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POSTERIOR FOSSA TUMORS: PANDORA'S BOX VIOLATED.

Surgery

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ABSTRACT

Objectives: Current study aims to use a retrospectively collected data registry of the surgically treated cases of the Posterior fossa lesions to clarify the relative frequencies of histological patterns in our setup over a period of 15-years.

Methods: Institutional patient database of 201-patients who underwent surgical intervention by the Department of Neurosurgery in a single referral center – King Hussein medical center-over a fifteen-year period from January 2002 to January 2017 scrutinized retrospectively. The clinical diagnosis was confirmed by radiological studies (CT scan, MRI…) and histopathological reports in all patients.

Results: Of total population, 104 were females and 97 were males. In our series, 61.7% of posterior fossa tumors were in children <14 years. Cerebellar symptoms were the most common manner of presentation followed by headaches and vomiting. All patients underwent surgical intervention employing posterior craniectomy with excision of tumor. Histopathological patterns identified were: Medulloblastoma reported in 82-cases, followed by Pilocytic astrocytoma 39-cases, while in descending order hemangioblastoma, Anaplastic ependymoma, Dermoid/ Epidermoid cyst, meningioma were recognized.

Conclusion: Posterior fossa tumor considered one of the most devastating forms of human illnesses, predominantly showing up in children with peak incidence in the first decade, clinical presentation is insidious. Advances in imaging modalities and treatment options have conveyed about a great improvement in survival in these patients over the past few decades, with recurrence rates very low due to a feasible gross total excision.

KEYWORDS

Posterior fossa; medulloblastoma; anaplastic astrocytoma; Choroid Plexus Papilloma.

Introduction:

Posterior fossa tumor is one of the most devastating forms of human illnesses. Cerebellum is the most common site of appearance of central nervous system tumors in children, reports state more than 60% of these tumors in children arising in the posterior fossa [1]. New progresses in imaging modalities and treatment options (particularly surgery and radiotherapy) have conveyed about a great improvement in survival in these patients over the past few decades [2-6]. Large multi-institutional reports illustrating the epidemiology and morbidity of posterior fossa tumor surgery are lacking [7]. Current study aims to use a retrospectively collected data registry of the surgically treated cases of the Posterior fossa lesions to delineate the relative frequencies of histological patterns in our setup over a period of 15-years. Special emphasis devoted to analyze the incidence, impact in such cases.

Methods:

This retrospective study was conducted under approval by the institutional ethical committee. Institutional patient database of 201-patients who underwent surgical intervention by the Department of Neurosurgery in a single referral center – King Hussein medical center-over a fifteen-year period from January 2002 to January 2017 scrutinized. The clinical diagnosis was confirmed by radiological studies (CT scan, MRI…) and histopathological reports in all patients. Patients with age ranging from neonate to 58 years were included in the study. Patients with posterior fossa metastasis were excluded.

Results:

Of the 201 total population, 104 were females and 97 were males giving a male: female of 1:1.1. In our series, 61.7% of posterior fossa tumors were in children <14 years with 23.9% in the <5 years, while 36.4% was in older children (5–15 years). Raised intracranial pressure was the cardinal clinical manifestation, headache and vomiting were the most common presenting symptoms with duration of symptoms ranged between 1-month and 16-months in extreme cases. Cerebellar symptoms were the most common manner of presentation followed by headaches and vomiting. Blindness was found in seven of our patients possibly due to long standing hydrocephalus. All patients underwent computed tomography (CT) and magnetic resonance imaging (MRI) [Figure. 1]. All patients underwent surgical intervention employing posterior craniectomy with excision of tumor. Pre-operative ventriculo-peritoneal shunt needed in few cases (in the case of obstructive hydrocephalus and raised intra-cranial pressure features) or in some cases post-operatively to the definitive tumor removal procedure. The majority did not need diversion procedure. In total, twenty eight patients underwent a second operation-resection during the follow-up within 1-68 months. Seven of these patients also had a third resection after further 1, 6, and 11 years of follow-up. The verdict for re-do surgery was based on clinical condition and/or progression on radiology images (MRI-based indications of increase tumor residual

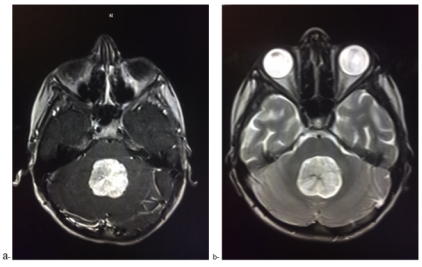

Figure. I: axial contrasted MRI(a) images showing typical frond like morphology with vivid homogenous enhancement(Choroid plexus papilloma), Axial T2WI (b) demonstrates a rather rounded multilobular fourth ventricular lesion, contains small cystic changes, appears isointense in T2WI.

Histopathological patterns identified were: Medulloblastoma reported in 82-cases [Figure. 2], followed by Pilocytic astrocytoma 39-cases, while in descending order hemangioblastoma, Anaplastic ependymoma, Dermoid/ Epidermoid cyst, meningioma were recognized [Figure. 3].

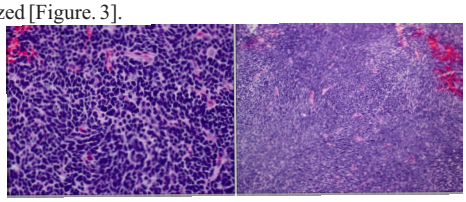

Fig. II: Histopathological examination of a resected specimen. Images show a Small blue round cell tumor with Syncytial arrangement of densely packed undifferentiated cells (embryonal cells) with high mitosis and apoptotic bodies

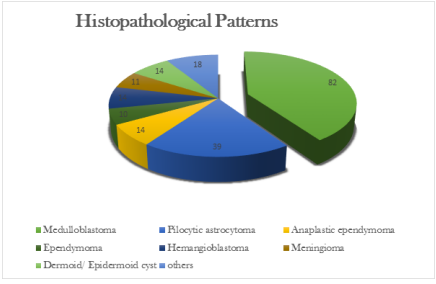

Fig. III: Diagram showing the histopathological patterns identified.

Discussion

Posterior fossa tumors are lethal conditions due to; vicinity and possible direct invasion or compression of the brainstem, on the other hand the tightness of the posterior compartment. The clinical presentation depends on the site of the tumor, biological behavior and aggressiveness of the tumor, and the rate of growth. Cerebellum is the most common site of appearance of central nervous system tumors in children, reports state more than 60% of these tumors in children arising in the posterior fossa [1].

New progresses in imaging modalities and treatment options (particularly surgery and radiotherapy) have conveyed about a great improvement in survival in these patients over the past few decades [2-6]. Large multi-institutional reports illustrating the epidemiology and morbidity of posterior fossa tumor surgery are lacking [7].

Clinical manifestations diverge from non- specific symptoms like headache and vomiting to more profound and serious presentations like lower cranial nerves palsies and ataxia due to direct involvement or compression [8]. In our review, Cerebellar symptoms were the most common manner of presentation followed by headaches and vomiting. Blindness was found in seven of our patients possibly due to long standing hydrocephalus.

In order to define the diagnosis based on medical history and physical exam, imaging tests obtained. All patients underwent computed tomography (CT) and magnetic resonance imaging (MRI). In our study these radiological studies were adequate in all cases to consider a surgical treatment.

Due to the limited pace of the posterior compartment, all lesions addressed utilizing surgical intervention, posterior craniectomy with excision of tumor executed. Pre-operative ventriculo-peritoneal shunt needed in few cases (in the case of obstructive hydrocephalus and raised intra-cranial pressure features) or in some cases post-operatively to the definitive tumor removal procedure especially before we introduced the cranial endoscope to perform endoscopic third ventriculostomy. The majority did not need diversion procedure [9]. In total, twenty eight patients underwent a second operation-resection during the follow-up within 1-68 months. Seven of these patients also had a third resection after further 1, 6, and 11 years of follow-up. The verdict for re-do surgery was based on clinical condition and/or progression on radiology images (MRI-based indications of increase tumor residual. In many series high incidence of recurrence ranging from 5% to 17% is evident, although long survival period is generally proved [10-12].

The histopathological approach is dramatically altering. Despite the fact that the former WHO classification of CNS tumors (2007) categorized the neoplasms based on histology, the most recent classification (2016) is conferring the combination of both histological and genetics conclusions [13, 14]. In our study a heterogeneity of pathological patterns identified due to the diversity of cells of origin. Medulloblastoma is typically a midline enhancing homogeneous posterior fossa mass on CT reported in 82-cases [Figure. 4], it is the most common malignant brain tumor in children and represents 30% of posterior fossa tumors. It is classified as grade-4 according to the WHO classification, and has a propensity to leptomeningeal dissemination. Almost 10% of cases are diagnosed in infancy. 75% occur in the midline; cerebellar location is associated with older age and desmoplastic histology [15, 16].

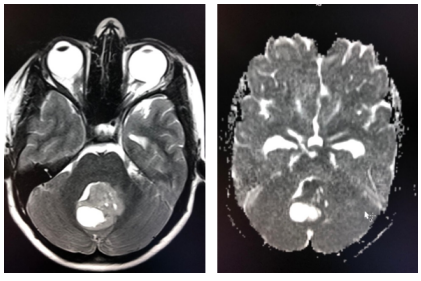

Fig. VI: Axial T2WI axial cut demonstrates a mass centered in the fourth ventricle, lesion shows heterogeneous intensity with a cystic component. ADC map shows hypo-intensity with low ADC value indicating restricted diffusion consistent with high cellularity.

According to several studies cerebellar astrocytomas reported as the most frequent posterior fossa tumors in children, accounting for up to 35% of these lesions [17]. They are circumscribed, discrete, slow-growing lesions, often associated with cysts within and around the tumor, in our review Pilocytic astrocytoma revealed in 39-cases [Figure. 5], while in descending order our analysis showed hemangioblastoma, Anaplastic ependymoma, Dermoid/ Epidermoid cyst, meningioma were recognized [Figure. 3].

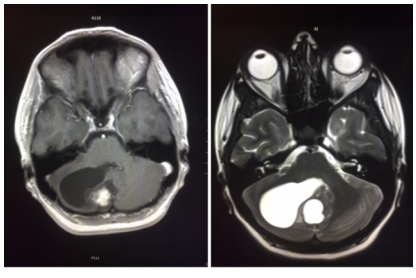

Fig. V: Axial T1WI post contrast shows vivid enhancement within the mural nodule. Axial T2WI shows a large cystic mass with solid mural nodule epicentered in the RT cerebellar hemisphere, displacing and effacing the 4th ventricle.

Posterior fossa is an area with dissimilar mass lesions harboring a divergent clinical and pathological manifestations.

Conclusion

Posterior fossa tumor considered one of the most devastating forms of human illnesses, predominantly showing up in children with peak incidence in the first decade, clinical presentation is insidious, with cerebellar mode and early morning vomiting. Advances in imaging modalities and treatment options have conveyed about a great improvement in survival in these patients over the past few decades, with recurrence rates very low due to a feasible gross total excision.

Limitations

This study has several limitations. First, a quantitative analysis could

not be established due to great heterogeneity in the included studies, different patient populations of different ages, multiple tumor subtypes.

Future Directions

We are planning more focused reviews on histological patterns and clinical forms. The diagnostic approach for brain tumors is dramatically changing.

Conflict of Interest Statement:

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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