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POSTERIOR CRANIAL FOSSA HEMANGIOBLASTOMAS: CLINICORADIOLOGICAL FEATURES, MANAGEMENT AND OUTCOME.

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Author's contribution :
MJ- Definition of intellectual content, Literature survey, Prepared first draft of manuscript, Implementation of study protocol, data collection, data analysis, manuscript preparation and submission of article, **KVK-** Concept, design , clinical protocol, manuscript preparation, editing and revision, statistical analysis, **SAvd-** preparation of tables, Coordination and manuscript revision, **SA-** Manuscript revision , **PS-** Manuscript revision

Conflicts of interest- nil

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Limitations of the study -
Patients with multi compartmental intracranial lesions, patients with spinal hemangioblastoms and patients who lost follow up are not included.

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Abstract
Background - Hemangioblastoms are benign tumors that primarily occur in the central nervous system (CNS). They account for approximately 1.5-2.5% of all brain tumors and 7-12% of infratentorial tumors in adults. The most frequent association of hemangioblastoms is with von Hippel-Lindau (VHL) syndrome, which is characterised by a chromosomal mutation on 3p. In cases of VHL syndrome, these tumors can be found in various locations both within and outside the CNS. On the other hand, sporadic solitary hemangioblastoms, although uncommon, are typically limited to the posterior cranial fossa. The primary treatment option for these tumors is complete surgical removal, making surgery the preferred and most effective approach.
Aims and objectives - The purpose of this study was to identify patients with solid and cystic posterior cranial fossa hemangioblastoms and explore the management challenges and outcomes associated with these tumors.
Material and methods - This retrospective study included 6 patients admitted with radiologically diagnosed cerebellar hemangioblastoms under department of neurosurgery , GRMC and associated J.A. group of Hospitals from May 2021 to April 2023 who underwent surgery. No randomisation done.
Results - The solid and cystic type of hemangioblastoma is typically the most prevalent form. Patients with this type often experience symptoms related to increased intracranial pressure (ICP). To address the tumors, all patients undergo complete removal through surgical resection. However, the occurrence of postoperative complications and mortality rates (16.6%) remains relatively high. This could be attributed to the limited literature available on adult posterior cranial fossa hemangioblastoms and the scarcity of surgical experience with such rare tumors.

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Conclusion- Overall, the prognosis and surgical outcomes for patients with hemangioblastomas are generally favourable. However, it should be noted that surgery for adult posterior cranial fossa hemangioblastomas is a demanding and challenging task. The complexity of the procedure, combined with the rarity of these tumors, contributes to the surgical challenges associated with their management.

Key words- Hemangioblastoms, Posterior cranial fossa, Cerebellar hemangioblastoms

INTRODUCTION:

Hemangioblastomas are benign tumors that primarily occur in the central nervous system (CNS). They account for approximately 1.5-2.5% of all brain tumors and 7-12% of infratentorial tumors in adults [(1)]. Hemangioblastomas can be classified as cystic, solid, or mixed type based on their content [(2)].

They are characterised by highly vascularised nodules within the tumour. The relative incidences of these morphological variants vary depending on the location, with cystic tumors more commonly found in the cerebellar hemispheres and solid tumors more frequently observed in the brain stem, cerebellar vermis, and spinal cord [(3)].

The spinal cord is the second most common location for hemangioblastomas, accounting for 2-3% of primary spinal cord neoplasms and 7-11% of spinal cord tumors [(4)]. However, occurrence in other locations, such as the supratentorial compartment, optic nerve, peripheral nerves, or soft tissues of extremities, is extremely rare [(5,6,7,8)].

Hemangioblastomas do not show a sex predilection and occur with roughly equal frequency in males and females [(9)]. They can manifest as solid or cystic tumors with varying amounts of solid components [(10)].

Clinical presentation of hemangioblastomas varies depending on the location and growth patterns. Cerebellar lesions often present with signs of cerebellar dysfunction, such as ataxia and coordination problems, or symptoms of increased intracranial pressure due to associated hydrocephalus caused by the expanding cyst [(11)].

The primary treatment approach for hemangioblastomas is complete surgical excision. Due to their highly vascular nature and proximity to vital structures in the brain stem and upper cervical spinal cord, complete removal may not always be possible, especially for larger tumors that are difficult to resect [(11)].

When managing these vascular tumors, two main aspects need to be considered: preventing spontaneous bleeding by complete embolisation when feasible and eliminating the space-occupying mass from the posterior cranial fossa either through surgery or radio-surgical shrinking. These approaches aim to preserve the functional integrity of important brain structures [(12)].

Gross examination of hemangioblastomas typically reveals a cherry red colour. The tumour may contain a cyst filled with clear or xanthochromic fluid. It grows within the parenchyma of the cerebellum, brain stem, or spinal cord, attached to the piamater, and receives a rich blood supply from the pial vessels. The cyst walls are non-neoplastic and consist of compressed cerebellar tissue.

MATERIAL AND METHODS:

This was a retrospective study on 6 patients who were admitted under department of neurosurgery, GRMC and associated J.A. group of Hospitals, Gwalior from May 2021 to April 2023 who underwent surgery and after analysing the patient's outcome the results were obtained.

A retrospective analysis was conducted on patients who underwent surgical intervention for the diagnosis of posterior fossa hemangioblastomas with solid and cystic components. Relevant data pertaining to patient demographics (age, sex), clinical manifestations (symptoms and signs), imaging results (Figure 1,2 and 3), and surgical findings were collected. The patients' status and progress were assessed at discharge and again at a follow-up period of 3 months post-discharge.

Clinical outcomes were classified based on the Glasgow Outcome Scale as either favourable (indicating good recovery or moderate disability) or unfavourable (representing severe disability, vegetative state, or death).

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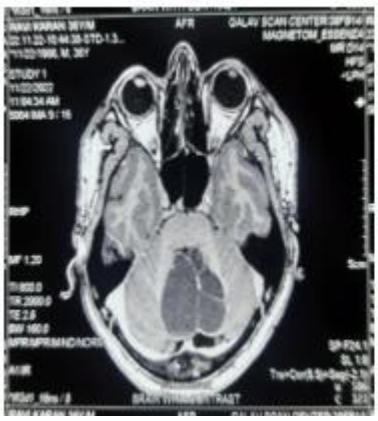

Fig. 1

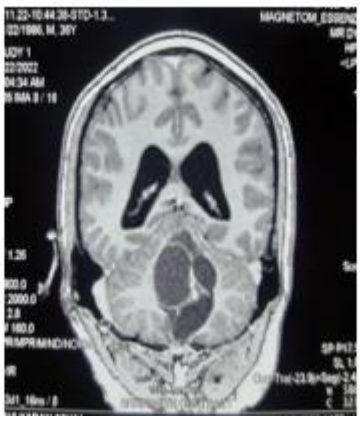

Fig. 2

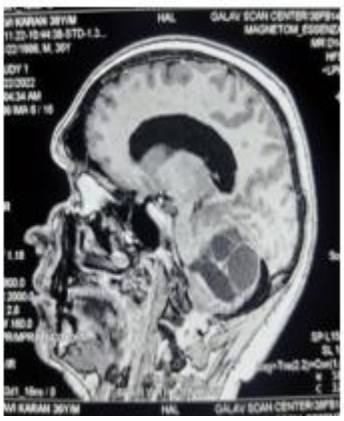

Fig. 3

Figure 1, 2 and 3 - MRI contrast images depicting axial, coronal, and sagittal cuts of a 36-year-old male patient with a posterior fossa hemangioblastoma.

Inclusion criteria :
All the patients with clinical and radiological diagnosis of posterior cranial fossa hemangioblastoma and underwent surgery.

Exclusion criteria :
Patients who did not underwent any surgical procedure, supratentorial or spinal hemangioblastomas and those not willing for participation.

RESULTS :
A non-randomised, retrospective, single institute study was conducted between May 2021 to April 2023, in GR Medical College and JA group of hospitals (GRMC), Gwalior over a period of two years in the department of Neurosurgery on 6 patients diagnosed with posterior cranial fossa hemangioblastoma. Based on the data collected and data analyses following observations were made.

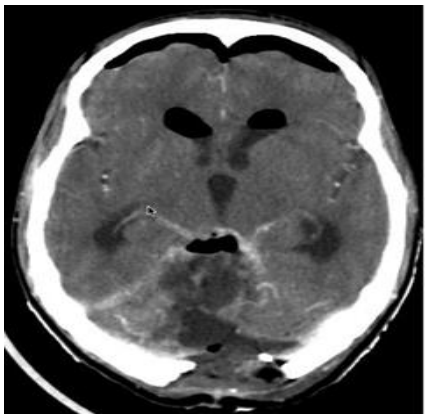

Fig. 4- Immediate Post operative plain CT head

In this study, a total of 6 patients were included, comprising 4 males and 2 females, ranging in age from 30 to 59 years. The most common symptoms and signs reported by the patients included headache (83.3% of cases), ataxia (66.6%), dysmetria (33.3%), and hydrocephalus (50%). The primary objective of the surgical intervention was to resect the hemangioblastoma, which was considered to be responsible for the observed symptoms.

Tumors associated with symptoms were found to be larger and more likely to exhibit peritumoral oedema or peritumoral cysts (100% of cases associated with oedema and cysts) compared to asymptomatic tumors. The majority of the tumors were located in the posterior half of the cerebellum (84%), with a smaller proportion found in the anterior half (16%).

At a follow-up period of three months after tumour resection, improvement in symptoms was observed. Preoperative hydrocephalus was resolved in 33% of cases following tumour removal, eliminating the need for cerebrospinal fluid diversion. However, 66% of cases required shunt surgery for the management of hydrocephalus. Notably, no genetic studies were conducted as part of this study.

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DISCUSSION:

The most common conventional MRI patterns of hemangioblastomas are traditionally described as enhanced mural nodules with adjacent non-enhanced surrounding cysts or enhanced solid lesions. Furthermore, hemangioblastomas commonly (60–69% of cases) have associated internal and/or peripheral signal flow voids, consistent with the pathologically identified dilated afferent and efferent vessels [(13)]. Traditionally, hemangioblastomas have been classified into four morphologic patterns based on macroscopic pathology. A cyst with a mural nodule type was the most common (60%), followed by a solid tumour type (26%) and a solid tumour with internal small cysts (9%), whereas a simple cyst type was the least common (5%).

In our current study, we observed that the most frequently reported symptoms and signs among the patients with cerebellar hemangioblastomas were headache, experienced by 83.3% of cases, followed by ataxia in 66.6% of cases, dysmetria in 33.3% of cases, and hydrocephalus in 50% of cases. Furthermore, we found that the majority of the tumors were located in the posterior half of the cerebellum, accounting for 84% of cases, while a smaller proportion was found in the anterior half, constituting 16% of cases. These findings provide insights into the clinical presentation and anatomical distribution of cerebellar hemangioblastomas in our study population.

Jagannathan et al [(14)] in his series of 80 patients of cerebellar hemangioblastomas in 2008 found that presenting symptoms were consisted with a mass in the posterior fossa and included the following: headache (75%), gait difficulties (55%), dysmetria (29%), hydrocephalus (28%), nausea/vomiting (28%), nystagmus (13%), dysarthria (9%), and/or dysphagia (8%). Lesions are preferentially distributed in the posterior half of the cerebellum, than they are in the brainstem and spinal cord, especially in cases of VHL disease. Also he reported that 70% of cerebellar lesions are cystic.

In a study conducted by Wanebo et al.[(15)] in 2003, which focused on 231 patients diagnosed with von Hippel-Lindau disease and a total of 650 hemangioblastomas, it was discovered that 72% of the symptomatic tumors were accompanied by cysts. Interestingly, in the majority of cases, the cyst size was larger than the corresponding tumour. The study also concluded that the development of signs and symptoms was primarily influenced by the rate of cyst growth and its overall volume. This indicates that the presence and characteristics of cysts play a significant role in the manifestation of clinical manifestations in patients with von Hippel-Lindau disease and associated hemangioblastomas.

In a retrospective analysis conducted by Rachinger et al.[(16)] in 2009, a total of 17 consecutive patients with 23 solid hemangioblastomas in the posterior fossa and spinal cord were examined. The study revealed that despite the frequent occurrence of these tumors in functionally important areas such as the brainstem and cerebellopontine angle , the clinical symptoms observed were rather unsystematic. Common symptoms reported by the patients included vertigo and headaches, with the exception of spinal cord lesions that typically presented with progressive quadriparesis. Notably, distinct cranial nerve symptoms were rarely observed, with only one patient demonstrating symptoms of vestibular nerve irritation. However, one patient with a hemangioblastoma in the medulla oblongata presented in a comatose state due to an acute hemorrhage. Apart from this particular case, distinct neurological deficits were infrequently observed for intracranial tumour locations. These findings suggest that the clinical presentation of hemangioblastomas in the posterior fossa and spinal cord can be variable and nonspecific, with symptoms such as vertigo and headaches being the most common, while distinct neurological deficits are less common except in cases involving the spinal cord.

In a report by Song and Lonser [(17)] in 2008, they presented four cases of posterior fossa hemangioblastomas that exhibited features of pathological satiety and atypical anorexia. All four lesions were localised to the obex region, which is a part of the medulla oblongata. Additionally, Lee et al.[(18)], in their seminal paper on Magnetic Resonance Imaging (MRI) of posterior fossa hemangioblastomas, classified them into four types. Type 1, which accounts for approximately 5% of posterior fossa hemangioblastomas, refers to a simple cyst without a macroscopic nodule. Type 2 represents a cyst with a mural nodule and is the most common type, constituting around 60% of cases. Type 3 corresponds to solid tumors (26% of cases), while type 4 refers to solid tumors with small internal cysts (9% of cases). These different types are observed in the cerebellum, with type 3 and type 4 tumors being more predominant in the spinal cord. Skull radiographs do not reveal any specific findings that suggest the presence of hemangioblastomas and therefore have limited diagnostic value in detecting these tumors. Instead, MRI is the preferred imaging modality for the evaluation and diagnosis of posterior fossa hemangioblastomas.

MRI with gadolinium enhancement is considered the best imaging study for screening and diagnosing hemangioblastomas, offering high sensitivity and specificity compared to CT and non-enhanced MRI. It allows for detailed visualization of the tumour and its characteristics. To achieve a high level of confidence in diagnosis, large-scale studies are often necessary. Intracranial hemangioblastomas can exhibit three distinct morphologic patterns based on macroscopic pathology. The cystic component surrounding the nodule appears hyperintense on T1-weighted images and hyperintense on T2-weighted images. The mural nodule, on the other hand, appears isointense on T1-weighted images and exhibits high signal intensity on T2-weighted images. Following the administration of gadolinium-based

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contrast medium, the nodule shows prominent enhancement, while the cystic portion does not enhance. MRI is particularly useful in differentiating between the cystic and solid components of the tumour and in delineating the extent of surrounding oedema. It provides valuable information regarding the tumour's location, size, and relationship with adjacent structures, aiding in accurate diagnosis and treatment planning[(13)].

The first successful surgical removal of a medullary hemangioblastoma was documented in 1936, predating the introduction of microneurosurgery techniques [(19)]. Over time, advancements in neuroimaging methods, refinements in microsurgical techniques, and the utilisation of preoperative immobilisation have contributed to improved outcomes and reduced morbidity and mortality associated with hemangioblastoma surgery. In general, long-term results of hemangioblastoma management have been favourable. However, local recurrences after complete tumour resection appear to be more common in patients with von Hippel-Lindau (VHL) disease, those diagnosed at a young age, and individuals with multiple hemangioblastomas. The recurrence rate can vary among different surgical series but typically remains below 25%. Recent studies have highlighted the correlation between histological subtype and the likelihood of hemangioblastoma recurrence. The cellular subtype demonstrates a higher recurrence rate of approximately 25%, while the reticular subtype exhibits a lower recurrence rate of approximately 8%. This finding suggests that the histological characteristics of the tumour may provide valuable prognostic information regarding the risk of recurrence. Overall, the combined efforts of improved imaging techniques, refined surgical approaches, and enhanced understanding of tumour characteristics have contributed to favourable long-term outcomes and reduced recurrence rates in the management of hemangioblastomas.

CONCLUSION

Cerebellar hemangioblastomas typically present with symptoms and signs that can be attributed to the presence of oedema and peritumoral cyst formation. The recommended and effective treatment for these cases is surgical resection. Complete removal of the tumour often alleviates the need for cerebrospinal fluid diversion in patients with preoperative hydrocephalus. It is worth noting that cerebellar hemangioblastomas have a preference for the posterior half of the cerebellum in their distribution. To prevent tumour recurrence, it is essential to perform meticulous extracapsular resection during the surgical procedure. If a planned staged treatment strategy is followed with a meticulous microsurgical technique, the long term outcomes can be as favourable as seen in small and cystic hemangioblastomas, even in eloquent locations.

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