



AN ARCHIVAL STUDY TO ANALYSE THE HISTOPATHOLOGICAL SPECTRUM OF CHILDHOOD CENTRAL NERVOUS SYSTEM TUMORS

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ABSTRACT

Background: Central Nervous System (CNS) tumors are the second most common tumors in children and a leading cause of mortality and morbidity worldwide. Till date there are very few published studies reporting the histopathological and the demographic profile of pediatric CNS tumors in India. **Aim:** This retrospective study was designed with the aim to analyse the histopathological and the demographic details of childhood CNS tumors reported at a tertiary care neurosciences hospital in India. **Material and Methods:** Data regarding age, gender, clinical features, tumor location and histopathology of 295 patients with CNS tumors (≤ 18 years of age) diagnosed between January 2004- December 2019 was collected retrospectively and analysed. **Results:** Amongst the 295 cases, mean age of presentation was 11.5 years. Slight male predominance was observed (M: F;1.8:1). Commonest clinical manifestations were symptoms and signs of increased intracranial pressure (81.4%) followed by focal neurological signs (18.6%). Commonest site was posterior fossa/cerebellum (34.9%), followed by cerebral hemispheres (23.7%), spine (14.2%), sellar/ suprasellar (6.8%), lateral ventricles (3.7%), cerebellopontine (CP) angle and cerebral convexities (3.1% each), 3rd and 4th ventricles (2.7% each), brainstem (1.7%), thalamus (1.4%), optic nerve and pineal region (0.7% each), cauda equina and sylvian fissure (0.3% each). Embryonal tumors (20.3%) constituted the most common tumor category. **Conclusion:** This study showed a male predominance with embryonal tumors being the most common tumor category and posterior fossa and the cerebellum being the most common sites. This study adds to the existing literature highlighting the histopathological and demographic profile of CNS pediatric tumors in India.

KEYWORDS : Central Nervous System, Pediatric tumors, World Health Organization, Histopathological types, Demographic details

Introduction

Tumors of the Central Nervous System (CNS) are the second most common tumors in the children after leukaemia, constituting approximately 15-35% of all childhood malignancies, and are a leading cause of mortality and morbidity in children worldwide.^{1,2} According to the Indian literature, CNS tumors account for 11.4% to 20.1% of childhood tumors.^{3,6}

These childhood CNS tumors differ significantly from adult CNS tumors in reference to their sites of origin, clinical presentation, tendency to disseminate early, histopathological features and their biological behaviour.

Over the years, an enhanced understanding of these biological differences between adult and childhood CNS tumors has led to various distinct molecular and genetic pathways and advanced therapeutic approaches for each tumor type creating a paradigm shift in the World Health Organisation classification of CNS tumors changing from WHO classification 2007 (based on histogenesis) to WHO classification 2016 (based on both, genotypic and phenotypic parameters). However, literature review has revealed only few Indian reports available related to the CNS tumors in children, thus highlighting the need for more and more institutional data to assess the actual disease load in India. In this purview, this present study reports the hospital-based histopathological and demographic profile of pediatric CNS tumors from our institute.

Objectives

- To study the histopathological and demographic profile of CNS tumors in children ≤ 18 years of age
- To study the clinical features, localization, histopathological types and WHO grades of paediatric CNS tumors

Materials and Methods

This is a retrospective study, based on the data collected from

a tertiary care neurosciences hospital of East Delhi, India. Data of CNS tumors in the pediatric age group (≤ 18 years of age) was collected from the Histopathology laboratory records where the samples were received from two tertiary care hospitals in India, including Institute of Human Behaviour and Allied Sciences and Guru Teg Bahadur Hospital, New Delhi. The histopathological records of 295 children ≤ 18 years of age with CNS tumors (January 2004- December 2019) were reviewed. Only patients with the proven histopathological diagnosis were included in the study. The cases were diagnosed with routine Haematoxylin and Eosin (HE) and immunohistochemistry and were categorized according to the most recent WHO classification prevalent at the time of diagnosis. In addition to the histopathological types and WHO grades of the tumors, location of the tumors, patients demographics including age and gender were also recorded. Patients with metastatic tumors, tumor like cystic lesions (arachnoid cysts, epidermoid cysts and colloid cysts), space occupying lesions of infectious etiology and vascular malformation were excluded from the study. Results were compared with the Indian and International data available as tumor registries and hospital-based studies. All statistical analyses were performed using the SPSS18.0 software package (SPSS).

Results

A total of 295 cases of pediatric CNS tumors were retrieved from the histopathology records of Department of Pathology from January 2004 to December 2019.

Age and sex distribution

Amongst the 295 cases the mean age of presentation was 11.5 years (ranged from 7 days to 18 years). The cases were subdivided into two age groups, 0-10 years, and 11-18 years. There were 117 patients (39.7%) in 0-10 years age group and 178 patients (60.3%) in 11-18 years age group. In the first decade (0-10 years), Embryonal tumors were the commonest (26.5%) followed by Diffuse astrocytic and oligodendroglial tumors (17.1%), Other astrocytic tumors (16.2%), Ependymal

tumors (11.1%), Tumors of the cranial and paraspinal nerves (7.7%), Choroid plexus tumors (5.9%), Meningiomas (4.3%), Mesenchymal nonmeningothelial tumors (3.4%), Craniopharyngiomas (2.6%), Neuronal and mixed neuronal glial tumors (2.6%), Tumors of the pineal region (1.7%) and Germ cell tumors (0.9%). Contrary to it in the second decade (11-18 years), category of Other astrocytic tumors [comprised of Pilocytic astrocytomas, Subependymal giant cell tumors (SEGA) and Anaplastic pleomorphic xanthoastrocytomas (PXA)] was the commonest (21.3%) followed by Embryonal tumors (16.3%), Diffuse astrocytic and oligodendroglial tumors (13.0%), Ependymal tumors (11.8), Tumors of the cranial and paraspinal nerves (8.9%), Craniopharyngiomas (6.7%), Meningiomas (6.2%), Neuronal and mixed neuronal glial tumors (4.5%), Mesenchymal nonmeningothelial tumors (4.5%), Lymphomas (2.2%), Choroid plexus tumors (1.7%), Germ cell tumors (1.7%), Tumors of the pineal region (0.6%) and Histiocytic tumors (0.6%) [Table 1].

There were 189 (64.1%) male cases and 106 (35.9%) female cases. In the first decade (0–10 years) there were 75 (64.1%) male cases and 42 (35.9%) female cases while in the second decade (11-18 years) there were 114 (64.1%) male cases and 64 (35.9%) female cases [Figure 1]. A male predominance was found in almost all the tumor categories, except for the Embryonal tumors and the Craniopharyngiomas where a slight female predominance was reported. Overall, the M: F ratio was 1.8:1. [Figure 2].

Clinical features

The most common clinical manifestations were symptoms and signs of increased intracranial pressure (headache, vomiting, seizures, visual disturbances and increased head circumference) manifesting in 240 cases (81.4%) followed by focal neurological signs (ataxia and weakness) manifesting in 55 cases (18.6%).

Tumor localization

Of the 295 patients, 103 patients (34.9%) had a tumor at the posterior fossa/cerebellum, the next common sites of involvement in descending order were cerebral hemispheres (70 patients; 23.7%), spine (42 patients; 14.2%), sellar/suprasellar (20 patients; 6.8%), lateral ventricles (11 patients; 3%), CP angle (09 patients; 3.1%), cerebral convexities (09 patients; 3.1%), 3rd ventricle (08 patients; 2.7%), 4th ventricle (08 patients; 2.7%), brainstem (05 patients; 1.7%), thalamus (04 patients; 1.4%), pineal region (02 patients; 0.7%), optic nerve (02 patients; 0.7%), cauda equina (01 patient; 0.3%), sylvian fissure (01 patient; 0.3%). [Figure 3]. Distribution of the tumors according to the original site of involvement is detailed in Table 2.

Histopathology

On comparing the histopathological subtypes of pediatric CNS tumors, it was found that the Embryonal tumors were the commonest [n 60, 20.3%]. Other astrocytic tumors including Pilocytic astrocytomas, SEGA, anaplastic PXA [n 57, 19.3%] comprised the second largest group. Diffuse astrocytic and oligodendroglial tumors [n 43, 14.6%] were the third most common tumors followed by Ependymal tumors [n 34, 11.5%], Tumors of the cranial and paraspinal nerves [n 25, 8.5%], Meningiomas [n 16, 5.4%], Craniopharyngiomas [n 15, 5.1%], Mesenchymal non-meningothelial tumors [n 12, 4.1%], Choroid plexus tumors [n 10, 3.4%], Neuronal and mixed neuronal-glia tumors [n 11, 3.7%], Lymphomas [n 4, 1.4%], Germ cell tumors [n 4, 1.4%], Tumors of the pineal region [n 3, 1.0%] and 16 Histiocytic tumors [n 1, 0.2%]; [Table 3, Figure 4]. In this study, 126 (42.7%) tumors were WHO grade I, 37 (12.5%) tumors were WHO grade II, 26 (8.8%) tumors were WHO grade III, 85 (28.8%) tumors were WHO grade IV. 21 (7.2%) tumors were not graded as these tumors have not been assigned any grade by WHO till date [Table 1].

Distribution of these pediatric tumors based on histopathology was compared with the published Indian literature (Table 4) and with the International literature (Table 5).

Discussion

The incidence of CNS tumors in children has been variable among different regions and countries. In India, according to Indian Council of Medical Research (ICMR), National Cancer Registry Data, incidence of childhood CNS tumors varies from 0-2.11%.⁷ The first multi-institutional study in India conducted by Jain et al in 2011 had collected the data from seven centres across the country in an attempt to represent the profile of the CNS pediatric tumors of the entire country. In their study, they found the incidence of pediatric CNS tumors ranging from 10-21%, with an average of 14.8%.⁸ The other single institution-based studies published by Rathi et al (2007), Bhalodia et al (2011), Shah et al (2015), Margam et al (2016), Madhavan et al (2016), Shirazi et al (2017) and Randale et al (2019) the incidence of pediatric CNS tumors was reported as 18.2%, 6.9%, 16.5%, 21%, 5.6%, 11.9% and 23.8% respectively.⁹⁻¹⁵ Jumaily et al (2019) in a single institution-based study from Iraq reported the incidence of pediatric CNS tumors as 14.8%.¹⁶ In our study, the incidence was found to be 19.4% showing concordance with majority of the existing Indian studies except for Bhalodia et al, Madhavan et al and Shirazi et al.

In contrast to the incidence, mean age of presentation has shown very little variation in different studies. Indian studies published by Mehta et al (2019) had reported it as 9.4 years, Shirazi et al (2017) had reported it as 10.5 years while Shah et al (2015) had reported the mean age of presentation as 10.7 years.^{17, 14, 11} In a study published by Gaidi et al (2012) from Egypt, mean age of presentation was reported as 7 years.¹⁸ In our study, the mean age of presentation was 11.5 years.

Male: female ratio had shown a male predominance in most of the studies published worldwide^{19-20, 14} but at the same time, some studies, albeit with smaller numbers of patients, have reported a slight female predominance.^{21, 22} In concordance to the existing literature, our study showed a male predominance (189 male cases compared to 106 female cases) with an overall M: F ratio of 1.8:1.

Clinically, regardless of the location, symptoms and signs of increased intracranial pressure (81.4%) followed by focal neurological signs (18.6%) were the leading clinical features in our study and the results were similar to the study published by Gaidi et al (2011) and Madhavan et al (2016), both these studies had reported clinical features related with increased intracranial pressure as commonest followed by focal neurological signs.^{18, 13}

Considering the location, in our study, posterior fossa/cerebellum was found to be the most common site followed by cerebral hemispheres, spine, sellar/suprasellar regions and ventricles. Each of these sites had a predilection for certain tumors, like Medulloblastomas and Pilocytic astrocytomas were the commonest tumors at posterior fossa/cerebellum. Similarly, Glioblastomas and Pilocytic astrocytomas were the commonest tumors involving the cerebral hemispheres. Benign peripheral nerve sheath tumors comprised of Schwannomas and Neurofibromas were the commonest tumors involving the spine. Craniopharyngiomas were the commonest tumors involving the sellar/suprasellar region while Ependymomas and Choroid plexus tumors were the commonest tumors found involving the lateral ventricles, 3rd ventricles and 4th ventricles. The results were almost consistent with the other studies published by Shah et al (2015), Katchy et al (2013), Rosemberg et al (2005) and Kaatsch et al (2001).^{1, 23-25}

Based on histopathology, our study has reported Embryonal

tumors as the most common tumor category comprised of Medulloblastomas, PNETs and ATRTs. Category of Other astrocytic tumors comprised of Pilocytic astrocytomas, SEGAs and pleomorphic PXAs was reported as the second most common. Third most common category of tumors was Diffuse astrocytic and oligodendroglial tumors comprised of Glioblastomas, Diffuse astrocytomas, Anaplastic astrocytomas, Oligoastrocytomas, Anaplastic oligoastrocytomas, Oligodendrogliomas, and Anaplastic oligodendrogliomas. Ependymal tumors were the fourth most common category comprised of Ependymoma and its variants. Fifth most common category was of Tumors of the cranial and paraspinal nerves comprised of Schwannomas and Neurofibromas. Meningiomas formed the sixth most common category followed by Craniopharyngiomas, Mesenchymal non-meningothelial tumors, Neuronal and mixed neuronal-glial tumors and Choroid plexus tumors, Lymphomas, Germ cell tumors, Tumors of the pineal region and Histiocytic tumors. The results of our study regarding distribution of tumors based on histopathology were in concordance with majority of other Indian studies published by Mehta et al (2019), Randale et al (2019) and Shah et al (2015).^{17, 15, 11} At the same time our results were found to be contrary to some of the Indian studies published by Madhavan et al (2016), Margam et al (2016) and Jain et al (2011) where astrocytomas were reported as the commonest category of pediatric tumors.^{13, 12, 8} Contrary to the previous Indian studies, our study has reported a higher incidence of Benign peripheral nerve sheath tumors and Meningiomas. Review of international data revealed that a good number of international studies had reported Astrocytomas and Medulloblastomas as the two most common CNS tumor categories in children. Our study has also reported these two as the commonest (Medulloblastomas followed by Astrocytic tumors), and Ependymomas as the third most common tumor category in concordance to a large meta-analysis done by Rickert and Paulus, where it was found that internationally, Ependymomas were the third most common tumors followed by Craniopharyngiomas occupying the fourth place.²⁶ The trend reported by Rickert and Paulus as depicted in Table 5 had shown similar data from Canada, HongKong, Germany, Sweden and Morocco,^{27-28, 23, 29-30} while data from India, Iraq, Egypt, Korea, Brazil, China and Japan had different results.^{8, 16, 18, 31, 1, 32-33} However, data from a single institute in Beijing, China, had reported Craniopharyngiomas as the second commonest tumor. [28] Rickert and Paulus had reported germ cell tumors to be the fifth most common type, but over a period of time it had been reported that the frequency of germ cell tumors varies markedly in different countries ranging from just 0.9% in Morocco to 14.3% in Japan.^{30, 33} In particular, all the three oriental Asian countries, Korea, China, and Japan, had shown higher frequency for Germ cell tumors and Craniopharyngiomas, suggesting environmental and/or genetic differences. Similarly, it is worth mentioning that though CNS lymphomas were very rare in majority of studies, including our study (1.4%), but CNS involvement by Burkitt lymphoma had constituted as high as 9.3% of intracranial tumors in a Nigerian study and 4.2% in an Iranian study.^{21, 34} This high prevalence of Burkitt lymphoma in tropical Africa has been attributed to the combined Epstein-Barr virus and Plasmodium falciparum malarial infection associated with other putative cofactors such as mosquito-borne arboviruses and plant tumor promoters.³⁵

Thus, to summarize there are considerable variations among different regions and countries regarding the current incidence of CNS tumors in children. Moreover, there are clear geographical and ethnic variations in the incidences of some specific pathological entities such as Germ cell tumors, Craniopharyngiomas and Lymphomas, which may be due to genetic or certain specific environmental risk factors and warrant further analytical epidemiological studies.

Multimodality approach including surgery, chemotherapy, and radiotherapy remains the mainstay in the management of childhood CNS tumors.³⁶

Conclusion

This study showed a male predominance with embryonal tumors being the most common tumor category along with posterior fossa and the cerebellum being the most common sites. Moreover, this study adds to the existing literature highlighting the histopathological and demographic profile of CNS tumors in children.

Table 1: Distribution of tumors by age group: 1st decade (0-10 years) and 2nd decade (11-18 years)

Histopathology	0-10 years	11-18 years	WHO grade
Other astrocytic tumors	(19)	(38)	I
Pilocytic astrocytoma	17	36	I
Subependymal giant cell astrocytoma (SEGA)	02	01	III
Anaplastic pleomorphic xanthoastrocytoma (PXA)	-	01	
Embryonal tumors	(31)	(29)	IV
Medulloblastoma	24	22	IV
Primitive neuroectodermal tumor (PNET)	05	05	IV
Atypical teratoid rhabdoid tumor (ATRT)	02	02	
Ependymal tumors	(13)	(21)	II
Ependymoma	05	14	III
Anaplastic ependymoma	08	07	
Diffuse astrocytic and oligodendroglial tumors	(20)	(23)	IV
Glioblastoma	11	12	II
Diffuse astrocytoma	05	06	III
Anaplastic astrocytoma	01	02	II
Oligoastrocytoma	01	01	III
Anaplastic oligoastrocytoma	01	-	II
Anaplastic oligodendroglioma	-	01	
Oligodendroglioma			
Tumors of the cranial and paraspinal nerves	(09)	(16)	I
Schwannoma	05	13	I
Neurofibroma	04	03	
Craniopharyngiomas	03	12	I
Meningiomas and its variants	05	11	I, II, III
Choroid plexus tumors	(07)	(03)	I
Choroid plexus papilloma (CPP)	06	02	II
Atypical CPP	01	01	
Mesenchymal non-meningothelial tumors	(04)	(08)	NA
Capillary haemangioma	02	01	
Ewing sarcoma	01	02	
Haemangioblastoma	-	02	
Lipoma	01	-	
Benign fibrous histiocytoma (BFH)	-	01	
Osteoma	-	01	
Chondrosarcoma	-	01	
Neuronal and mixed neuronal-glial tumors	(03)	(08)	I
Ganglioglioma	02	05	I
Dysembryoplastic neuroepithelial tumor (DNET)	01	01	I
Paraganglioma	-	01	II
Central Neurocytoma	-	01	
Lymphomas	-	(04)	
Diffuse large B cell lymphoma (DLBCL)	-	04	

Germ cell tumors	(01)	(03)	NA
Teratoma	01	02	
Yolk sac tumor	-	01	
Tumors of the pineal region	(02)	(01)	IV
Pineoblastoma	02	-	III
Pineal Parenchymal tumor of intermediate differentiation (PPID)	-	01	
Histiocytic tumors	-	(01)	NA
Langerhans cell histiocytosis		01	

Table 2: Distribution (frequency and percentage) of paediatric CNS tumors according to the primary site of involvement

Location	Number of cases	Tumors (In decreasing order of magnitude)
Posterior fossa/cerebellum	103 (34.9%)	Medulloblastoma (40), Pilocytic astrocytoma (34), Diffuse astrocytic and oligodendroglial tumors (8), Ependymoma (6), PNET (5), CPP (3), Hemangioblastoma (2), ATRT (2), Anaplastic pleomorphic xanthoastrocytoma (2), Ganglioglioma (1), Meningioma (1), Ewing sarcoma (1)
Cerebral hemispheres	70 (23.7%)	Glioblastoma (19), Pilocytic astrocytoma (8), Diffuse astrocytic and oligodendroglial tumors (14), Ependymoma (4), PNET (4), Anaplastic ependymoma (3), Ganglioglioma (3), DLBCL (3), Capillary hemangioma (3), DNET (2), Choroid plexus tumors (2), Medulloblastoma (1), Mature teratoma (1), ATRT (1), Craniopharyngioma (1), Meningioma (1)
Spine	42 (14.2%)	BPNST (18), Ependymoma (7), Meningiomas (4), Mesenchymal nonmeningothelial tumors (04), Anaplastic ependymoma (3), Pilocytic astrocytoma (3), Mature teratoma (1), DLBCL (1), Ganglioglioma (1)
Sellar/suprasellar	20 (6.8%)	Adamantinomatous craniopharyngioma (12), Pilocytic astrocytoma (4), Ganglioglioma (2), PNET (1), Pineoblastoma (1)
Lateral ventricle	11 (3.7%)	Choroid plexus tumors (04), Anaplastic ependymoma (2), SEGA (2), Tanyctic ependymoma (1), Transitional meningioma (1), Central neurocytoma (1)
CP angle	9 (3.1%)	BPNST (07), Meningiomas (2)
Cerebral convexities	9 (3.1%)	Meningioma (07), Benign fibrous histiocytoma (1), Langerhans cell histiocytosis (1), Ewing sarcoma (1)
3rd ventricle	8 (2.7%)	Anaplastic ependymoma (03), Craniopharyngiomas (2), SEGA (1), Choroid plexus papilloma (1), PPID (1)
4th ventricle	8 (2.7%)	Medulloblastoma (05), Pilocytic astrocytoma (2), Ependymoma (1)
Brainstem	5 (1.7%)	Diffuse fibrillary astrocytoma (02), Glioblastoma (1), ATRT (1), Anaplastic astrocytoma (1)

Thalamus	4 (1.4%)	Glioblastoma (03), Pilocytic astrocytoma (1)
Pineal region	2 (0.7%)	Pineoblastoma (01), Yolk sac Tumor (01)
Optic nerve	2 (0.7%)	Pilocytic astrocytoma (01), Neurofibroma (01)
Cauda equina	1 (0.3%)	Paraganglioma (01)
Sylvian fissure	1 (0.3%)	Meningioma (01)

PNET: Primitive neuroectodermal tumor, ATRT: Atypical teratoid rhabdoid tumor, DLBCL: Diffuse large B cell lymphoma, DNET: Dysembryoplastic neuroepithelial tumor, SEGA: Subependymal giant cell tumor, PPID: Pineal parenchymal tumor of intermediate differentiation

Table 3: Distribution (frequency and percentage) of paediatric CNS tumors based on histopathology

Tumor Category with frequency and percentage	Tumor type with frequency
Embryonal tumors [n60, 20.3%]	Medulloblastoma and its variants (46), PNET (10), ATRT (04)
Other astrocytic tumors [n57, 19.3%]	Pilocytic astrocytoma (53), SEGA (03), Anaplastic pleomorphic xanthoastrocytoma (01)
Diffuse astrocytic and oligodendroglial tumors [n43, 14.6%]	Glioblastoma (23), diffuse astrocytoma (11), anaplastic astrocytoma (03), oligoastrocytoma (02), anaplastic oligoastrocytoma (02), oligodendroglioma (01), anaplastic oligodendroglioma (01)
Ependymal tumors [n34, 11.5%]	Ependymoma (19), anaplastic ependymoma (15)
Tumors of the cranial and paraspinal nerves [n25, 8.5%]	Schwannoma (18), neurofibroma (07)
Meningiomas [n16, 5.4%]	Transitional meningiomas (10), rhabdoid meningiomas (03), fibrous meningioma (01), metaplastic meningioma (01), atypical meningioma (01)
Craniopharyngiomas [n15, 5.1%]	Adamantinomatous craniopharyngiomas (15)
Mesenchymal non-meningothelial tumors [n12, 4.1%]	Capillary haemangioma (03), Ewing sarcoma (03), Haemangioblastoma (02), BFH (01), lipoma (01), osteoma (01), chondrosarcoma (01)
Neuronal and mixed neuronal-glial tumors [n11, 3.7%]	Ganglioglioma (07), DNET (02), Paraganglioma (01), central neurocytoma (01)
Choroid plexus tumors [n10, 3.4%]	Choroid plexus papilloma (08), atypical choroid plexus papilloma (02)
Lymphomas [n4, 1.4%]	DLBCL (04)
Germ cell tumors [n4, 1.4%]	Mature teratomas (03), yolk sac tumor (01)
Tumors of the pineal region [n3, 1.0%]	Pineoblastomas (02), PPID (1)
Histiocytic tumors [n1, 0.3%]	Langerhans cell histiocytosis (01)

PNET: Primitive neuroectodermal tumor, ATRT: Atypical teratoid rhabdoid tumor, SEGA: Subependymal giant cell

tumor, BFH: Benign fibrous histiocytoma, DNET: large B cell lymphoma, PPID: Pineal parenchymal tumor of Dysembryoplastic neuroepithelial tumor, DLBCL: Diffuse intermediate differentiation.

Table 4: Incidence Of Paediatric CNS Tumors As Reported In Various Indian Studies

Tumor category	Mehta et al 2019 (n 242)	Randale et al 2019 (n179)	Madhavan et al 2016 (n 250)	Margan et al 2016 (n 239)	Shah et al 2015 (n76)	Jain et al 2011 (n 3936)	Our study 2022 (n295)
Embryonal tumors (Medulloblastomas, PNETs, ATRT)	33.5	24.5	21.6	18.4	29	22.4	20.3
Diffuse astrocytic and oligodendroglial tumors	24.8	13.9	58.8	56.8	25.0	35.8	14.6
Other astrocytic tumors (Pilocytic astrocytoma, SEGA, PXA)	14.9	22.9			22.3		19.3
Ependymomas	16.1	13.4	10.4	12.5	6.6	9.8	11.5
Craniopharyngiomas	5.0	7.8	3.6	9.2	11.8	10.2	5.1
BNPSTs (Schwannomas, Neurofibromas)	1.2	6.1	NA	2.9	2.6	3.6	8.5
Meningiomas	1.7	2.8	0.4	2.9	1.3	3.2	5.4
Choroid plexus tumors	0.4	5.5	0.4	0.8	2.6	1.8	3.4
Neuronal and mixed neuronal glial tumors	0.4	2.1	NA	1.7	1.3	2.4	3.7
Lymphomas	NA	NA	NA	0.8	NA	NA	1.4
Germ cell tumors	NA	1.6	1.6	1.3	NA	2	1.4
Tumors of pineal region	0.8	0.5	2.4	0.8	NA	1.3	1.0

PNETs: Primitive neuroectodermal tumors, ATRT: Atypical teratoid rhabdoid tumor, SEGA: Subependymal giant cell astrocytoma, PXA: Pleomorphic xanthoastrocytoma, BPNST: Benign peripheral nerve sheath tumor

Table 5: Frequency Of Various Types Of Paediatric CNS Tumors As Reported In Different Countries (In Percentage)

Tumor	Brazil	Canada	China	Egypt	Germany	Hong Kong	India	Iraq	Japan	Korea	Morocco	Sweden	Our study
Astrocytomas	32.5	39.4	30.5	35	41.7	57	34.7	31.5	35.7	27.8	37.1	51	33.9
Embryonal tumors (MBs, PNET, ATRT)	13.9	15.4	14.6	18.8	25.7	23	22.4	40.7	10	19.8	28.9	17	20.3
Ependymomas	7.4	7	5.6	10	10.4	8	9.8	3.7	4.8	8.1	12	8	11.5
BNPST	NA	3.1	2.8	0.7	NA	NA	3.6	0	0	0.4	NA	1.1	8.5
Meningiomas	3	< 2	3.1	2.4	1.2	NA	3.2	0	1.9	2.6	2.2	1.6	5.4
Craniopharyngiomas	11	6.8	18.4	11.3	4.4	6	10.2	0	10.5	9.2	6.6	4.6	5.1
Neuronal and mixed Neuronal glial tumors	7.6	< 2	3.1	1.3	3.2	NA	2.4	0	0	6.2	1.3	0	3.7
Choroid plexus tumors	3	2.3	1.8	2.4	NA	NA	1.8	1.9	0	2.2	NA	1.9	3.4
Germ cell tumors	3.6	3.1	7.9	2.4	NA	2	2	9.3	14.3	8.1	0.9	1.5	1.4
Tumors of the pineal region	NA	0.5	0.6	0.9	1.3	NA	1.3	1.9	0	NA	0.7	2.7	1

Mbs: Medulloblastomas, PNET: Primitive neuroectodermal tumor, ATRT: Atypical teratoid rhabdoid tumor, BPNST: Benign peripheral nerve sheath tumour

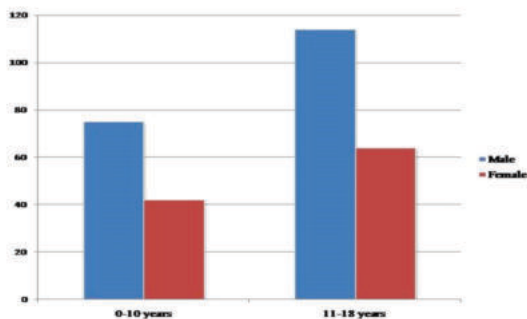


Figure 1: showing distribution of CNS paediatric tumors according to the age groups and gender

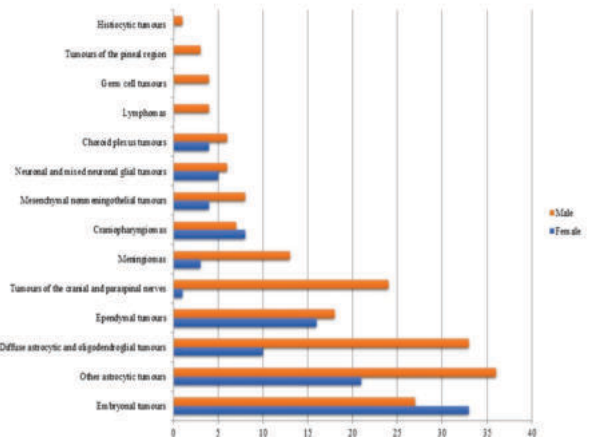


Figure 2: Showing The Male: Female (M: F) Ratio For Various Tumor Categories Of CNS Paediatric Tumors

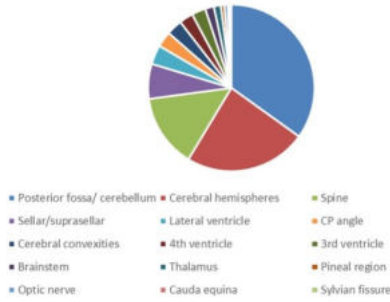


Figure 3: showing distribution of CNS paediatric tumors according to the location

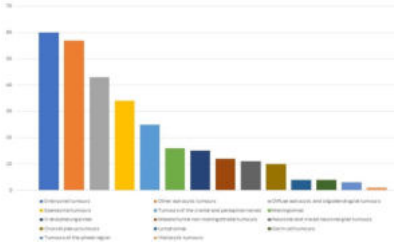


Figure 4: showing distribution of CNS paediatric tumors according to the histopathological types

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