the left lobe of the liver. The paraaortic lymph nodes were not enlarged. X-rays of the chest and right femur were normal. The urinary metanephrine level was elevated to 91 mmol/24hrs (normal: 0-7 mmol/l). Bone marrow examination and trephine biopsy showed an infiltrate of small, dark, round cells with the presence of an occasional rosette.

A cervical lymph node biopsy was performed. The excised lymph node measured 3 x 2 cm. The cut surface showed areas of necrosis and haemorrhage. Histological examination revealed features characteristic of neuroblastoma. The lymph node was infiltrated by small cells which had slightly pleomorphic, round, hyperchromatic nuclei and scanty cytoplasm set against a faint fibrillary background. Homer-Wright rosettes with central fibrillary polarisation were also noted (Fig. 1). No ganglionic maturation was observed.

The patient was diagnosed to have Stage IV neuroblastoma based on the system proposed by Evans *et al.*¹ and was commenced on combination chemotherapy. However, he continued to deteriorate clinically and developed a palpable hepatomegaly. Ultrasonography confirmed the enlarged liver which was thought to be the result of further tumour infiltration. He succumbed 4 months from the date of initial diagnosis. A post-mortem examination was not consented to.

DISCUSSION

Neuroblastoma is essentially a tumour of childhood, usually occurring before the age of 4 years. There are only a few isolated reports of its occurrence in adults.²⁻⁵

The diagnosis of neuroblastoma in this case was not particularly difficult. The clinical presentation of prolonged fever, bone pains abdominal mass and pallor was very much like that in childhood cases previously reported from this hospital.⁶ The characteristic histology seen on biopsy of a metastatic lymph node in the presence of a raised urinary metanephrine level confirmed the diagnosis. In addition, the finding of a suprarenal mass on computerised axial tomography supported an adrenal origin.

In the adult patient the diagnosis of neuroblastoma can sometimes elude the unwary. Although a high urinary metanephrine level, when present, aids in the diagnosis as well as in predicting relapse, it is not commonly elevated in the adult variant of this tumour.² The histological features on tissue sections frequently defeat attempts to differentiate it from other small, dark, round cell tumours on light microscopical examination. In the adult, tumours which can be confused with a neuroblastoma include

lymphomas, undifferentiated small cell carcinomas and rhabdomyosarcomas. If the diagnosis is in doubt, the pathologist can resort to further investigatory techniques to help in confirming or refuting the diagnosis. In contrast to the other small, round cell tumours, neuroblastoma produces catecholamines which can be demonstrated in tissue sections or touch preparations by formaldehyde-induced fluorescence.⁷ Immunohistochemistry has its limitations in differentiating neuroblastoma from undifferentiated small cell carcinomas due to the expression of similar antigens (neuron specific enolase, S-100 protein and bombesin) by both these tumours.⁸ Nevertheless immunohistochemistry is useful in differentiating neuroblastomas from lymphomas and rhabdomyosarcomas. The presence of dendritic processes, dense core granules and desmosomes on electron microscopy also readily distinguishes the neuroblastoma.⁵ Tissue culture shows characteristic outgrowths of neurites from even the most poorly differentiated neuroblastoma cells.⁷

As a final note, the diagnosis of neuroblastoma in an adult patient rests on awareness of this condition.

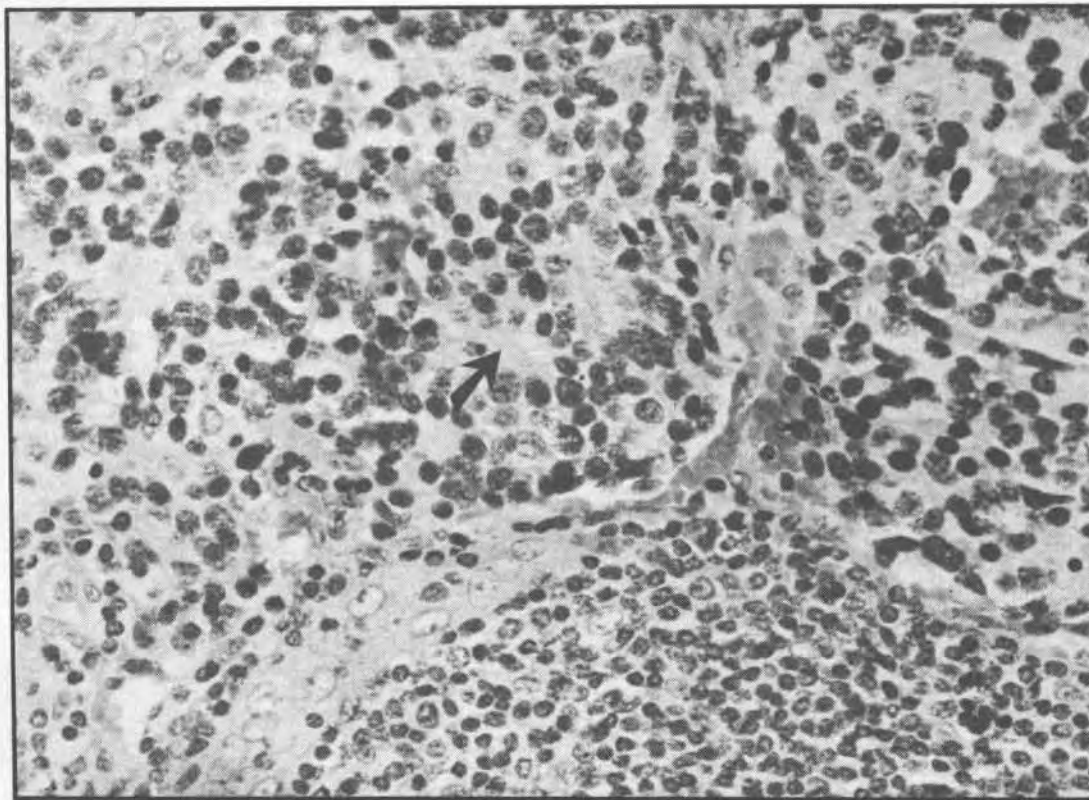


FIG. 1: Lymph node biopsy showing small, round tumour cells set against a fibrillary matrix with Homer-Wright rosettes (arrow). Residual lymphoid tissue is shown on the lower part. (H & E x 200).

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