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`` Infratentorial pediatric brain tumors: An institutional experience``

KEYWORDS: Infratentorial tumor, Children under 14 year

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ABSTRACT *OBJECTIVE: To investigate the prevalence of infratentorial tumors and their age and gender- wise distribution of the lesions.*

MATERIAL AND METHODS: A retrospective review of pediatric primary infratentorial brain tumors, encountered over 3 years (January 2013 to December 2015).

RESULTS: Study includes 32 pediatric infratentorial tumors. Male to female ratio were 1.9:1. The cases were divided into 5 age groups each covering three years of life (0-2, 3-5, 6-8,9-11 and 12-14 years). Most of the patients were in age group 3 (40.6%)and the least number of patients were found in age group 1 (6.25%) and 4 (12.5%). The 32 tumors were categorized based on histology and morphological basis. The most frequent tumor was medulloblastoma (56%) followed by ependymoma (18%).

CONCLUSION: Demographic and histopathological profile of cohort in the present study do not differ substantially from that found in other hospital-based and population-based studies except for slight higher frequency of ependymoma.

INTRODUCTION

The Infratentorial or posterior fossa compartment of the cranial vault, which extends from tentorium cerebelli superiorly to foramen magnum inferiorly, housing the cerebellum and most of the brainstem, specifically the pons and medulla. As the name indicates, the midbrain or mesencephalon is more centrally located and crosses through the tentorial incisure or notch, straddling the supratentorial and infratentorial compartments. Osseous boundaries of the posterior fossa include the clivus anteriorly, the petrous ridge of the temporal bones anterolaterally, the mastoid portion of the temporal bones laterally, and the occipital bone posteriorly-inferiorly.

Primary brain tumors are the most common solid tumors in the pediatric population, comprising 20% to 25% of all childhood cancers. About 60% to 70% of all pediatric brain tumors originate in the posterior fossa[1]. The reason that pediatric brain tumors have a propensity to occur in the posterior fossa has not yet been elucidated. By far, the most common posterior fossa tumors of childhood are medulloblastomas, epen- dymomas, and astrocytomas. As for uncommon tumors, these include atypical teratoid/rhabdoid tumors (AT/RTs), teratoma, hemangioblastoma, dermoid, and epidermoid. Each tumor may have a vastly different prognosis and response to treatment depending on the pathologies.

Despite researchers' best efforts, the true prevalence of infratentorial tumors remains uncertain in Gwalior region. The present study was designed to investigate the prevalence of infratentorial tumors and their age and gender- wise distribution of the lesions.

MATERIALS AND METHODS

This was a retrospective study based on data collected from Department of Pathology, G R medical college Gwalior. The case summaries, histopathological record and discharge reports of all children up to 14 years of age, were reviewed who were operated for infratentorial tumors during period from January 2013 to December 2015. In addition to the location and histological types of the tumor, patient demographics including age, sex, and symptoms were also recorded. Patients with metastatic tumor and vascular malformation were excluded from the study. Furthermore, patients who were diagnosed as a case of primary brain tumor clinically and on imaging (e.g., brain stem glioma) but not biopsied and treated by chemoradiation were also excluded from the study. All histopathological diagnoses were made according to the 2007 World Health Organization classification system. Thus, with all above considerations we had a cohort of 32 pediatric patients with histopathologically proven brain tumors. . The data was compiled, summarized and analyzed using frequency distribution and percentage proportion.

RESULTS

Study includes 32 pediatric infratentorial tumors. Out of 32 patients 21(65.35) were male and 11(34.37%) were female. Male to female ratio were 1.9:1. The age of the patients was divided in to five groups (Table 1). The mean age of patients was 7.14 years (range, 2-14 years). Most of the patients were in age group 3 (40.6%)and the least number of patients were found in age group 1 (6.25%) and 4 (12.5%) as shown in Table 1. A distinct overall male predominance was noted in all tumor types except brain stem astrocytoma as shown in table 2.

Table 1: Age wise distribution of infratentorial tumors in children under 14 years of age

Age	Medulloblastoma	Ependymoma	Brain stem glioma	Cerebellar Pilocytic astrocytoma	Tuberculoma	Dermoid	Epidermoid
<1y-2y	4	0	0	0	0	0	0
3y-5y	5	0	2	0	0	0	0
6y-8y	9	1	1	2	0	0	0
9y-11y	0	2	0	0	0	0	0
12y-14y	0	3	0	0	1	1	1
Total	18	6	3	2	1	1	1

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Table 2: Gender wise distribution of infratentorial tumors in children under 14 years of age

Year	Medulloblastoma		Ependymoma		Brain stem glioma		Cerebellar Pilocytic astrocytoma		Tuberculoma		Dermoid		Epidermoid	
	M	F	M	F	M	F	M	F	M	F	M	F	M	F
2013	4	2	1	2	0	1	0	2	0	0	0	0	1	0
2014	2	2	0	0	0	1	0	0	1	0	0	0	0	0
2015	7	1	3	0	0	1	0	0	0	0	0	1	0	0
Total	13	5	4	2	0	3	0	2	1	0	0	1	1	0

It was evident from histology that medulloblastoma (56%, figure 1) was the most common group of infratentorial tumors in children (Table 3). The mean age of patients for medulloblastoma was 6 years with most patients in age group 3. Ependymoma was the second most dominant tumor type in our study (18%, Figure 2). The mean age of patients for ependymoma was 10 years (range, 8-14), with most patients in age group 5 and 4. Brain stem astrocytoma ranked third in the list with a frequency of 9.3% (Figure 3), other less frequent tumors were tuberculoma (Figure 4) and epidermoid (Table 3, Figure 5).

Table: 3 Histological diagnoses of infratentorial tumors in children less than 14 years of age

SN	Diagnosis	Numbers	Percentage
1	Medulloblastoma	18	56
2	Ependymoma	6	18
3	Brain stem astrocytoma	3	9.3
4	Cerebellar Pilocytic astrocytoma	2	6.25
5	Cerebellar Tuberculoma	1	3.1
6	Dermoid	1	3.1
7	Epidermoid	1	3.1
8	Total	32	100

Headache was most frequent complaint (87%) followed by imbalance (84%) and vomiting (78%). In some, a history of visual disturbances, enlarge head size was reported by the parents, including 6th and multiple cranial nerve palsy (Table 4).

Table 4: The presenting symptoms and signs of infratentorial tumors in children under 14 year of age

Symptoms/ Signs	Numbers	%
Headache	28	87
Imbalance	27	84
Vomiting	25	78
Visual disturbances	10	31
Enlarge head size	14	43
6th cranial nerve palsy	6	18.75
Seizure	2	6.2
Loss of consciousness	2	6.2
Multiple cranial nerve palsy	2	6.2

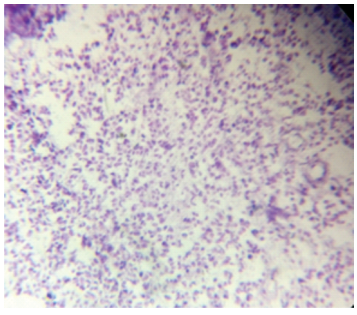

Figure 1: Medulloblastoma: Section reveals undifferentiated looking elements with little definable cytoplasm and occasional homer wright rosettes formation (10x view)

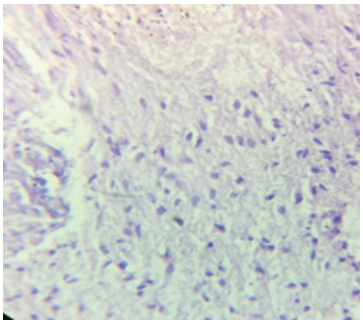

Figure 2: Ependymoma: Section showing true ependymal rosettes containing a well defined central lumen

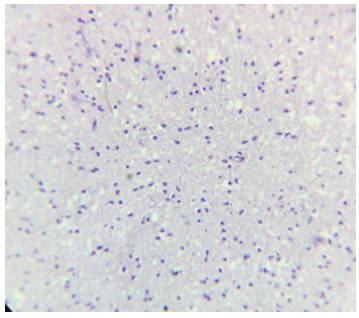

Figure 3: Astrocytoma: Section shows cells with mild nuclear pleomorphic and only modest hyperchromasia. The absence of mitotic activity supports it as low-grade lesion

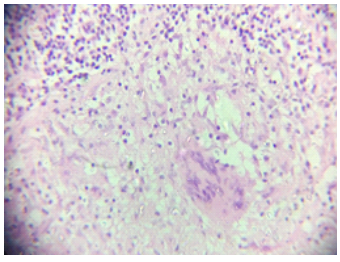

Figure 4: Tuberculoma: section showing an ill-defined granuloma with giant cell

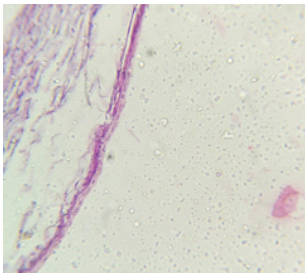

Figure 5: Epidermoid: section-showing cyst lining with thinning at places

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DISCUSSION

Tumors of the central nervous system are the second most common childhood tumors (20%) after leukemia (37%) and the most common solid pediatric tumors comprising 40-50% of all tumors. Posterior fossa (infratentorial) tumors comprise 54% to 70% of childhood brain tumors compared to 15%-20% in the adult population [2].

The present study was designed to determine the frequency of infratentorial tumor among children less than 14 year of age. Our study revealed male predominance with over all male to female ratio 1.9:1, which is inline with previous studies (Kadri et al.[3], Ahmed et al.[4], 2007;Menon et al.[5], 2007;Rehman et al.[6], 2009).

Present study revealed most of the patients in age group 6-8 years, which is nearly in accordance with a previous study done by Ahmed et al. [4](2007) who reported most cases in age group 5-9 years. However Velema and Percy [7](1987) and Menon et al.[5], (2007) reported most cases in lower age group.

The medulloblastoma was the most frequent tumor of our study constituting 56% of total cases. A predominance of medulloblastoma was reported by Packer et al in 1990 [8]with 74(36%) PNET; 57(28%) cerebellar astrocytoma and 18(9%) ependymoma. Mosso et al[9] in 1992 produced the results of childhood cancer registry of Torino (Italy) over a period of 20 years. In this series medulloblastoma were 56/293 (19.11%), astrocytoma 80/293 (27.3%), ependymoma 14/293 (4.78%) while a significant number of cases [80/293 (27.3%)] were labelled as "not otherwise specified". A few studies have also reported a preponderance of astrocytoma in their series. This is most often seen in studies where the upper limit of the pediatric age was 20 years. Farewell et al[10], in 1977 reported a series of 488 pediatric CNS tumors (467 intracranial) over a period of 39 years. In their study astrocytoma comprised 28%, medulloblastoma 25% and ependymal tumors 9% of the total. The controversies over the morphological hierarchy are more diverse with medulloblastoma and astrocytoma competing for the coveted highest frequency. These controversies arise probably because arbitrary definitions are used by individual scientists, whilst defining the studies, thus making them non-comparable. In the case of pediatric brain tumors the age limits used for certain developmental periods (e.g. the widely divergent ages of 12, 15 and 20 years for the upper limit of childhood) and the definition of age itself (meaning age at the time of first clinical symptoms or at diagnosis) and in the different approaches to histopathological diagnosis, as the terminology and classification of certain tumors have either changed or been handled arbitrarily over the years (Rickert et al[11]., 1997).

Symptoms of infratentorial tumors usually result from either compression of vital structures or raised intracranial pressure. Common presenting features include cerebellar symptoms, multiple cranial nerve palsies, headaches, vomiting and blindness due to raised intracranial pressures[12]. Similarly the most frequent symptoms in our study were headache (87%) followed by imbalance (84%) and vomiting (78%).

Although Tabatabaei SM et al[2] 2012 has reported Cerebellar symptoms were the most common cause of presentation (80.9%) followed by headaches (73.8%) and vomiting (38.1%). Visual problems were found in 7.14% of their patients probably due to late presentation.

CONCLUSION

From the present series, we conclude that, the frequencies of major histologic types of pediatric infratentorial tumors found in the study do not differ substantially from that found in other developed and developing countries except for slight lower frequency of cerebellar astrocytoma. Medulloblastomas and ependymoma, which form the major histologic types in pediatric patients, need special attention. Epidemiological surveillance of various histological types of pediatric infratentorial tumor is of great importance from public health perspective. It helps in planning the distribution of infrastructure and resources toward the disease management and preventive programs. In countries like India where there is a scarcity of data because of inadequacies in tumor registration, such hospital based studies have a major role to play in such planning.

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