CASE REPORT

Primary Neuroectodermal Tumour of Scalp in Infancy

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Abstract

A case of orimary neuroectodermal tumor of scalp in infancy is presented.

Key words

Neuroectodermal, Scalp

Introduction

Tumours of scalp and skull are rare. These tumours can be primary or secondary. These tumours are totally invasive. Primary Neuroectodermal tumours are seen in children and young adults. These tumours are aggressive in nature and their prognosis is poor. Aggressive surgery and chemotherapy may improve survival rate. Histopathology of these tuours is akin to medulloblastoma.

Case Report

A three month old baby presented with a progressively enlarging swelling in occipital region of two months duration. On examination, there was a swelling 4" x 3" in occiput which was non-inflammatory, firm with underlying bone defect. Skin over the swelling was intact. CT scan showed a soft tissue mass with involvement of bone, no intracranial extension was seen (Fig. I). At operation, swelling was completely excised. The involved portion of the bone was removed, but dura was completely intact. Histopathology of the tumor proved it to be a case of PNET (Primary Neuroectodermal Tumour) (Fig. 2). Child was subjected to post-operative radiotherapy in view of invlvement of bone. There is no recurrence locally and elsewhere after three months of surgery.

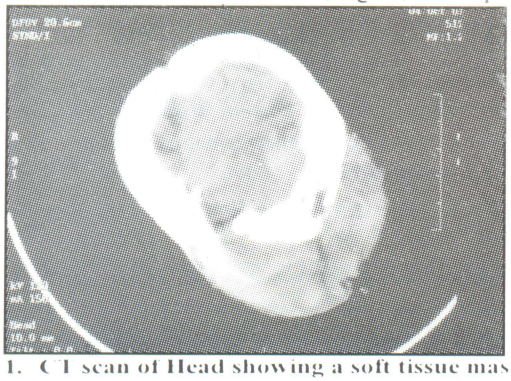

Fig 1. CT scan of Head showing a soft tissue mass in occipital region with destruction of bone.

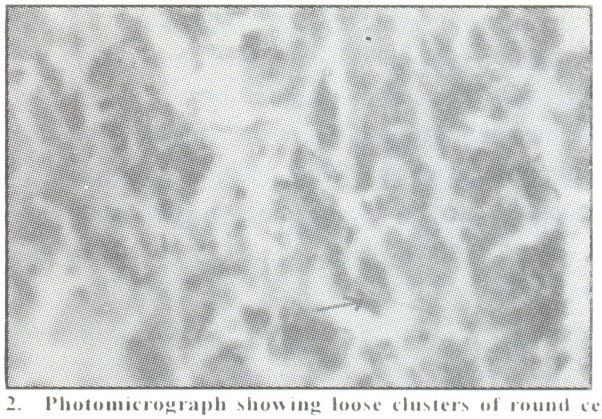

Fig 2. Photomicrograph showing loose clusters of round cells with a tendency to form acinar structures-arrow (x400)

Discussion

Primitive neuroectodermal tumour is a rare soft tissue neoplasm occurring in children and young adults. It is derived from a carcinogenic alteration of pleuripotent neural crest cells. The term primitive neuroectodermal tumour was first used in 1973 for tumours that were primitive but contained foci interpreted as glial or neuroblastic

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differentiation of primitive neuroectodermal cells. These tumours are morphologically identical to medulloblastoma, but arise in sites other than cerebellum. Intradermal primitive neuroectodermal tumours in adults not only are rare but tend to metastasize intracranially. These tumours respond poorly to therapy. Distinction between PNET and medulloblastoma may seem semantic. Caution is recommended in the application of the term PNET to those cerebral hemispheric lesions in which foci of differentiation are prsent in an otherwise largely primitive neoplasm. Zimmerman *et. al.* (1) reported 10 cases over a period of 8 years. Parados *et. al.* (2) reported treatment of high risk medulloblastoma and other primitive neuroectodermal tumours with reduced dose craniospinal radiation therapy and multi-agent nitrosourea based chemotherapy. Kosnik *et. al.* (3) reported a series of 18 cases of which 3 were spinal. They opined that these tumours are highly malignant (40% survival at 6 months, 10% survival at one year and all patients died in a span of two years). Russo *et. al.* (4) reported comparative genetic hybridization in patients with supratentorial and infratentorial Primitive neuroectodermal tumours. Parados *et. al.* (5) reported hyperfractionated craniospinal radiation therapy for primitive neuroectodermal tumours. Kimber *et. al.* (6) reported Primitive neuroectodermal tumours : Anatomic and outcome. Tumours arising from head and neck had an intermediate prognosis (37%). Authors have reported that complete excision reduced chance of local recurrence but did not prevent metastatic spread. Banerjee *et. al.* (7) reported that neuroectodermal tumours are rare. A correct diagnosis can be made by demonstrating neuroendocrine granules on electron microscopy and a combination of Mic_2, $Beta_2$ microglobulin. Jones *et. al.* (8) reported that primitive neuroectodermal tumours are aggressive round cell tumours whose prognosis is poor. Aggressive surgery and chemotherapeutic agents may improve long term survival. Dick *et. al.* (9) reported imaging of non-CNS neuroectodermal tumours : Diagnostic features and correlation with outcome. Womer *et. al.* (10) reported extracranial primitive neuroectodermal tumours. Fear *et. al.* (11) reported role of prenatal and neonatal factors in the etiology of childhood malignant neoplasms of the brain. Paulino *et. al.* (12) stated that supratentorial primary neuro ectodermal tumours have worse overall survival.

In our case, histopathology of tumour was akin to medulloblastoma. Since dura was intact, this patient was labelled primary neuroectodermal tumour. It is rarity of the case and its confusing histopathology that prompted us to report this case.

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