

MEDULLOBLASTOMA: A LIFE THREATENING CANCER OF INFANCY AND CHILDHOOD

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ABSTRACT

Medulloblastoma is a rare and aggressive tumor of the brain that usually occurs in children or young adults with a tendency to metastasize. The incidence in infant is rare. We came across one such child who presented with slight disorientation, vomiting, irritable personality and somewhat large size head. The clinical, radiological preoperative diagnosis was space occupying lesion in the posterior fossa. On histological examination it turned out to be Medulloblastoma of infratentorial region in posterior fossa.

KEY WORDS: Medulloblastoma, Posterior fossa, Tentorium.

INTRODUCTION

Medulloblastoma was first described by Bailey and Cushing. They suggested that cerebellar medulloblastoma cells may develop along spongioblastic or neuroblastic lines.¹ Currently, it is still controversial whether medulloblastoma is a primitive neuroepithelial tumor that is incapable of differentiation or a tumor with a bipotential capacity for histologic differentiation.² Chromosomal gains and losses are more common in anaplastic medulloblastoma than in non anaplastic ones.³ Medulloblastoma reveals clusters and isolated small, round malignant cells with hyperchromatic nuclei and indistinct cytoplasm. Typical Homer-Wright rosettes are seen in the well

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differentiated type, but they are poorly formed in the undifferentiated type.⁴ At present, children with medulloblastoma are divided into two disease groups: Average risk patients are the ones who are older than the age of three years with non-metastatic and totally or nearly totally excised (d²1.5 cm²) tumor on post operative MRI. The patients not falling in this criteria and including those age below three years are regarded as high-risk group (HR). Predicting favorable outcome has been difficult and improved stratification clearly is required to avoid both under-treatment and over treatment.⁵ Two rare sub types at extreme ends of the histologic spectrum are medulloblastomas with extensive nodularity and large cell/anaplastic medulloblastoma.⁶

We diagnosed a case of medulloblastoma with presentation of large anaplastic type cells, one of rare entity on histopathology, at Gulab Devi Hospital, Lahore. The case is reported with a review of available literature.

CASE REPORT

A child of 1.4 years was admitted with history of slight disorientation, intermittent vomiting, irritable personality and some what large size head for the last one year. There was no history of trauma, fever or weight loss. Laboratory investigation revealed an ESR of 45mm

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at the end of 1st hour and Hemoglobin of 10.6gm/dl. X-Ray skull revealed a solitary mass in the posterior fossa. Computed Tomography (C.T Scan) showed a solitary lesion in the lateral hemisphere of the cerebellum. After the clinical and radiological diagnosis of intracranial tumor, surgical intervention was planned.

On general examination, the material revealed multiple, firm, gray black pieces of tissue, measuring 0.8x0.8x0.6cm in aggregate, weighing 09 gms. The entire specimen was processed in two blocks in automatic tissue processor, embedded in paraffin and four to five microns thick sections were cut. The sections were stained with hematoxylin and eosin. Light microscopic examination showed highly cellular small to large size cells crowding with hyperchromatic nuclei and indistinct cytoplasm (Fig-1). In one area pseudorosette formation was seen (Fig-2). There was increased abnormal mitotic activity. Vascular proliferation was also evident.

DISCUSSION

The significant feature of the case under view was the occurrence of a medulloblastoma in the posterior fossa of a boy of 1.4 years age. Medulloblastoma is a rare tumor of infants. It is uncommon under the age of three years.⁶ Central nervous system malignancies comprised 13% of infant cancer, with an average

annual incidence rate of nearly thirty per million infants. Out of these, medulloblastoma accounts for five per million infants. There is around a two-fold higher average annual incidence in males than in females. The peak incidence in children is between five and ten years of age group.⁷

Bailey and Cushing¹ theorized that medulloblastoma was derived from nests of undifferentiated cells capable of both neuronal and glial differentiation. Schindler and Gullotta⁸ suggested that the positive staining is a result of GFAP remnants from degenerated astrocytes, which were phagocytized by macrophages. Medulloblastoma on the basis of morphology divided into two varieties-classic and desmoplastic. Classic medulloblastomas are solid masses composed of small closely packed sheets of the cells. Nuclei are often densely hyperchromatic and round, with no definable cytoplasm. Peripheral pseudorosette formation is often seen. Desmoplasia is usually present in childhood and is accompanied by Homer-Wright rosettes. Two rare subtypes at extreme ends of the histologic spectrum i-e medulloblastomas with extreme nodularity and largely anaplastic are associated with better and worse clinical outcomes, respectively.⁷ c-myc, N-myc, 2q14-22, 3p23, 5p14-pter, 8q24, 9p22-23, 10p12-pter, 12p11-12, 17p11-12 and Xp11 chromosomal amplification are detected in

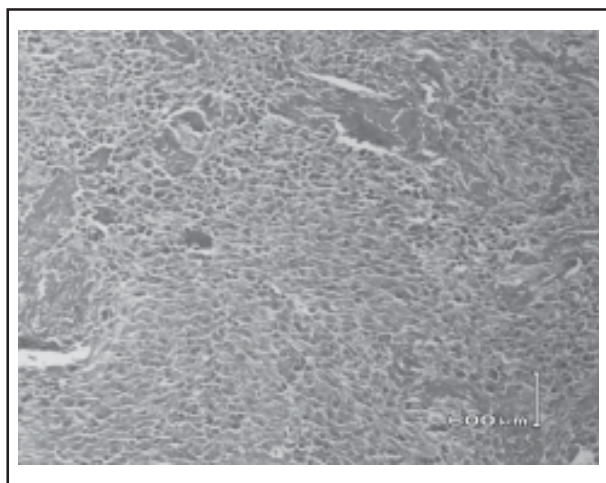


Fig-1: Medulloblastoma with difused clusters of small round malignant cells without typical rosette formation X20

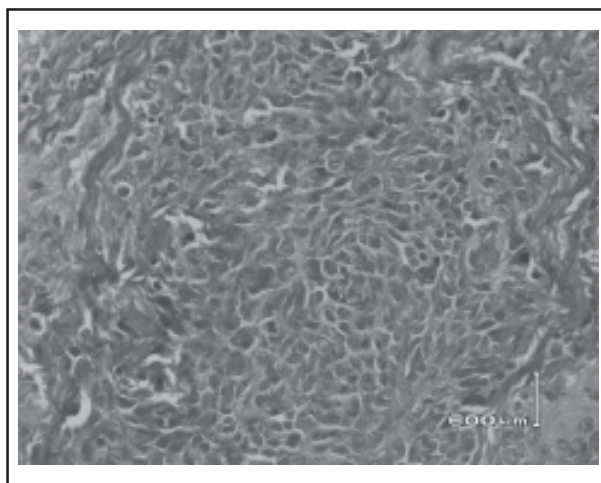


Fig-2: Medulloblastoma with modularity and large size anaplastic malignant cells without typical differentiation X40

different medulloblastomas. Angiogenic factors also contribute a direct role in PNET/MB tumor growth.⁹

Current therapy for this disease includes maximum surgical resection, whole neuraxis radiation and chemotherapy. The prognosis of medulloblastoma in children under three years of age is poor, because of the high morbidity of radiotherapy in children under three years old. Despite this aggressive treatment, only 60% of children with medulloblastoma will be cured and most of these will suffer long-term side effects. One study shows that children who survive medulloblastoma suffer a loss of normal-appearing white matter, an associated decline in intellectual function and long term endocrine deficiencies.¹⁰

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