SUCCESSFUL MANAGEMENT OF RUPTURED CAVERNOUS MALFORMATION OF THE PONS

Manejo exitoso de un cavernoma roto de la protuberancia cerebral

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ABSTRACT

Introduction: Brain stem cavernomas constitute 18-35% of intracranial cavernomas and have the highest bleeding rate of all brain cavernomas. Its annual rebleeding rate is 21 to 60%. Asymptomatic injuries should be treated conservatively, while symptomatic and accessible injuries surgery is recommended. Surgical resection prevents neurological deterioration caused by recurrent bleeding.
Clinical Case: We present the case of a 10-year-old female patient with headache, ataxic gait, left LV PC paresis, and vertigo. A magnetic resonance imaging of the brain (MRI) was carried out, where a ragged expansive process was evidenced in T2 in the posterior protuberance region with intra-tumoral and extra-tumoral hemosiderin deposits. Brain angiography was negative. A medial suboccipital craniotomy with a telovelar approach was performed, achieving total cavernoma resection. The patient was extubated on the 2nd postoperative day. Non-contrast brain tomography (CT) showed no acute complications, only minimal bleeding from the operative bed. Post-surgical brain MRI showed absence of the lesion. At 7 months, she continued without motor deficit, with adequate gait and Glasgow 15 points.
Conclusion: Brain stem cavernomas are a rare disease with high morbidity. Proper patient choice, surgical approach, and time to surgery decrease post-surgical morbidity.

Keywords: *Hemangioma, Cavernous, Central Nervous System, Brain Stem, Craniotomy (Source: MeSH NLM)*

RESUMEN

Introducción: Los cavernomas del tronco encefálico constituyen el 18-35% de los cavernomas intracraneales y son los que tienen la tasa más alta de sangrado de todos los cavernomas cerebrales. Su tasa anual de resangrado es del 21 a 60%. Las lesiones asintomáticas deben ser tratadas de manera conservadora, mientras que en lesiones sintomáticas y accesibles se recomienda la cirugía. La resección quirúrgica evita el deterioro neurológico causado por el sangrado recurrente.
Caso Clínico: Se presenta el caso de una paciente mujer de 10 años, con cefalea, marcha atáxica, paresia del VI PC izquierdo y vértigo. Se realizó una resonancia magnética cerebral (RMN) donde se evidenció en T2 un proceso expansivo arrosariado en región protuberancial posterior con depósitos de hemosiderina intratumoral y extratumoral. La angiografía cerebral fue negativa. Se realizó una craneotomía suboccipital medial con abordaje telovelar logrando la resección total del cavernoma. La paciente fue extubada en el 2° día postoperatorio. La tomografía cerebral (TAC) sin contraste no evidenció complicaciones agudas, solo mínimo sangrado de lecho operatorio. La RMN cerebral postquirúrgica mostró muestra ausencia de la lesión. A los 7 meses, continuaba sin déficit motor, con marcha adecuada y Glasgow 15 puntos.
Conclusión: Los cavernomas del tronco encefálico son una patología poco frecuente con alta morbilidad. La elección adecuada del paciente, el abordaje quirúrgico y el tiempo hasta la cirugía disminuyen la morbilidad posquirúrgica.

Palabras Clave*: Hemangioma Cavernoso del Sistema Nervioso Central, Tronco Encefálico, Craneotomía. (Fuente: DeCS Bireme)*

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Cavernomas have an incidence of 0.5% in the general population and constitute 5-10% of all intracranial vascular malformations. The prevalence varies between 4 to 35%. Cavernomas are made up of thin-walled dilated sinusoidal vascular channels covered by a simple endothelium and a fibrous adventitia.[2,6] 18-35% of intracranial cavernomas are in the brainstem, the protuberance being the most frequent location.[1] This type Cavernoma have the highest rate of bleeding of all intracranial cavernomas. [3]

Xie et al. mention that the risk of annual bleeding is 2.3 to 13.6% and that of rebleeding is 5-21.5%. It also mentions

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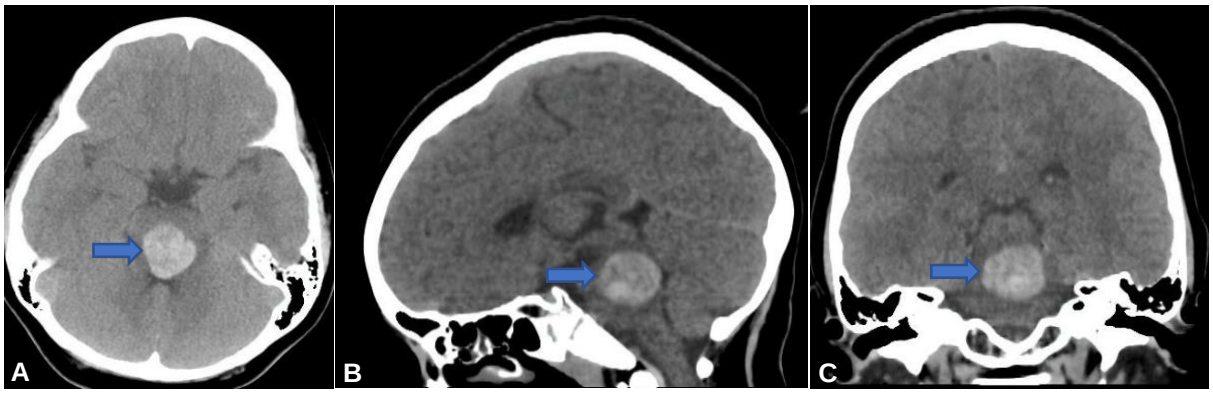

Fig 1. Brain CT without contrast showing a hyperdense lesion (arrow) in the posterior protuberance slightly lateralized to the left, compatible with acute bleeding. **(A)** Axial view. **(B)** Sagittal view. **(C)** Coronal view.

that the risk factor for rebleeding is a previous history of bleeding in a brainstem lesion, while considering as possible risk factors the female sex, being young, perilesional edema, large lesion, developmental venous anomaly, and hypertension arterial.[2] On the other hand, it is reported that after the first hemorrhage the incidence of rebleeding can be 21 to 60% per year, and after the 1st year the incidence of rebleeding is 8% per month. [1]

Lesions with a low risk of bleeding, asymptomatic or incidental injuries should be treated conservatively. [2] On the other hand, Gui et al. recommends that brain stem cavernoma resection should be carried out in symptomatic patients where the lesion is surgically accessible. Although new neurological deficits may appear postoperatively, these are usually transitory and resolve over time.

Surgical resection avoids neurological deterioration caused by repetitive bleeding.[1] The use of radiosurgery is controversial because studies mention that it reduces the risk of bleeding after the first 2 years, but not in aggressive lesions. Doses of 12 to 14 Gy is used. [2]

We present the case of a patient with a cavernoma of the cerebral protuberance treated surgically, with success in the Almenara Hospital, Lima, Peru.

CLINICAL CASE

History and examination: 10-year-old female patient, native and from Lima, with a medical history of asthma and allergic rhinitis. She refers a history of 13 days characterized by dizziness and gait disturbance, to which is subsequently added global and oppressive headache of intensity 4/10 on the visual analog scale (VAS). On physical examination: she was oriented, with a 15-point Glasgow Coma Scale (GCS), without meningeal or focalization signs, ataxic gait, left sixth nerve palsy and vertigo of central origin. The computerized axial tomography (CT) showed a hyperdense image of the posterior pons of 2.52 x 2.49 cm (PAxT), moderate perilesional edema, slight mass effect, and non-collapsed basal cisterns (*Figure 1*).

A cerebral angiography was performed which was negative for vascular lesion (*Figure 2*). Magnetic resonance imaging (MRI) with contrast showed a lesion compatible with a beaded expansive process in the posterior pontine region, with multiple intratumoral and extratumoral hemosiderin deposits, suggestive of a ruptured brain stem cavernoma (*Figure 3*)

Treatment: It was decided to perform a medial suboccipital craniotomy with a telovelar approach, achieving total resection of the ruptured cavernoma (in the 3rd week

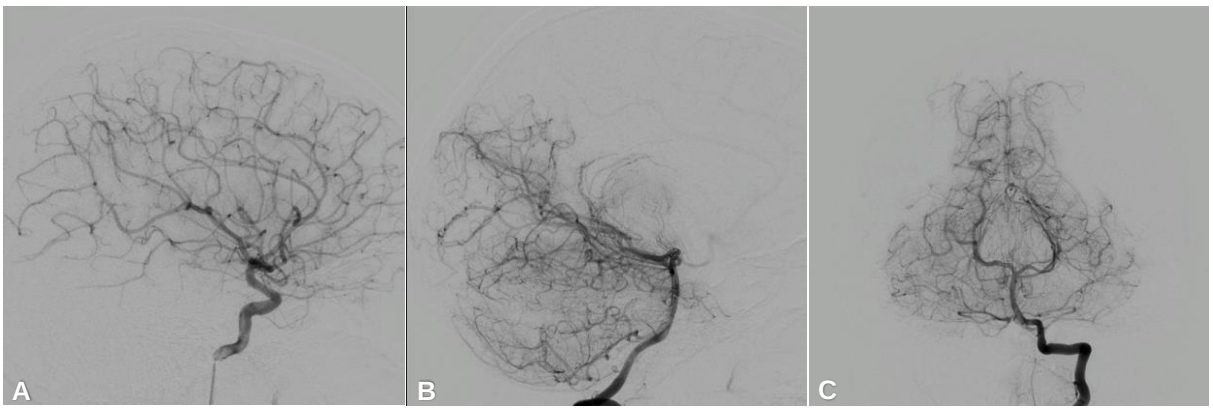

Fig 2. (A) Angiography of the left internal carotid artery in lateral incidence where no vascular pathology is evidenced. **(B)** Angiography of the left vertebral artery (LVA) in lateral incidence where no vascular pathology is evident. **(C)** Angiography of the left vertebral artery (LVA) in anteroposterior incidence where vascular pathology is not evident.

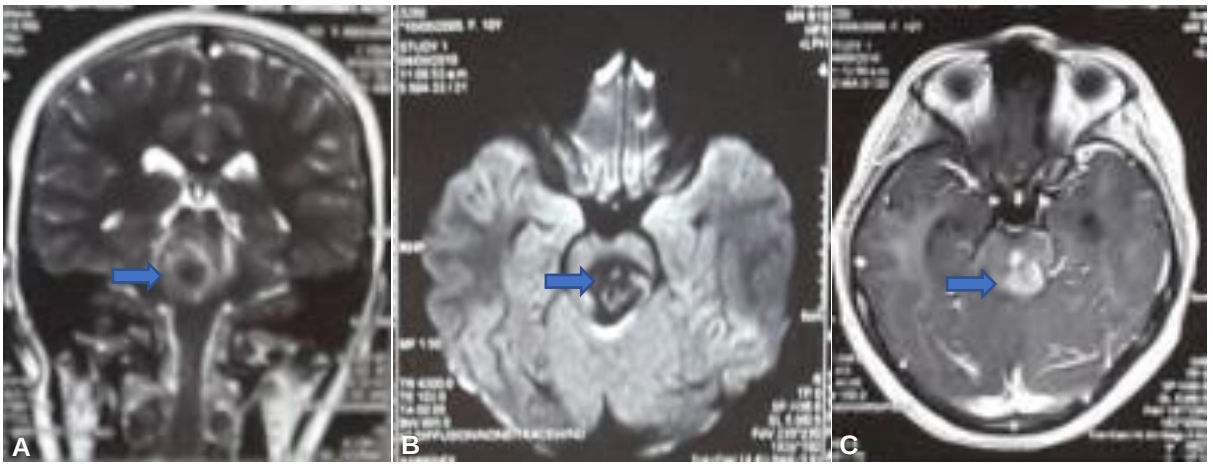

Fig 3. MRI of the brain shows a beaded lesion in a posterior pons slightly lateralized to the left (arrow) compatible with a cavernoma. (A) T2 sequence in coronal view that shows perilesional hemosiderin deposits, in relation to extralesional bleeding, with rupture of the capsule. **(B)** Flair sequence in axial view. **(C)** T1 sequence with contrast in axial view.

of the hemorrhage). She was transferred to the pediatric intensive care unit and was extubated on the 2nd postoperative day. Brain CT without contrast showed little bleeding in the operative bed and minimal edema, without the presence of acute complications. (*Figure 4*)

Clinical evolution: On the fourth postoperative day, the patient adopted the sitting position without major problems. On the fifth day she began ambulation with slight difficulty. On the sixth postoperative day, she was discharged, with GCS 15 points, left hemiparesis 4+/5, without meningeal signs, left sixth cranial nerve paresis of the same magnitude preoperatively, with an operative wound in good condition. Brain MRI with contrast at 7 months showed total resection of the cavernoma, without complications (*Figure 5*). At the clinical evaluation, the patient was oriented (GCS 15 points), without motor deficit, without sensory deficit, without oculomotor deficit, wandering normally.

DISCUSSION

Commonly seen brainstem lesions include cavernomas, gliomas, and hemangioblastomas. Among these lesions, cavernomas have a high incidence and are benign lesions that have an acute presentation with high morbidity. Cavernomas can be cured by surgical resection, but their location in the brainstem makes it difficult to decide the most appropriate surgical approach. An optimal surgical technique aims not only to resect the lesion, but also to minimize damage to the adjacent brain tissue, to improve the prognosis after resection. Neuronavigation and intraoperative neurophysiological monitoring improve the outcome of brain stem cavernoma surgery. [1]

Kearns et al in his study showed a mean presentation of 36.5 years for brain stem cavernomas, with a predominance of females, with the pons being the most frequent location (60%), followed by the midbrain (24.5%) and the bulb (15.8%) . In this study, he mentions that he had a complete resection in 92.3% of cases, with improvement of the neurological symptoms in 57.9% and with stable neurological symptoms in 25.9%. [7] Lashkarivand et al found in his study of 22 patients, a slight predominance of males, with a mean age of 58 years, total resection in all cases, without the presence of intraoperative complications except for one case of postoperative CSF fistula that had to be reoperated. [6]

Midbrain injuries are divided into ventral, lateral, and dorsal. Ventral lesions are lateral to the emergence of the III cranial nerve and medial to the pyramidal tracts, in these, the Transylvian approach is used. The lateral lesions are lateral to the midbrain sulcus, covered by the lateral mesencephalic vein, and for these, a transtentorial subtemporal approach is used. The dorsal lesions are two thin horizontal lines immediately above and below the quadrigeminal lamina, for which a supratentorial suboccipital approach is used. [2]

Pons lesions are divided into ventrolateral and dorsal. Ventrolateral ones in a peri-trigeminal area located medial to the V cranial nerve and lateral to the pyramidal tract, for which a subtemporal transtentorial, anterior transpetrous, retrosigmoid and presigmoid approach can be used. The dorsal ones are found in the median sulcus above the facial colliculus, the supra-facial triangle, and the infra-facial triangle, which can be treated by a midline posterior suboccipital approach. [2]

Bulbar lesions are also ventrolateral and dorsal in type. The ventrolateral ones are at the level of the retro-olivary groove or between the XII cranial nerve and C1 in the anterolateral groove and can be surgically approached by a far lateral approach. The dorsal ones are located in the posterior median fissure below the obex, in the posterior intermediate sulcus between the gracile and cuneiform fasciculus, in the posterior lateral sulcus between the cuneiform fascicle medially and the spino-trigeminal tract laterally, being treated by a posterior suboccipital midline approach. [2]

In 1994 Zabramski et al. presented a radiological classification of cerebral cavernomas, but this does not provide a degree of severity or a prediction of the postoperative result. This classification divides them into 4 types. Type I is subacute bleeding that is hyperintense on T1 and hypo- or hyperintense on T2. Type II is the typical "popcorn" lesion, being the most common, it represents bleeding in different phases, which is evidenced as a mixed

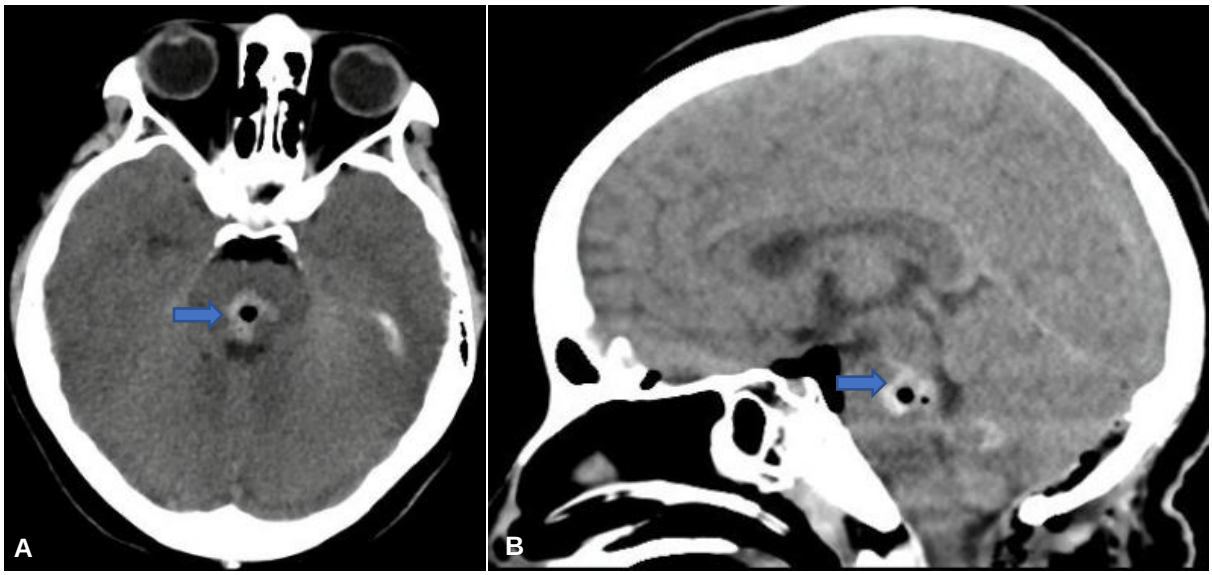

Fig 4. Brain tomography without contrast in the immediate postoperative period in **(A)** Axial view and **(B)** sagittal view, showing complete resection of the posterior pontine lesion with minimal bleeding on the operative bed (arrow) and mild perilesional edema.

signal centrally in T1 and T2, but in T2 a radiant hypointense halo is added. Type III is chronic hemorrhage seen on T1 as a centrally hypo to isointense lesion and on T2 as a centrally hypointense lesion with a radiant hypointense halo. Type IV is multiple stippled microbleeds that are difficult to identify on T1 and T2 but are "black dots" with irradiation on the echo gradient sequence. [6]

In 2015 Lawton et al. presented a more practical and clinical classification of brain stem cavernomas that includes the size of the lesion, if it crosses the midline in the axial plane, if there is an abnormality of venous development, age of the patient, bleeding time, which predicts the outcome postoperative. It is graduated from 0 to VII, having a good result from 0 to II and a high surgical risk in patients from V to VII. This classification is achieved with a sum of scores, the size is assessed and if it is 2cm or less it is scored as 0, if it is greater than 2cm it is scored as 1; It is also assessed if it crosses the midline in the axial plane, giving it 1 point; if there is a developmental anomaly, giving 1 point; age if he is 40 years old or less is awarded 0 points, and if he is older than 40 years is awarded 2 points; Finally, the bleeding time is evaluated, giving 0 points if it is acute (less than 3 weeks), 1 point if it is subacute (3 to 8 weeks) and if it is chronic (more than 8 weeks) 2 points. [2]

Al-Shahi et al. mentions that the definition of hemorrhage from a cavernoma is unclear because the authors do not mention that, as a requirement, the hemorrhage must be symptomatic or if the hemorrhage must extend outside the lesion. For this reason, it mentions that it requires the appearance of acute or subacute symptoms (headache, impaired consciousness, new neurological deficit or worsening of a previous one) plus radiological or pathological or surgical evidence or in cerebrospinal fluid of a recent intralesional or extralesional bleeding. [2]

Surgery should be performed between 3 and 4 weeks after the hemorrhage, because part of the hematoma can liquefy in this period of time and an area of gliosis forms around the lesion, which facilitates the isolated resection of the lesion of the peripheral brainstem tissue. In patients who undergo surgery between the 3rd and 6th month after bleeding, it has been observed that the hematoma solidifies and is firmly adhered to the tissue of the peripheral brainstem, which can lead to increased surgical injury. [1]

Brown et al. suggests the 2-point method to determine the optimal approach to brainstem injuries, where the first point represents the center of the injury and the second point represents the site where the injury is closest to the surface of the brainstem or the site considered as safest for

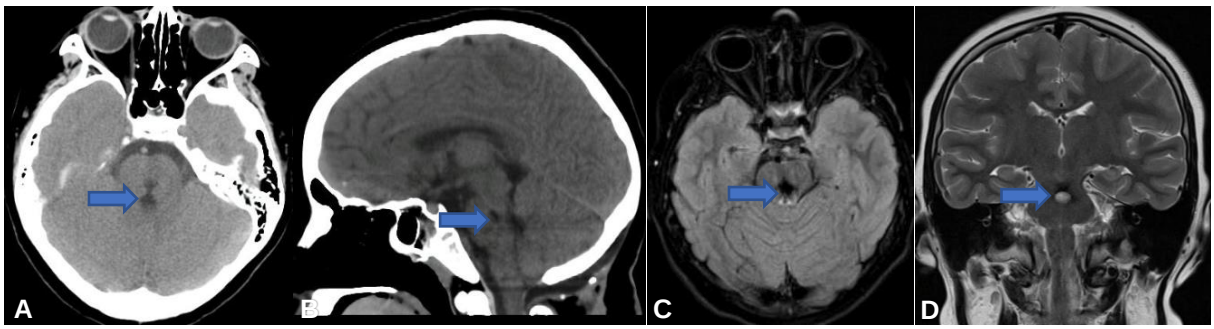

Fig 5. (A) Non-contrast cerebral tomography in axial view showing absence of the lesion (arrow) in the posterior pons. **(B)** Sagittal view of the brain CT without contrast showing the absence of the lesion (arrow). **(C)** MRI of the brain in flair sequence in axial view with absence of the lesion (arrow). **(D)** MRI of the brain in T2 sequence in coronal view with absence of lesion (arrow).

brainstem incision. By communicating these 2 points and directing it outwards, the craniotomy can be planned. [1]

For lesions located in the midbrain and pons, the lateral approach is suggested, since the midline suboccipital approach should be avoided because there are important nuclei in the mid-dorsal region of the brainstem. [1]

In this case, we present a patient with a ruptured pontine cavernoma, the most frequent of the brainstem according to the literature. achieving clinical improvement in the follow-up after 7 months. This result is similar to what is mentioned in other studies.

CONCLUSION

Brain stem cavernomas are a rare pathology but have high morbidity and high risk of rebleeding. The right choice of both the patient and the surgical approach reduces postsurgical morbidity. Surgery should be performed between the 3rd and 4th week to reduce intraoperative complications since the perilesional gliosis plane allows the lesion to be resected more easily.

REFERENCES

1. Gui S, Meng G, Xiao X, Wu Z, Zhang J. Surgical management of brainstem cavernous malformation: report of 67 patients. **World Neurosurg. 2019**; 122: e1162-e1171.
2. Xie MG, Li D, Guo FZ, Zhang LW, Zhang JT, Wu Z, et al. Brainstem cavernous malformations: surgical indications based on natural history and surgical outcomes. **World Neurosurg. 2018; 110**: 55-63.
3. Yeh YC, Wei KC, Chen KT. Transmastoid presigmoid retrolabyrinthine approach for removal of pontine cavernous malformation: how i do it. **Acta Neurochir (Wien). 2020; 162** (5): 1131-1135.
4. London D, Lieberman S, Tanweer O, Pacione D. Transclival approach for resection of a pontine cavernous malformation: 2-dimensional operative video. **Oper Neurosurg (Hagerstown). 2020**; pii: opaa025.
5. Abunimer AM, Lak AM, Aboul-Al-Shaar H, Patel NJ. Anterior petrosectomy for resection of brainstem cavernous malformation. **World Neurosurg. 2020; 135**: 1
6. Lashkarivand A, Ringstad G, Eide PK. Surgery for brainstem cavernous malformations: association between preoperative grade and postoperative quality of life**. Oper Neurosurg (Hagerstown). 2019**; pii: opz337.
7. Kearns KN, Chen CJ, Tvrdik P, Park MS, Kalani MYS. Outcomes of surgery for brainstem cavernous malformations: a systematic review. **Stroke. 2019; 50**(10): 2964-2966.
8. García R, Ivan M, Lawton M. Brainstem cavernous malformations: surgical results in 104 patients and a proposed grading system to predict neurological outcomes. **Neurosurgery. 2015; 76(3)**: 265-277.
9. Li D, Yang Y, Hao SY, Wang L, Tang J, Xiao XR, et al. Hemorrhage risk, surgical management, and functional outcome of brainstem cavernous malformations. **J Neurosurg. 2013; 119**: 996-1008.
10. Li D, Hao SY, Tang J, Xiao XR, Jia GJ, Wu Z, et al. Surgical management of pediatric brainstem cavernous malformations. **J Neurosurg Pediatr. 2014**; **13**: 484-502.
11. Gross BA, Batjer HH, Awad IA, Bendok BR, Du R. Brainstem cavernous malformations: 1390 surgical cases from the literature. **World Neurosurg. 2013; 80**: 89-93.
12. Kin T, Nakatomi H, Shojima M, Tanaka M, Ino K, Mori H, et al. A new strategic neurosurgical planning tool for brainstem cavernous malformations using interactive computer graphics with multimodal fusion images**. J Neurosurg. 2012; 117**: 78-88.
13. Kondziolka D, Lunsford LD, Kestle JR. The natural history of cerebral cavernous malformations. **J Neurosurg. 1995; 83**: 820-824.
14. Flemming KD, Link MJ, Christianson TJ, Brown RD Jr. Prospective hemorrhage risk of intracerebral cavernous malformations. **Neurology. 2012; 78**: 632-636.
15. Gross BA, Du R. Cerebral cavernous malformations: natural history and clinical management. **Expert Rev Neurother. 2015**; 15: 771-777.
16. Moriarity JL, Clatterbuck RE, Rigamonti D. The natural history of cavernous malformations. **Neurosurg Clin N Am. 1999; 10**(3): 411-417.
17. Batra S. Lin D, Recinos PF, Zhang J, Rigamonti D. Cavernous malformations: natural history, diagnosis, and treatment. **Nat Rev Neurol. 2009; 5**(12): 659-670.
18. Robinson JR, Awad IA, Little JR. Natural history of the cavernous angioma. **J Neurosurg. 1991; 75**(5):709-714.
19. Akers A, Al-Shahi Salman R, A Awad I, Dahlem K, Flemming K, Hart B, et al. Synopsis of guidelines for the clinical management of cerebral cavernous malformations: consensus recommendations based on systematic literature review by the angioma alliance scientific advisory board clinical experts panel. **Neurosurgery. 2017; 80**: 665-680.
20. Abla AA, Turner JD, Mitha AP, Lekovic G, Spetzler RF. Surgical approaches to brainstem cavernous malformations. **Neurosurg Focus. 2010; 29**: e8.

Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Authors Contributions

Conception and design: All authors. *Drafting the article:* Vargas. Critically revising the article: Palacios. *Reviewed submitted version of manuscript:* Vargas. *Approved the final version of the manuscript on behalf of all authors:* Vargas.

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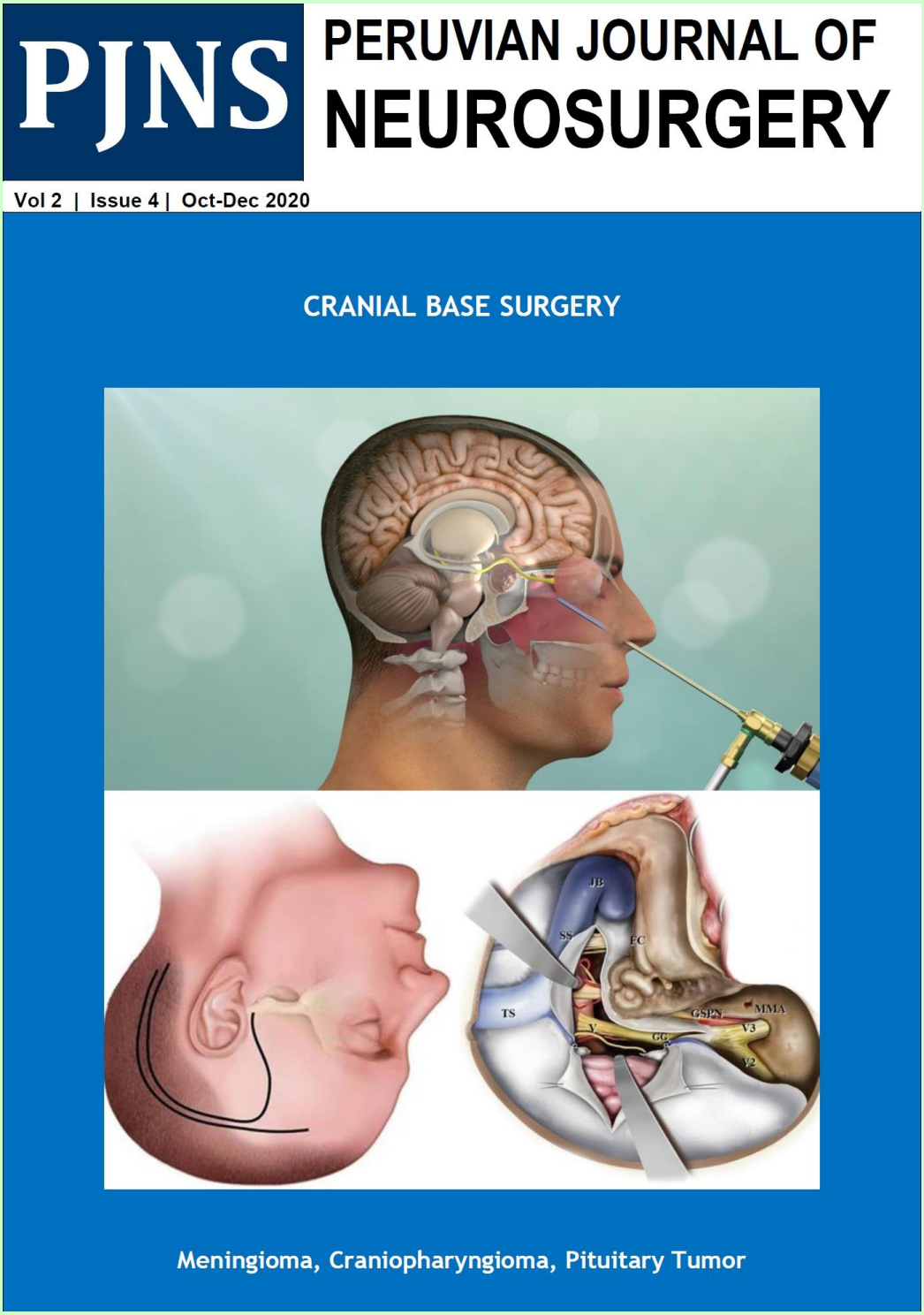
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