

## Medullomyoblastoma : A Case Report

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**Abstract :** A rare tumour occurring in the cerebellar midline and consisting of a mixture of medulloblastoma and mature and immature striated muscle is usually referred to as "medullomyoblastoma". The origin and nature of medullomyoblastoma are controversial.

In this report, we describe the histological, and immunohistochemical features of medullomyoblastoma occurring in the cerebellar vermis of an 11-year-old boy.

**Key Word:** Medullomyoblastoma

### INTRODUCTION

A rare tumour occurring in the cerebellar midline and consisting of a mixture of medulloblastoma and mature and immature striated muscle is usually referred to as "medullomyoblastoma". The origin and nature of medullomyoblastoma are controversial (1-3,5-7,9,12,13).

### CASE REPORT

An 11-year-old boy was admitted with headaches, and vomiting for 30 days. During examination the child was drowsy with bilateral papilledema and bilateral cerebellar signs. Examination of other systems revealed no abnormality.

Routine hematological investigation, serum chemistry and chest x-ray were normal. A cranial computerized tomographic scan showed a high-density, contrast-enhancing lesion in the cerebellar midline. Suboccipital craniectomy and C1 laminectomy were performed. A large, grayish white, moderately vascular tumour was seen in the vermis that was filling the fourth ventricle. Nearly total excision was accomplished.

Postoperatively, the patient had an uneventful recovery and was discharged. At follow-up 2 months after operation, the patient was well.

Tissue from the cerebellar tumour was fixed in 10% buffered formalin and embedded in paraffin. Most sections were stained with haematoxylin and eosin and selected sections with phosphotungstic acid-haematoxylin (PTAH), periodic acid-Schiff (PAS) with and without diastase. Immunoperoxidase stains for glial fibrillary acidic protein (GFAP, Dako), desmin (Dako) and neuron-specific enolase (NSE, Dako) were performed on selected sections with appropriate positive and negative controls.

**Light Microscope Findings:** The tumour showed two distinct patterns in different areas. One area looked like typical medulloblastoma with closely packed groups of small cells separated by a sparse amount of connective tissue stroma (Fig. 1). The cells had round to oval hyperchromatic nuclei and scanty cytoplasm. Numerous mitotic figures were seen.

Some regions of the tumour were less cellular and contained two populations of cells: small undifferentiated cells with pleomorphic nuclei and scant cytoplasm, and oval or elongated strap-like cells with conspicuous eosinophilic cytoplasm. Some round cells with similar cytoplasm were also present. There were pleomorphism, and occasional giant cells. Some of the strap-shaped cells showed typical cross-striations with PTAH, indicative of rhab-

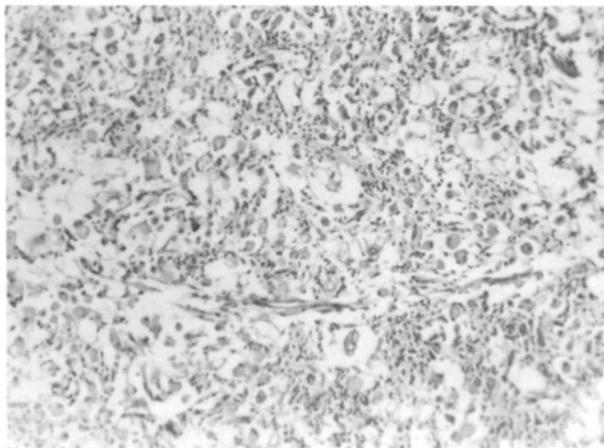


Fig. 1: Two populations of cells: small undifferentiated cells and oval or elongated cells with eosinophilic cytoplasm.

omyoblast (Fig. 2). Diastase digestible PAS positivity was seen in the cytoplasm of these large, eosinophilic cells. There was definite orientation of the muscle cells around blood vessels.

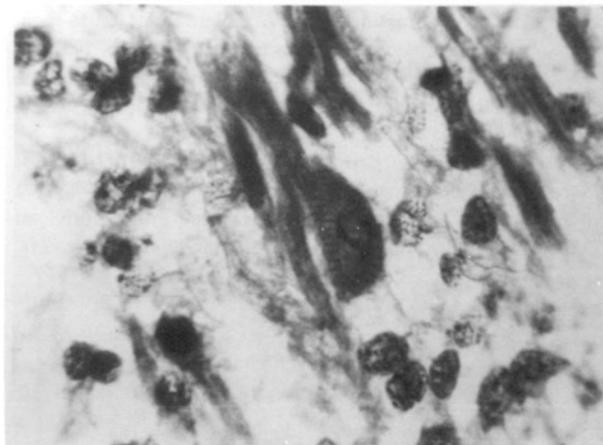


Fig. 2: Strap-like cells with cross striations.

**Results of Immunoperoxidase Staining:** In the medulloblastoma areas of the tumour, the cells did not exhibit positive staining for GFAP. The stain for desmin was positive in large eosinophilic cells. NSE stain revealed diffuse cytoplasmic staining of some small cells with hyperchromatic nuclei. Also some large eosinophilic cells were positive for this enzyme.

#### DISCUSSION

The clinical and cranial CT findings in this patient indicated the presence of a cerebellar midline tumour believed to be either a medulloblastoma or an

astrocytoma. The pathological diagnosis of medullomyoblastoma was based on the presence of a densely cellular primitive neuroectodermal component that contained less-cellular areas in which striated muscle fibre was found.

The histogenesis of the myogenic component of medullomyoblastoma is controversial. The concept that this tumour might represent a variant of a malignant teratoma or a teratoid tumour, was first proposed by Ingraham and Bailey (5) and has been supported by others (8). This interpretation was criticized by Stahlberger and Friede (12) because the nonectodermal component in these tumours is restricted to cross-striated muscle fibres. Other mesodermal or endodermal components are not seen. However, a recent report by Chowdhury et al (2) brings notable support to the teratoma hypothesis.

Some authors have suggested that the muscle fibres in medullomyoblastoma may be derived from embryonal pluripotential mesenchymal cells present within or near the tumour (7,12,13).

In the central nervous system such a pluripotential mesenchymal cell has been called neuromesenchyme or ectomesenchyme, which is derived from the neural crest. The leptomeninges are considered to be an ectomesenchymal derivative, and rarely have both non-neoplastic striated muscle and various mesenchymal tumours, including rhabdomyosarcoma (10). The leptomeningeal component that invests the perforating blood vessels is deemed the likely source of both primary intraparenchymal rhabdomyosarcomas and the myoblastic component of medulloblastoma. In the case of medullomyoblastoma, it has been suggested that the neoplastic medulloblastoma cells somehow induce perivascular ectomesenchymal cells to differentiate into muscle fibres (7,12). Indeed, there is a tendency for the muscle fibres to localize around blood vessels in some cases. The presence of melanin-containing cells in some cases of medullomyoblastoma is seen as further support for a neural crest-derived ectomesenchymal origin (1,3).

Walter and Brucher propose that the myoblastic component is derived from the multipotential endothelial cells(13). A final hypothesis concerning the histogenesis of the muscle fibres in medullomyoblastoma is that they originate directly from the primitive neuroepithelial (medulloblastoma) cells (11).

Lenon and Peterson (6) have shown skeletal muscle differentiation in cultures of rat glial cells derived from nitrosoethylurea-induced brain tumours. There is a report of rhabdomyoblastic differentiation in a cell line derived from a classical vermian medulloblastoma (4). These findings demonstrate the myogenic potential of medulloblastoma cells.

In summary, we believe that the most likely explanation for the origin of muscle fibres in medullomyoblastoma is that they arise through the induction of perivascular ectomesenchymal cells or directly from the medulloblastoma cells. Indeed, in this case there is a tendency for the muscle fibres to localise around blood vessels. Also, we observed positive staining for NSE in some large eosinophilic cells. Thus, medullomyoblastomas are probably best regarded as either mixed neuroepithelial-mesenchymal neoplasm or a variant of medulloblastoma rather than malignant teratoid tumours.

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