PRIMARY NEONATAL INTRACRANIAL NEUROBLASTOMA CASE REPORT

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SUMMARY :

Primary intracerebral neuroblastoma is an uncommon neoplasm. It is difficult to make a differential diagnosis from other intracranial neoplasm preoperatively or histologicaly because of its undifferentiated cellular features.

In this report a case of primary intracranial neuroblastoma in the neonatal period is presented and the literature reviewed.

KEY WORDS :

Neonatal intracranial tumours, Neuroblastoma.

INTRODUCTION

Intracranial tumours are not infrequent in childhood and adolescence (6,7) but during the first year of life are extremely rare (10,14,15,16,23,31,32,37). Different reports suggested that the incidence ranges between 1.4% and 8.5% of all pediatric tumours in infants (10,1,16,32).

The most frequent intracranial tumour of the neonatal period is teratoma (46.1 percent) (21,34). Approximately *6.5 percent of neoplasms are of neuroepithelial origin (17,20). After the neonatal period, teratomas are less common and most tumours are of neuroepithelial origin with a large proportion of astrocytomas (13,14,32).

The incidence of primary cerebral neuroblastoma in the neonatal period is unknown because of its rarity.

CASE REPORT

A one-month-old boy presenting with recurrent generalized convulsions and enlarging head was admitted to hospital. His mother was 28 years old and had 4 children. Her pregnancy had been normal.

Our patient was born in a State Hospital and his first check-up had been made there. Clinical findings had shown that he had a bigger head circumference than normal.

In the third week of life, first convulsive attack had been observed and he had been referred to hospital.

Examination. Physical and neurological examinations showed that all findings were consistent with an increased intracranial pressure syndrome. When the contrast enhancement computed tomography (CT) was performed, it showed a mass filling the right middle fossa spreading out to the anterior fossa with surrounding brain edema and shifting to the left side (Figure 1). Open right carotid angiography performed for localization of the mass and its blood supply, revealed severe vasospasm.

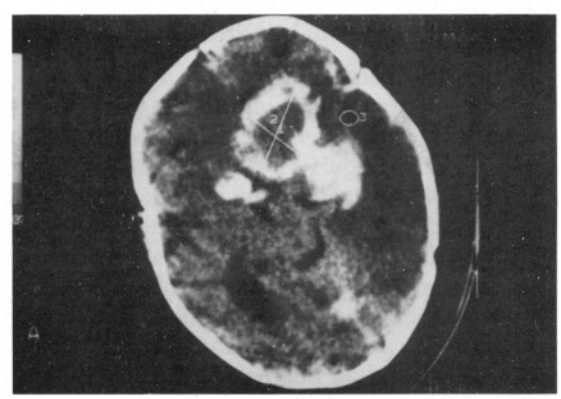

Figure 1 : Contrast enhancement CT scan showing a mass in the right middle fossa spreading out to the right anterior fossa with surrounding brain oedema. Necrosis also seen in the middle of the mass.

However, the other routine laboratory examinations including abdominal ultrasonography did not reveal any pathological findings (except mild homogeneous hepatomegaly).

Operation. A large right frontotemporoparietal craniotomy was performed the presumptive diagnosis being congenital intracranial neoplasm. Right temporal lobectomy showed that a hard, fragile,

reddish-white tumour with circumscribed organized haematoma had invaded the surrounding brain tissue and this was resected subtotally. The mass was adherent to the skull base.

Postoperative Course. There were no significant changes in the patient's condition postoperatively. A control CT scan two days after the operation showed minimal residual tumour and severe peripheral oedema (Figure 2). The general condition deteriorated and death occurred 5 days after the operation.

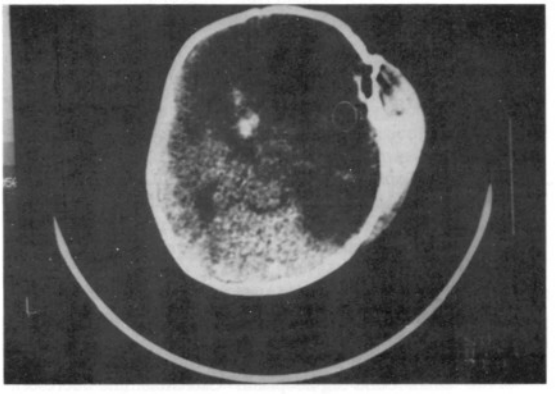

Figure 2 : Postoperative CT scan.

Pathological Examination. Cells which exhibited a neuroblastic nature were seen between the zones of haemorrhage and necrosis. The atypical cells had small, round and deeply staining nuclei and a dense chromatin network. Surrounding central nervous tissue was invaded by the tumour cells. There was no reticulin network or rosette formation (Figure 3).

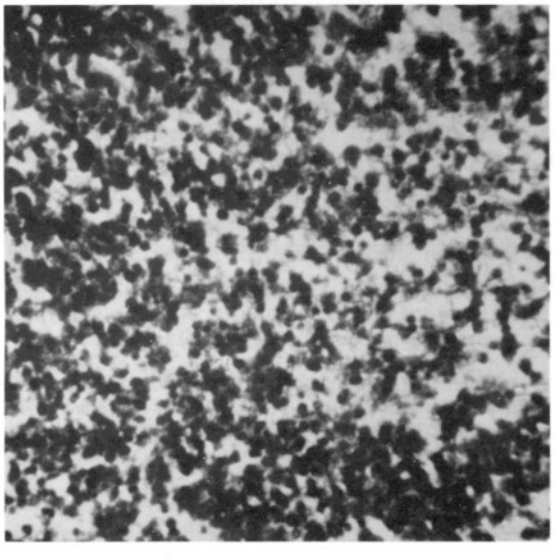

Figure 3 : The atypical cells have small, round and darkly staining nuclei and dense chromatin network. H.E.X250.

DISCUSSION

Primary cerebral neuroblastoma is an uncommon tumour in the neonatal period. Therefore, histopathological features are essential for differential diagnosis. The genesis and maturation of neurons from multipotential homogeneous cells occur in there stages. Stage 1 or the neurocytogenesis phase exhibits neural development with active cell division but cellular differentiation is not seen at this stage. The origin of neuroblast is performed in stage II. In stage III, the neuroblasts have potential for maturation but are unable to divide. Thereafter they become neurons (9,28).

In these circumstances, neuroblastoma emerges during the second stage of cytogenesis and shows a different maturation along one cell line toward neurons (8,12,19,35). In some cases, beginning of differentiation toward neurons has been observed (1). Maturation to adult cell forms is sometimes seen, but is more often lacking. Frequently these tumours are so difficult to recognize that some neoplasms which were originally described as medulloblastoma, ependymoma, undifferentiated primary sarcoma or poorly differentiated oligodendroglioma by some authors were indeed neuroblastomas (5,28).

The gross appearance of neuroblastoma is well defined even though crisply demarcated in its place. It is often lobulated with soft pearly-gray cut surfaces, usually with extensive areas of haemorrhage, necrosis and gelatinous cystic degeneration. Our case also presented extensive wide haemorrhage and necrosis with Islands of tumour cells in these areas. Secondary attachment to the dura is occasionally found (27). Microscopically, there are collections of small regular cells which contain round and darkly staining nuclei. There is little cytoplasm and cytoplasmic outlines are poorly defined. Necrotic areas are often present. Rosette formation is seen in about a quarter to a third of cases (30).

The histological features of primitive ectodermal tumours such as glial differentiation, high percentage of cellular indifferentiation, pleomorphism and less prominent connective tissue component can be difficult to confirm differential diagnosis using a light microscope, but ultrastructural analysis makes is successful (1,11,26).

Ultrastructural examinations will show, in addition, neurosecretory granules and synaptic endings (33). In vitro maturation of neuroblastoma has sometimes been used as a diagnostic aid (25).

Neuroblastoma may arise anywhere in the cerebral hemispheres but the left hemisphere is the slightly dominant location (2).

Thirty-five cases of primary cerebral neuroblastoma were presented by Horten and Rubinstein (12,29) and eleven cases were reported retrospectively by Berger and colleagues (2). These tumours were found in any lobe but had a predilection for the frontal and parietal regions. Age incidence ranges between 2 1/2 months to 26 years and sex incidence is about equally divided (18,22,23,24,27,36).

The CT appearance of neuroblastoma has a tendency to be a rounded, well circumsribed and commonly calcified lesion. Contrast enhancement CT scan exhibits heterogeneous lack of opacification of the cystic and necrotic foci. But these findings are not useful in differentiating primary cerebral neuroblastoma from other primary neuroectodermal neoplasms. ANgiogram reveals an avascular mass lesion (4,23). Our case presented similar findings.

Primary treatment of neuroblastoma is still surgical removal. Chemotherapy, radiotherapy and immunotherapy may also augment the success of the treatment. Unfortunately we could not apply all these treatments in our case.

Neuroblastoma should be considered in the differential diagnosis of neonatal period tumours.

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