

## STUDY OF EMBRYONIC TUMOURS OF THE CENTRAL NERVOUS SYSTEM

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### ABSTRACT

#### BACKGROUND

Primary brain tumours do not spread to other body sites, and can be malignant or benign. Secondary brain tumours are always malignant. Both types are potentially disabling and life threatening. There are a number of distinct types of brain cancers within the brain, and the treatments and their outcomes vary greatly based on pathologic and histologic diagnosis. Embryonal tumours are the least studied tumours. So a study was done in our institution to understand the same

#### METHODS

One hundred cases of brain tumours that have turned up in the Department of Medicine, A. J. Shetty Institute of Medical Sciences, Mangalore.

#### RESULTS

In the present study embryonal tumours constituted 1(8.33%) of Neuroepithelial tumours. This tumour occurred in Paediatric age group.

#### CONCLUSION

The embryonal tumour forms a very small share of Intra-cranial tumours. The future of the study is very good and further studies has to be done to understand the demographic patterns of the disease.

#### KEYWORDS

Embryonal, Tumours, Central Nervous System, Histology.

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**INTRODUCTION:** Primary brain tumours do not spread to other body sites, and can be malignant or benign. Secondary brain tumours are always malignant. Both types are potentially disabling and life threatening.<sup>1</sup> There are a number of distinct types of brain cancers within the brain, and the treatments and their outcomes vary greatly based on pathologic and histologic diagnosis. More recently, researchers are identifying new therapies based on increased knowledge of cellular and molecular biology.<sup>2</sup>

One of the least studied type of the brain tumours are the embryonal tumours. There is no clear cut classification to describe these. It is considered as a form of neuroepithelial tumour.<sup>7</sup>

The Embryonal tumours are a type of Neuro-Epithelial tumours and the types are described below as follows.

**Medulloblastoma [WHO grade IV]:** A malignant, invasive embryonal tumour of the cerebellum with preferential manifestation in children, predominantly neuronal differentiation, and an inherent tendency to metastasize via CSF pathways.

**Central Nervous System Primitive Neuroectodermal Tumours [WHO grade IV]:** A heterogenous group of tumours occurring predominantly in children and adolescents. They may arise in the cerebral hemispheres, brain stem or spinal cord and are composed of undifferentiated or poorly differentiated neuroepithelial cells which may display divergent differentiation along neuronal, astrocytic and ependymal lining. CNS/supratentorial PNET is an embryonal tumour composed of undifferentiated or poorly differentiated neuroepithelial cells. Tumours with only neuronal differentiation are termed cerebral neuroblastomas or if ganglion cells are also present cerebral ganglioneuroblastomas. Tumours that recreate features of neural tube formation are termed medulloepitheliomas. Tumours with ependymoblastic rosettes are termed ependymoblastomas. Features common to all CNS PNET variants include early onset and aggressive clinical behaviour. Tumours with only neuronal differentiation are termed cerebral neuroblastomas. Precise incidence is difficult to determine because of differing viewpoints regarding classification and the rarity of these tumours. One percent of 933 primary paediatric CNS neuroepithelial tumours were found to be located in the cerebrum or suprasellar region; among CNS PNETs, 10 of 178 [5.6%] were located in these regions.

The age range for CNS PNET is 4 weeks to 20 years with a mean of 5.5 years. One hundred twenty patients were >1 year at diagnosis in a study conducted by Cheung YI, Sahota

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A, Cheung KN et al.<sup>2</sup> In a series of 35 cases, personally examined by Rubinstein et al, the age distribution was as follows: 30 of 35 cases (85%) occurred in the first decade; 23 or (65%) occurred within the first half of the first decade.<sup>3</sup> In a study conducted by Ahadevaara P et al 8 were males and 5 were females among 13 individually reported cases. The male: female ratio for supratentorial PNETs and cerebral neuroblastomas is 1.2:1. These tumours are found most commonly in the cerebrum, but can also be encountered in the spinal cord or suprasellar region.

**Atypical Teratoid/Rhabdoid tumour [WHO grade IV]:**

A highly malignant CNS tumour predominantly manifesting in young children, typically containing rhabdoid cells, often with primitive neuroectodermal cells and with divergent differentiation along epithelial mesenchymal neuronal or glial lines; associated with inactivation of the NI1/Hsnf5 gene in virtually all cases.

**Embryonal tumours:**

**Medulloblastoma:**

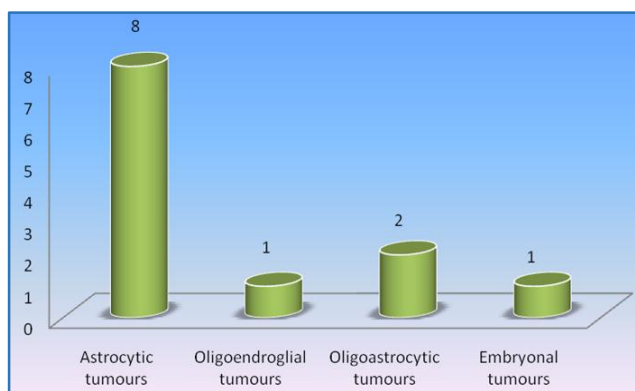
1. Desmoplastic nodular medulloblastoma.
2. Medulloblastoma with extensive nodularity.
3. Anaplastic medulloblastoma.
4. Large cell medulloblastoma.

**RESULTS:**

**Tumours of Neuroepithelial Tissue:**

Histological types	Cases	% of intracranial tumours	% of neuroepithelial tumours	Gender	
				M	F
Astrocytic tumours	8	8	66.7	3	5
Oligodendroglial tumours	1	1	8.3	-	1
Oligoastrocytic tumours	2	2	16.7	2	
Embryonal tumours	1	1	8.3		1
<b>Total</b>	<b>12</b>	<b>12</b>	<b>100</b>	<b>5</b>	<b>7</b>

*Table 1: Showing breakdown of tumours of neuroepithelial tissues*



**Fig. 1: Breakdown of neuroepithelial tumours**

**CNS primitive Neuroectodermal tumour:**

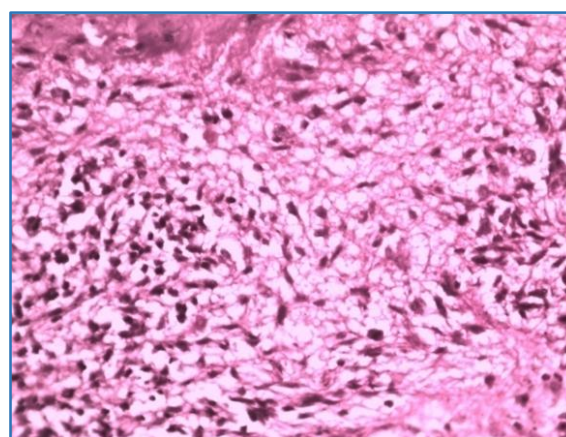
1. CNS neuroblastoma.
2. CNS ganglioneuroblastoma.
3. Medulloepithelioma.
4. Ependymoblastoma.

**Atypical teratoid/rhabdoid tumour.**

In the present study an effort is made to study the embryonal tumours.

**AIMS AND OBJECTIVES:** To study the embryonal tumours of the central nervous system.

**MATERIALS AND METHODS:** One hundred cases of brain tumours that have turned up in the Department of Medicine, A. J. Shetty Institute of Medical Sciences, Mangalore. The cases were diagnosed based on the histopathological reports of the specimen that were surgically removed and was diagnosed in the Department of Pathology. The neuroepithelial cells then were segregated and further studied for embryonal tumours.

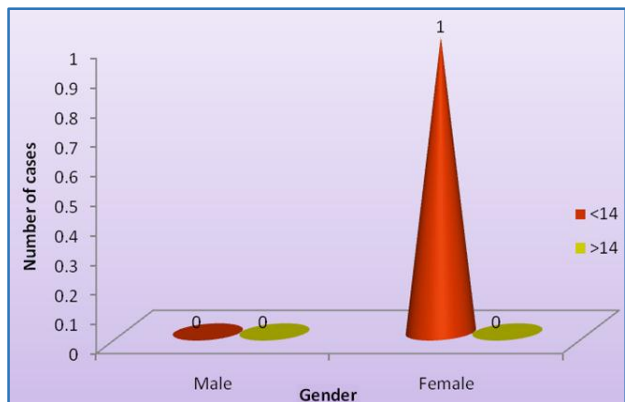


**Fig. 2: Microphotograph of differentiating neuroblastoma. Small round cells (Neuroblasts) in clusters separated by shwannian elements**

**Embryonal Tumours:**

Age in years	No. of cases	Gender	
		M	F
<14	1	0	1
>14	0	0	0
<b>Total</b>	<b>1</b>	<b>0</b>	<b>1</b>

**Table 2: Showing age and Gender distribution of Embryonal tumours**



**Fig. 3: Age and gender distribution of oligoastrocytic tumours**

In the present study embryonal tumours constituted 1 (8.33%) of neuroepithelial tumours. This tumour occurred in Paediatric age group. It was found in a female patient aged 5 years. Lesion was Located in right cerebral hemisphere. Child presented with visual disturbances and unsteady gait. Microscopically showed sheets of small blue cells with fibrillary background with areas showing neuronal and Schwann cell differentiation. (Fig. 1).

**DISCUSSION:** In the present study Embryonal tumour comprised 1(8.33%) of all Neuroepithelial tumours. This was diagnosed in patient aged 5 years. Embryonal tumours were grouped under “Gliomas” by Pal and Chopra et al<sup>4</sup> and Banerjee et al.<sup>5</sup> In this study Embryonal tumour is grouped

under neuroepithelial tumours as per WHO classification. The percentage of Embryonal tumours reported by Banerjee et al was 9.2% of all gliomas, with an average age of 8.5 years. Pal and Chopra et al<sup>4</sup> observed incidence of 9.1% of all Gliomas with an average age incidence of 13 years. In the present study it occurred in a 5-year-old female in cerebral location.

**CONCLUSION:** The embryonal tumour forms a very small share of Intra-cranial tumours. The future of the study is very good and further studies has to be done to understand the demographic patterns of the disease.

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