Neurology Case Series

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Dear Doctor,

Greetings from Micro Labs Limited.

We are happy to bring you "Neurology case series", which contains latest and interesting cases in the field of Neurology.

This issue contains

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Should you require any specific article or medical information, please feel free to contact us at medicalservices@microlabs.in

Looking forward to your valuable feedback

Best Regards,

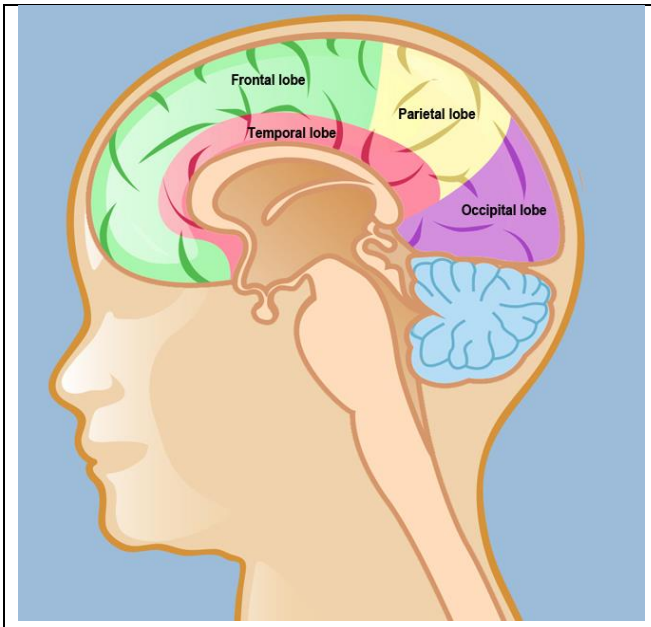
Medical Team,

1. Case Report on Gliomatosis Cerebri Presented as Parkinsonism Syndrome

Introduction:

Gliomatosis cerebri (GC) should be considered when parkinsonism is accompanied by other focal neurological disorders.

Gliomatosis cerebri (GC) is a rare neoplastic condition with no clear clinical pattern. The World Health Organization (WHO) defines it as a pattern of broad infiltrative growth that affects at least three cerebral lobes bilaterally and may extend to the brainstem, cerebellum, and spinal cord. The estimated incidence is 0.10 per million people, peaking at 0.43 per million after 65 years of age and males are more likely to be affected.[1] Further, the Glioma therapy and prognosis depends on the degree of malignancy. So, predicting histological grading before surgery is crucial for managing brain glioma and has a significant impact on survival rates. Conventional magnetic resonance imaging (MRI) can distinguish between high-grade and low-grade gliomas, but offers minimal insight into the tumor's biology.[2] A histological analysis confirms the suspected diagnosis, even though it is predicated on clinical presentation and neuroimaging results. In addition, patients with GC may exhibit neurocognitive and personality abnormalities, increasing headaches, and dementia-like symptoms.[1] This case report describes a 78-year-old patient who first developed Parkinson's illness and had GC with an atypical presentation.



Case Presentation:

In 2018, a 78-year-old Mexican woman with no medical history reported gait disturbances, including imbalance, slowness of walking, falls, and head tremors. One year later, she

experienced memory loss and periodic psychotic episodes. By 2022, her health had worsened to a walking handicap, rigid-akinetic syndrome, necessitating assistance for ambulation, and becoming wheelchair-bound. Non-motor symptoms included limited speech, impaired comprehension, emotional vulnerability, and dependence on everyday tasks. During that year, a brain MRI revealed widespread hyperintensity in the supratentorial white matter in T2/FLAIR sequences throughout all cerebral lobes (Figure 1). The T1 sequence showed no contrast enhancement, while the DWI sequence showed no diffusion restriction, excluding other plausible differential scenarios. CSF study showed slightly high protein levels of 56.6 mg/dL (normal range: 15-45 mg/dL), with normal cell count and glucose levels of 0/mm^3 and 59 mg/dL, respectively. Tests for KOH, bacterioscopy, culture, cytology, and JC virus in CSF were negative. Based on the clinical presentation, CSF tests, and radiological findings, GC was suspected.

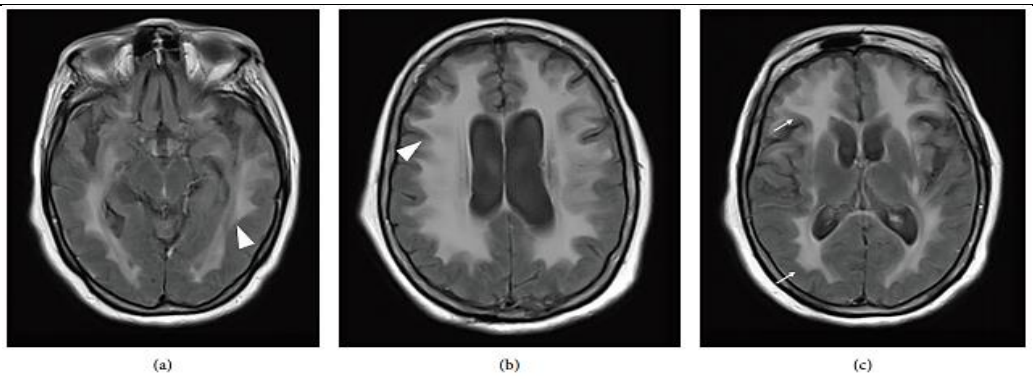


Figure 1: An MRI of the patient's brain in the T2/FLAIR sequence reveals broad white matter involvement. (a) A slice at the midbrain level reveals white matter involvement in the temporal lobes. (b) The lateral ventricle segment reveals hyperintensity that extends into the cortex. (c) A slice across the insula and thalamus reveals white matter involvement in the frontal and parietal lobes.

A neuroendoscopic biopsy confirmed GC with a pathology report of diffuse astrocytoma (Figure 2). GC was diagnosed in May 2022. Currently, the patient has moderate cognitive impairment, improved parkinsonism symptoms, but still requires assistance with ambulation and experiences sporadic neuropsychiatric alterations. Her treatment with levodopa-carbidopa, olanzapine, and escitalopram has resulted in positive outcomes.

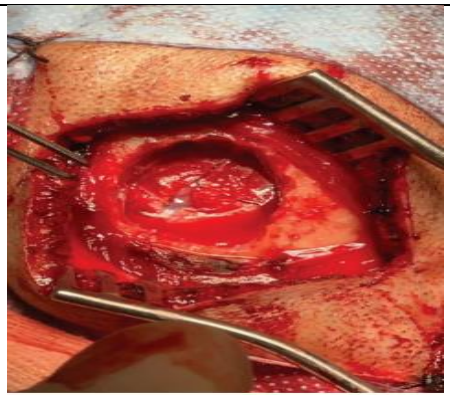


Figure 2: A frontal lobe biopsy was done via a craniotomy.

Discussion:

GC was initially defined as the excessive and diffuse development of neoplastic cells in both cerebral hemispheres. There is no established treatment protocol for GC, as it is non-resectable and surgery is only used for diagnosis. Radiation and chemotherapy are currently recommended for cancer treatment; however, the prognosis is generally poor. A histological analysis confirms the suspected diagnosis, even though it is predicated on clinical presentation and neuroimaging results.[1] A previous study showed that, in gliomatosis, amino acid PET can also be utilized to track the effectiveness of treatment. Additionally, using PET imaging with radiotracers may improve patient outcomes and lead to more tailored therapy by shedding light on the molecular mechanisms involved in the progression of gliomatosis and glioblastoma.[3]

Conclusion:

GC is a disease with nonspecific clinical symptoms, making clinical diagnosis difficult. It should be considered in cases of parkinsonism accompanied by other localized neurological diseases. This results in a delayed diagnosis and a low incidence. The importance of MRI as a diagnostic tool is highlighted, with a biopsy required to confirm the diagnosis.

Reference:

1. Garcia DG et al. Gliomatosis Cerebri Manifested as Parkinsonism Syndrome: A Case Report. Case Rep Neurol Med. 2024 Nov 28;2024:3375867.
2. Shakir TM et al. 1H-MR spectroscopy in grading of cerebral glioma: A new view point, MRS image quality assessment. Acta Radiol Open. 2022 Feb 26;11(2):20584601221077068.
3. Bonomi R et al. Multimodal neuroimaging of gliomatosis cerebri: a case series of four patients. Acta Radiol Open. 2020 Aug 21;9(8):2058460120942789.

2. Case Report on multilevel thoracic spinal epidural angiolipoma

Introduction:

Spinal epidural angiolipoma (SEAL) is an uncommon benign tumor that consists of vascular and fat elements. The incidence ranges between 0.04% and 1.2% for all spinal axis cancers and 2% to 3% for extradural spinal tumors.[1] These tumors are typically seen in the trunk and extremities' subcutaneous tissues. The dorsal mid-thoracic spine is the most prevalent location.[2] In this case report, a 52-year-old female came with back pain and paraparesis due to magnetic resonance (MR)-documented T7-T10 dorsal thoracic lesions. After a laminectomy for tumor excision, the patient was neurologically intact within 6 months.

Spinal epidural angiolipomas (SEAL) are a rare dorsal extradural spinal tumor that necessitates thorough complete resection.

Case Presentation:

A 52-year-old female reported with 3 months of back discomfort (Visual Analog Scale 5/10 and Karnofsky performance status 80/100) and a 1-month paraparesis (motor 3/5 proximal/distal) that worsened throughout the week. Thoracic MRI revealed a lesion around vertebrae T7-T10. T1-weighted MRI revealed a well-defined, homogeneous dorsal epidural compressive mass. T2-weighted imaging revealed strong intensity and distinctness from adjacent structures (Figure 1a-c). Spinal MR myelography indicated a full spinal block at T10 level (Figure 1d). The patient had a C-arm laminectomy from T7 to T9. A 6 × 2 × 0.5 cm dorsal extradural tumor with a reddish-yellow color and distinct borders from surrounding structures

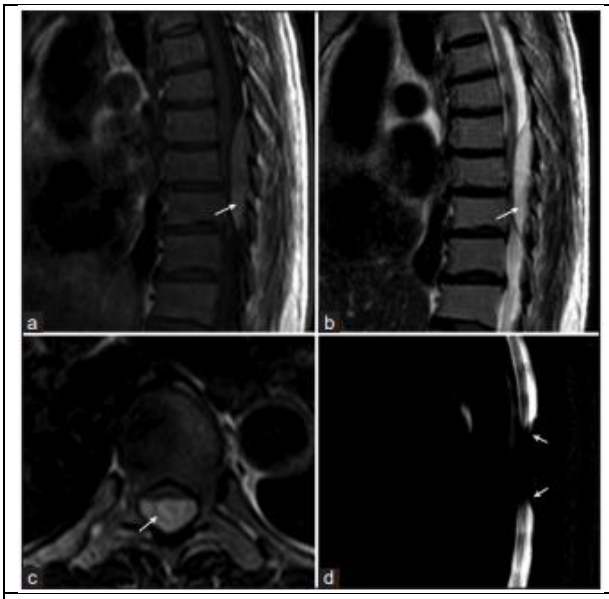


Figure 1: Magnetic resonance imaging reveals a focal mass at the dorsal extradural space at thoracic 7-10. (a and b) Coronal view with T1-weighted and T2-weighted images displaying the mass (white arrow). (c) Axial T2-weighted images reveal a dorsally positioned mass (white arrow). (d) Magnetic resonance myelography identifies the obstruction site (white arrow).

was found (Figure 2a). Microscopic inspection revealed a SEAL consisting of mature adipose tissue and abnormal blood arteries (Figure 2b and 2c). On the fourth day after surgery, the patient was discharged with 4/5 strength in the lower extremities. After 6 months, bilateral strength improved to 5/5.

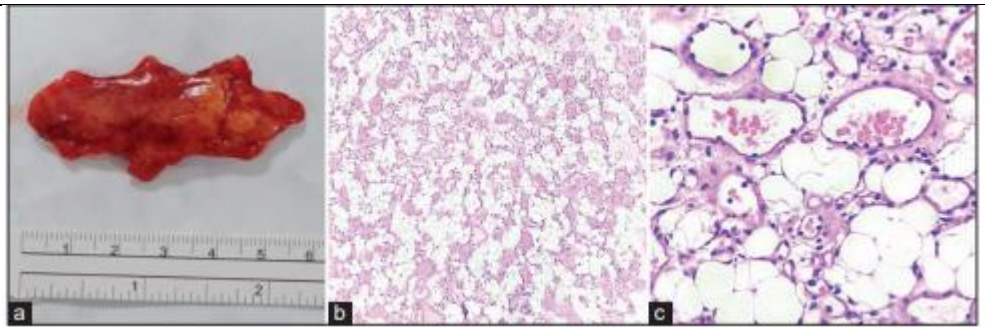


Figure 2: Histological results. (a) The bulk appears reddish-yellow when seen macroscopically. (b) A microscopic view reveals a mix of mature adipose cells and blood vessels (Hematoxylin and Eosin (HE) stain; ×100 magnification). (c) The vascular component has small, thin-walled arteries surrounded by mature adipose cells (HE stain; ×400 magnification).

Discussion:

SEAL tumors are rare and mostly found in the dorsal thoracic spine and more commonly diagnosed in women between 40 and 60 years old. In the present case, macroscopic view showing a reddish-yellow extradural tumor (6 × 2 × 0.5 cm) was found with distinct borders from surrounding structures.[2] A previous investigation found that SEALs are well-defined extradural masses that seem soft, reddish, and may be encapsulated on macroscopic examination. They can be easily separated from the dura mater.[3] The gold standard treatment for SEALs is gross complete resection, which can be performed using anterior or posterior surgical methods.[2] Because of SEALs possible high vascularity, the infiltrating type may need a more thorough excision and perhaps preoperative embolization.[4] In most documented cases, patients recover completely after complete tumor excision.[2]

Conclusion:

SEAL is an uncommon extradural dorsal thoracic spine tumor that can be easily detected with MRI. The gold standard of treatment is gross complete resection through laminectomy.

References:

1. Lee CH et al. Spinal epidural angiolipoma: a case report on sudden hemorrhagic onset and review of the literature. Journal of Korean Society of Geriatric Neurosurgery. 2021 Jun 15;17(1):20-4.
2. Wardhana DPW, Lauren C, Rosyidi RM. Multilevel thoracic spinal epidural angiolipoma. Surg Neurol Int. 2024 Jul 26;15:260.
3. Rkhami M et al. Epidural angiolipoma: A rare cause of spinal cord compression. Int J Surg Case Rep. 2018;45:72-6.
4. Maduri R et al. Spinal angiolipomas in pregnancy: Natural history and surgical treatment. Clin Neurol Neurosurg. 2019;178:25–30.

3. A rare case of Horner's Syndrome in Giant Cell Arteritis

Introduction:

Giant Cell Arteritis (GCA) is a polygenic immune-mediated disease that affects people over 50 years old and its cause remains unknown. Exaggerated immunological responses to vascular endothelial damage, including lymphocyte proliferation and giant cell development, are likely to be the cause. These responses cause luminal narrowing and disruption of the internal elastic lamina, resulting in reduced blood flow and end-organ ischemia. Common symptoms include headaches, scalp pain, jaw claudication, and vision loss.[1] The most severe complication is blindness caused by optic nerve ischemia. The American College of Rheumatology (ACR) has established diagnostic criteria for GCA based on the radiological and clinical criteria. GCA is a medical emergency that needs to be diagnosed and treated immediately due to the possibility of serious consequences. Diagnosis can be complicated due to overlapping symptoms with other common illnesses.[2] In this case report, an 82-year-old female with hypertension, atrial fibrillation, and arthritis reported experiencing right eye ptosis, blurred vision, dizziness, and throbbing eye pain within 24 hours.

In patients over 50 with Horner's syndrome and GCA symptoms, it is advised to measure inflammatory markers and obtain vessel wall imaging of the head and neck vasculature.

Case Presentation:

An 82-year-old female with a history of hypertensive and atrial fibrillation, taking dabigatran and aspirin, experienced right eye ptosis for 24 hours. Upon arrival, the patient's blood pressure was 162/91 mmHg, heart rate was 65 beats per minute, respiration rate was 20 breaths per minute, and she was stable. The patient experienced persistent drooping of her right eyelid, impaired vision, discomfort, and dizziness. Examination revealed right temporomandibular discomfort but no temporal scalp soreness, perceptible pulse, or stiffness of the superficial temporal artery. Her neurological exam showed slight right eye ptosis and miosis without anhidrosis, indicating incomplete Horner's syndrome. No other focal neurologic deficits were identified. The initial serum blood counts and metabolic panel were unremarkable. However, ESR was raised at 68 mm/h and CRP at 16 mg/L. CT angiography (CTA) revealed uneven thickness of the proximal major vessels, including the subclavian arteries, up to the carotid artery bifurcation, and the distal right internal carotid artery. The CTA head showed increased luminal constriction in the left middle cerebral artery MCA compared to the right one. MRI brain with contrast and MR angiography (MRA) with contrast vessel wall imaging revealed moderate stenosis over bilateral MCAs in the M1 segments (Figure 1) and vessel wall enhancement in the bilateral M1s, basilar artery, and petrosal extracranial internal carotid artery (Figure 2), indicating an inflammatory process. There was no evidence of acute ischemic stroke or hemorrhage. To address concerns about GCA, the patient was prescribed 60 mg of oral prednisone daily. An outpatient temporal artery biopsy revealed a rupture of the internal elastic lamina, multinucleated giant cells in the media, lymphocytic inflammation in perivascular soft tissue, and isolated medial calcifications, confirming the diagnosis of GCA. At a two-week follow-

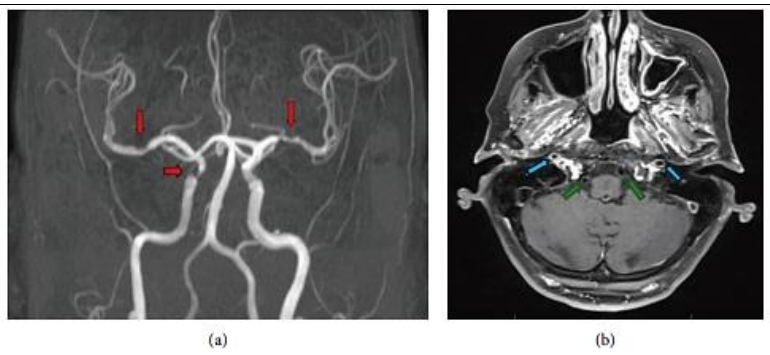


Figure 1: (a) MRA brain showing paraclinoid right internal carotid artery (horizontal arrow) and bilateral M1 segment (downward arrow) constriction. (b) Bilateral internal carotid arteries (blue arrows) and nonenhancing vertebral arteries (green arrows) show concentric enhancement on MRA brain imaging.

up, the patient reported improvement in impaired vision, ptosis, and jaw pain, as well as normalization of inflammatory markers. Later, the patient was started on weekly subcutaneous tocilizumab 162 mg. At the one-month follow-up visit, her Horner's syndrome was resolved.

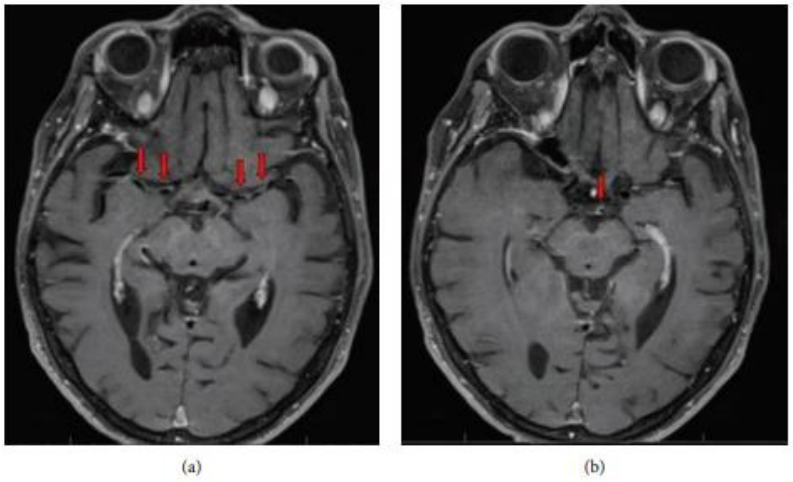


Figure 2: (a) Bilateral MCA vessel walls are enhanced in the MRI brain T1 post-contrast picture. (b) MRI brain T1 post-contrast picture shows enhancement of the basilar artery vessel wall.

Discussion:

Horner syndrome is a rare manifestation of GCA. Advancements in neuroimaging provide significant benefits for early diagnosis of vasculitis.[2] A previous report by Sverdlichenko et al. suggests that neuroradiological assessment of the entire sympathetic chain, in addition to a review of the relevant GCA history, symptomatology, and trending of inflammatory markers, is crucial for patients over 50 who present with new-onset Horner's syndrome.[3] This case study supports this assertion because the MRI brain of this patient showed mild to moderate stenosis and enhancement over the basilar artery and bilateral MCAs, indicating an inflammatory process. This study shows that MRI and MRA may accurately quantify artery supply in patients with GCA and Horner's syndrome.[2]

Conclusion:

For patients over 50 years with GCA symptoms and Horner's syndrome, it's recommended to assess inflammatory markers and perform vascular wall imaging of the head and neck vasculature. Early detection of GCA is crucial for initiating therapies and preventing long-term complications, including as blindness.

References:

1. Li KJ, Semenov D, Turk M, Pope J. A meta-analysis of the epidemiology of giant cell arteritis across time and space. Arthritis Res Ther. 2021 Mar 11;23(1):82.

2. Barr E, Levesque J, Badir J, Dunston R, Ranasinghe T. Unveiling the Uncommon: A Case Report of Horner's Syndrome as a Rare Glimpse Into Giant Cell Arteritis. Case Rep Neurol Med. 2024 Dec 10;2024:2503963.
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4. Association between bihemispheric posterior inferior cerebellar artery and vertebral artery stump syndrome

Introduction:

Posterior inferior cerebellar artery (PICA) changes should be considered while treating acute cases of bilateral cerebral infarction (CI) caused by VASS.

The carotid artery stump has been widely recognized as an embolic source for ischemic strokes. Vertebral artery stump syndrome (VASS) is an uncommon condition that can lead to an embolic stroke in the posterior circulation with a high recurrence rate.[1] It is potentially fatal and the best treatment for VASS has not yet been determined.[2] This is the first instance of VASS with a bihemispheric posterior inferior cerebellar artery (PICA), an uncommon type managed with endovascular therapy successfully.

Case Presentation:

A 63-year-old man with a history of hypertension presented to the Emergency Department with dizziness, vomiting, and gait disruption. Head magnetic resonance imaging (MRI) showed bilateral cerebellar hemispheres and vermis acute cerebral infarctions (CIs) [Figure 1a]. A poorly visible left V4 segment was seen by MR angiography [Figure 1b]. Upon admission, a conservative aspirin-based therapy was given. Digital subtraction angiography (DSA) of the left subclavian artery showed significant stenosis in the VA origin, along with

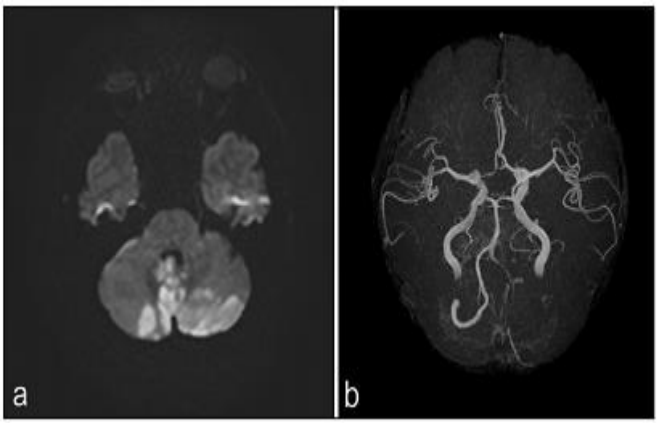

Figure 1: (a) Bilateral cerebellar infarctions were identified by initial head magnetic resonance imaging (MR). (b) The left V4 segment of the vertebral artery (VA) is not seen on the MR angiography.

collateral flow from the deep cervical artery to the V2 segment and a delayed anterograde flow in the VA. The V3 segment experienced a reduction in anterograde flow. With a to-and-flow lesion in the proximal section of the V4 segment, right VA angiography revealed retrograde flow from the right to the left V4 segment through the union. A bihemispheric PICA variation was discovered using three-dimensional rotational angiography. A diagnosis of bihemispheric PICA was made using VASS. The medication regimen was continued. The symptoms of dizziness and gait disturbance worsened on day 14 of hospitalization. An enlarged CI was observed in MRI. The left V4 segment was shortened, according to MR angiography. Poor left PICA condition and an extension of the to-and-flow lesion from the VA's V3 segment to the PICA orifice were found by repeated late-phase VA angiography. This study came to the conclusion that a stagnant flow that can result in the production of new thrombi was suggested by the difference between the VA as revealed by MR angiography and the late phase of VA angiography and used parent artery occlusion (PAO) to stop recurring embolisms and strokes.

A 6 Fr sheath introducer was placed into the right femoral artery while the patient was under general anesthesia. The appropriate VA received a 6 Fr ENVOY catheter. The VA close to the union was then provided with an intermediate catheter. A microwire (Figure 3a) was used to guide a microcatheter to the left VA's V3 segment. The V3 region was blocked by PAO using coils [Figure 3b]. Following treatment, DSA showed excellent PICA filling and retrograde flow from the right VA to the left V4 segment. Following treatment, symptoms did not worsen, and a head MRI on the next day revealed no new acute CIs [Figure 3c]. On day 32 of hospitalization, the patient received rehabilitation and was sent home with a modified Rankin scale score of 1.

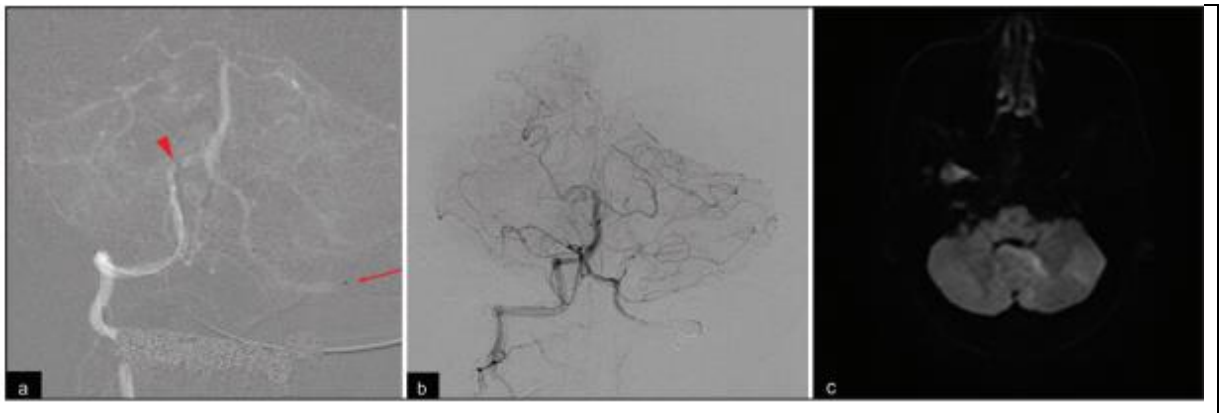

Figure 3: (a) The right V4 and left V3 segments were inserted with an intermediate catheter (arrowhead) and microcatheter (arrow), respectively. (b) The left extracranial vertebral artery underwent parent artery ligation using coils. (c) No new ischemic infarctions are detected by postoperative magnetic resonance imaging.

Discussion:

This is the first reported case of VASS-related bihemispheric PICA perfusion in the cerebellum. There is no standardized treatment for VASS.[1] A previous study by Suzuki et al. reported an antiplatelet therapy-treated patient with VASS.[3] Similarly, in this case, the etiology was artery-to-artery embolism caused by atherosclerotic alterations. Aspirin was initially prescribed as a conservative treatment. Furthermore, Medical care needs to be evaluated on a case-by-case basis.[1]

Conclusion:

This is the first case of VASS linked to a bihemispheric PICA that has been documented. Following endovascular PAO in the extracranial VA, no recurrent CI was noted. The potential role of PICA variants should be considered when treating patients who have acute bilateral CIs caused by VASS.

References:

1. Morimoto T et al. Vertebral artery stump syndrome associated with a bihemispheric posterior inferior cerebellar artery. Surg Neurol Int. 2024 Sep 27;15:350.
2. Katayama M et al. Vertebral artery stump syndrome: A 7-year follow-up case report. Radiol Case Rep. 2022 Jun 17;17(9):2923-2926.
3. Suzuki M et al. Vertebral Artery Stump Syndrome. Intern Med. 2018 Mar 1;57(5):733-736.