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

1. **Case report on Heidenhain variant of Creutzfeldt-Jakob disease.**
2. **Case report on Cerebrospinal fluid oculorrhea: a rare complication of orbital exenteration for cavernous sinus meningioma with orbital extension and radiation-induced hydrocephalus**
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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,

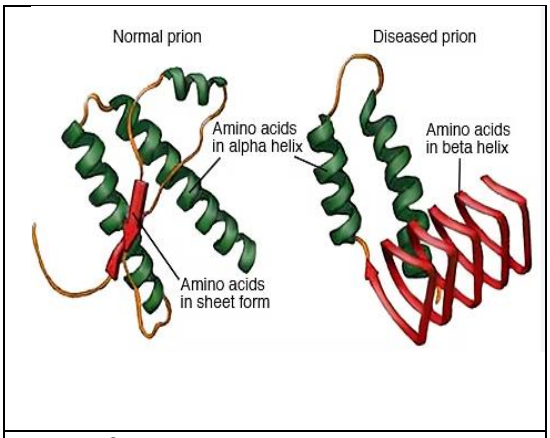
Medical Team,

1. Case report on Heidenhain variant of Creutzfeldt-Jakob disease

Introduction:

Since there is no treatment for CJD, palliative care and goal setting are critical for providing compassionate care to patients and their families.

Creutzfeldt-Jakob disease (CJD) is the most prevalent prions disease in humans. The normal prion protein (PrP^{c}) undergoes a conformational change to become the pathogenic protein (PrP^{sc}), which then aggregates in the central nervous system. Early symptoms are vague and can include headaches, anxiety, and behavioral changes. Diffuse neurological dysfunction later manifests as myoclonus, pyramidal and extrapyramidal impairments, cognitive decline, cerebellar ataxia, and visual abnormalities.[1] The prevalence of sporadic CJD (sCJD) is from one to two cases per million people. In the Heidenhain variant of sCJD (HvCJD), prions can cause visual disturbances in the occipital cortex, as evidenced by preferential diffusion restriction and T2-weighted FLAIR changes on MRI sequences.[2] This is a case of Heidenhain variant sCJD (HvCJD), which is likely to proceed quite rapidly.



Creutzfeldt-Jakob disease

Case Presentation:

A 72-year-old woman who had a medical history of melanoma, hypertension, hypothyroidism, and type 2 diabetes mellitus arrived as an outpatient transfer for additional neurologic decline management over three weeks. When the patient first arrived, they had visual abnormalities. Her husband was initially concerned when she struggled to sign on the appropriate line at an outpatient session. She started skipping lines when reading and had problems seeing objects within days. Her left hemibody weakness, visual hallucinations, paranoia, confusion, and short-term memory issues appeared a week later, along with a shaky, wide-based gait. Over the course of two more weeks, the patient's symptoms progressively got worse to the point that she was totally reliant on her husband for daily activities. The patient underwent extensive neurologic testing at two hospitals, including two contrast-enhanced MRI studies, a lumbar puncture for protein, cell count, glucose, meningitis polymerase chain reaction (PCR), cultures, Lyme disease, Rocky Mountain spotted fever, and an electroencephalogram (EEG).

The patient's workup was unremarkable except for past epileptic activity on EEG. She was prescribed lacosamide and then levetiracetam. Upon discharge, she was able to provide her history and maintain a conversation, but her family reported irritability and mood shifts.

Four days later, her husband took her to hospital for a second opinion because she was still declining despite taking anti-seizure drugs. The patient was not alert but easily arousable by voice and small stimuli. She had trouble focusing and paying attention, although she could obey basic instructions. Responses were restricted to single words and the speech was dysarthric. A cranial nerve exam identified a left lower face droop and thickening of the nasolabial fold. The left hemibody showed significantly higher muscle tone and rigidity than the right. Hyperreflexia (3+) was detected both diffusely and symmetrically. Although the patient struggled to complete strength, sensory, and coordination tests, no significant deficiencies were identified. Gait testing was delayed due to fall risk. The differential diagnosis included vascular insults, rapidly progressive degenerative processes (CJD, normal pressure hydrocephalus, and dementia variant), toxic/metabolic insults, central nervous system infection, and inflammatory processes based on the time course and symptoms. On Day 1, the initial workup of the metabolic panel, blood counts, urine, inflammation indicators, and thiamine level were not clinically significant. The serum autoimmune encephalopathy panel was negative. CSF tests revealed modestly increased protein at 54, glucose at 127, no nucleated cells, and negative results for autoimmune encephalopathy, fungal and bacterial

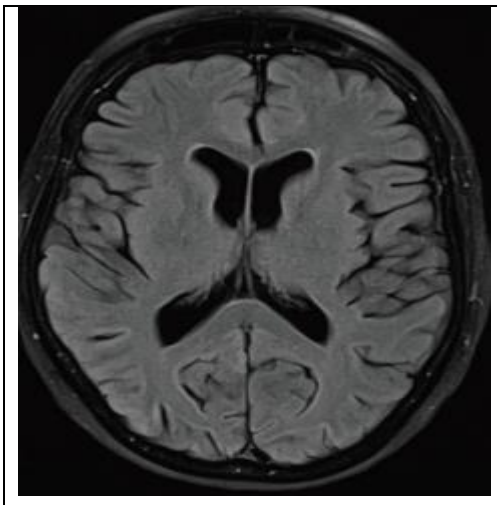


Figure 1. Magnetic resonance imaging of the patient's brain shows minimal FLAIR changes

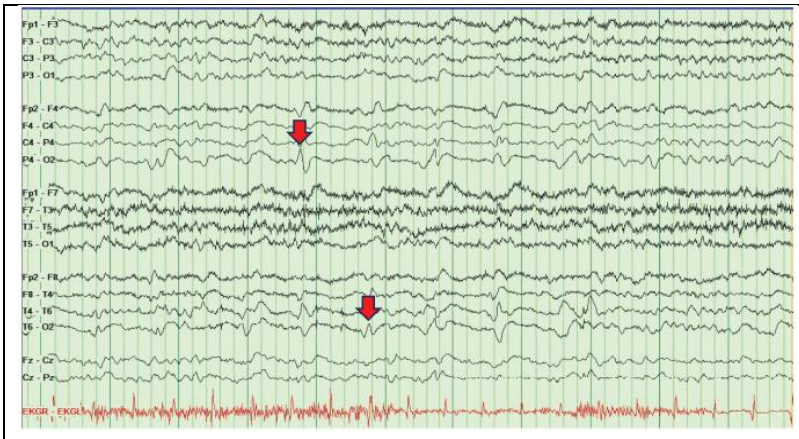


Figure 2. Electroencephalogram (EEG) completed around Week 4 of symptoms

cultures, meningitis PCR, and VDRL investigations. To test for CJD, a tube of CSF was delivered to the National Prion Disease Center on ice. Repeated MRI of the brain with and without contrast revealed gyriform cortical-restricted diffusion along the right occipital lobe, with patches of T2/FLAIR signal in the bilateral occipital lobes (Figure 1). Prolonged EEG revealed intermittent sharp wave complexes at 1 Hz in the right-sided posterior leads (Figure 2). These results led to a very high level of clinical suspicion for HvCJD. After consulting with the palliative care team on Day 2, the patient was moved to comfort care in accordance with the family's desires. The patient was discharged on Day 3. On Day 9, she died at her home. On Day 10, the patient's CSF sample showed positive RT-QuiC, increased T-tau protein, and 14-3-3 gamma, confirming the diagnosis of CJD.

Discussion:

The initial clinical symptoms of sCJD are generally unclear and unpredictable. In the present case, CSF investigations were shown to be highly sensitive in confirming the diagnosis of the condition. The CSF 14-3-3 and tau tests have low specificity (28% and 67%, respectively), whereas the RT-QuiC has a specificity of 99%-100%.[2] A previous study concluded that plasma Nfl and t-tau are effective indicators for the differential diagnosis of CJD. Plasma t-tau has also emerged as a potential predictive marker for disease duration.[3]

Conclusion:

As there is no cure for CJD, palliative care and goal setting are crucial for providing compassionate care to patients and their families. Discussion of supportive care interventions should begin even before confirmation with RT-QuiC due to high clinical suspicion.

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2. Case report on Cerebrospinal fluid oculorrhea: a rare complication of orbital exenteration for cavernous sinus meningioma with orbital extension and radiation-induced hydrocephalus

Introduction:

Cavernous sinus meningiomas (CSMs) are the most frequent primary cavernous sinus tumors with approximately 0.5 per 100,000 people in the general population. Their clinical appearance, specific to their site, frequently includes vision impairment, oculomotor disturbances, and face sensory alterations. Endocrine malfunction that necessitates long-term hormone replacement may also occur.[1] Radiotherapy is recommended for asymptomatic tumor growth, small-size tumors, and elderly patients due to the increased risk of postoperative complications. Although radiotherapy is commonly used to treat CSMs, it might lead to long-term complications such as hydrocephalus. Controlling cerebrospinal fluid (CSF) leakage in hydrocephalus patients is challenging and requires shunt treatment. CSF leakage from the orbit to the outside is referred to as "oculorrhea," as compared to otorrhea and rhinorrhea.[2] This is a rare case of right cavernous sinus meningioma with CSF oculorrhea that requires a lumboperitoneal shunt to treat hydrocephalus after tumor removal with orbital exenteration.

Radiotherapy can effectively treat cavernous sinus meningiomas in the short term, but long-term complications may include hydrocephalus.

Case Presentation:

A 71-year-old male patient with hypertension and diabetes mellitus reported a transitory visual field impairment as his primary complaint. The patient presented to the hospital with bilateral superior quadrantanopia. A physical examination revealed a 22 mm right CSM. A brain MRI showed no ventricular enlargement. After 10 months, an MRI showed an increase in size to 26 mm and expansion into the right sphenoid bone and optic nerve tract. A gamma knife surgery with a prescription dose of 12 Gy was done. After 13 months, the tumor size decreased to 23 mm. He experienced trouble opening his right eye after 5 years. MRI revealed a 32 mm tumor size, requiring stereotactic radiation (50.4 Gy/28 Fr). After gradual tumor growth, a second SRT (36 Gy/12 Fr) was conducted 8 years later to address right eyelid ptosis, mobility disturbance, tumor extension into the right orbit, and right sphenoid wing tumor enlargement. The tumor measured 50 mm in diameter. Nine years later, he developed right exophthalmos and lost his right sight. MRI revealed a significant rise in the

right intraorbital tumor and a little increase in the right sphenoid wing lesion, with a tumor diameter of 73 mm. In addition, the patient had gait apraxia due to communicating hydrocephalus. After the tap test, the patient's gait quality and speed improved. The CSF pressure was 25 cmH2O, and the protein concentration was 208 mg/dL, indicating radiotherapy-related spinal fluid malabsorption. To reduce the danger of infection, tumors were removed before hydrocephalus surgery. A month later, an orbitozygomatic craniotomy was performed to remove the tumor from the right orbit and sphenoid wings. Preoperative examination revealed a completely blind right eye, resulting in orbital exenteration. The tumor-adhering portion of the right middle skull base dura mater was also removed. An artificial dural substitute was used to cover the optic nerve canal for intraorbital repair. An osteoperiosteal flap was then applied to the intraorbital floor and secured with fibrin glue. The postoperative pathology revealed atypical meningioma. The intraorbital tumor was excised, leaving only the right cavernous sinus. However, there was postoperative CSF leakage in the orbit (Figure 1). The right eye experienced CSF leaking the day after surgery, which proved challenging to manage. After 7 days of treatment with eyelid sutures and spinal drainage, the patient's CSF leakage did not improve, therefore a closure was performed. During the initial procedure, an artificial dural substitute was used to correct the middle fossa dural defect and open the optic nerve canal. However, the substitute failed to adhere and resulted in leakage. The dural defective region was sutured as much as possible, and the intraorbital space was filled with fat and fascial graft, which was not done in the initial operation. The CSF leakage was eventually controlled, although it continued. Hydrocephalus was considered a possible source of CSF leaking. Eyelid sutures and a lumboperitoneal shunt were used to treat hydrocephalus, reducing CSF leaking and improving the condition. Two months following surgery, the patient was discharged home after rehabilitation. He continues to be monitored on an outpatient basis. After 16 months, there has been no return of CSF leaking following the first operation. The tumor did not spread to the opposite cavernous sinus, and the left visual field was retained. He died 20 months after primary surgery from sepsis caused by aspiration pneumonia, unrelated to complications from tumor removal and hydrocephalus surgery.

Discussion:

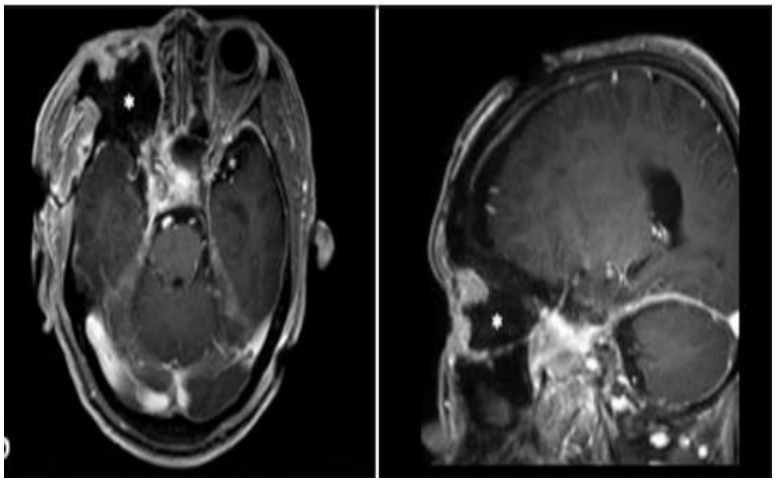

Figure 1. On improved T1-weighted images (left: axial, right: sagittal), all tumors were removed except those in the cavernous sinus. However, CSF retention was observed in the orbit (asterisk).

The patient underwent three rounds of radiotherapy for a right cavernous sinus meningioma, but the tumor continued to develop. Despite the patient's advanced age, a tumor resection was necessary to prevent total blindness from tumor progression. Hydrocephalus is a known radiotherapy complication.[2] In a previous study of 91 patients with atypical meningioma, one individual experienced hydrocephalus following radiotherapy, but symptoms resolved with a ventriculoperitoneal shunt.[3] In another study on cerebellar pontine angle tumors, 1.4% of meningiomas treated with radiation without resection developed hydrocephalus.[4]

Conclusion:

Radiotherapy can effectively treat cavernous sinus meningiomas in the short term, but long-term complications may include hydrocephalus. Hydrocephalus patients are at a significant risk of having intractable CSF leakage after tumor excision with orbital exenteration. Early surgical therapy for hydrocephalus may be important to prevent intractable CSF oculorrhea.

References:

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2. Ichinose T et al. Cerebrospinal fluid oculorrhea: A rare complication after orbital exenteration for cavernous sinus meningioma with orbital extension and radiation-induced hydrocephalus. Surgical Neurology International. 2025;16(9):1-6.
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3. Case report on Granulomatous Amoebic Encephalitis

Introduction:

A simple microscopic examination of CSF can sometimes detect amoeboid microorganisms. Early diagnosis and multi-agent treatment can improve prognosis and increase survival rates.

Granulomatous amebic encephalitis (GAE) is an uncommon central nervous system infection that is typically fatal with a mortality rate of over 90%.[1] GAE caused by the free-living amoeba Balamuthia mandrillaris was first identified in humans in 1990. The clinical presentation and neuroimaging in this illness are nonspecific, and B. mandrillaris is rarely found in conventional bacteriological testing for cerebrospinal fluid (CSF), making GAE diagnosis extremely challenging.[2] The most effective treatment for this infection has yet to be determined.[1] This case report presents a patient involved in an early CSF investigation that led to a rapid diagnosis.

Case Presentation:

A 48-year-old immunocompetent female with no medical history presented to the emergency department with the symptoms of stroke, including altered mental status (Glasgow Coma Scale (GCS) score of 12/15), slurred speech, severe proportional right hemiplegia, facial droop, and focal to bilateral Tonic-Clonic seizures. The patient had a high fever of 40°C, deteriorated overall health, and a history of headaches and coughing before experiencing neurological symptoms. The initial CT scan revealed no evidence of a stroke. However, Magnetic Resonance Imaging (MRI) indicated scattered nodular lesions above and below the tentorium (Figure 1). The patient was hospitalized in Neurology Department for additional investigation. Following admission, the patient experienced severe altered states of consciousness, with a GCS score of 9/15 and a prolonged high temperature of 39°C to 40°C. The initial CSF profile showed a leukocyte count of 17 cells/mm^3, with 80% lymphocytes, an

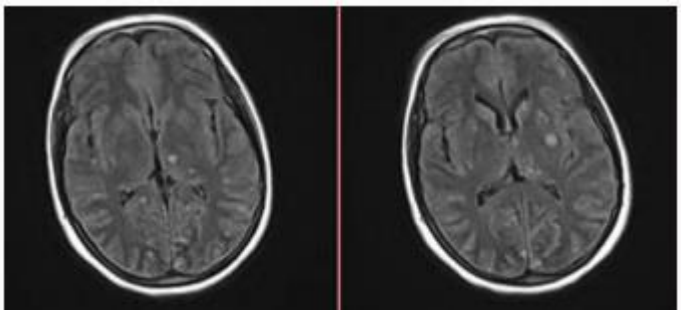

Figure 1: Gadolinium-enhancing nodular lesions in various cortical and sub-cortical locations are shown on T1 weighted brain MRI sections.

erythrocyte count of 778 cells/mm^3, a glucose content of 0.7 g/l, and a total protein concentration of 0.4 g/l. Gram staining and CSF culture yielded negative results. PCR tests for herpes simplex virus and India ink for Cryptococcus neoformans were also negative (Figure 2).

Brain MRI scans revealed several isointense nodular lesions on T1-weighted imaging and hyperintense lesions on T2-weighted imaging, with restricted diffusion and no ADC restriction. Some of the lesions showed ring enhancement and hemorrhage, with meningeal enhancement also present. An infectious origin was initially suspected. Five days later, a second lumber puncture was performed, and the CSF profile showed the following: erythrocyte count of 0 cells/mm^3, glucose concentration of 0.5 g/l, total protein concentration of 0.6 g/l, leukocyte count of 27 cells/mm^3 with 80% lymphocytes. CSF testing indicated amoeboid bacteria and cyst formation, indicating a telluric amoeba infection (Acanthamoeba sp./ Balamuthia mandrillaris). A Computed Tomography (CT) scan of the lungs was conducted based on the patient's coughing history to determine the route of entry and revealed alveolar consolidation and air bronchograms, indicating an infectious origin based on the clinical circumstances. The treatment plan included fluconazole and trimethoprim-sulfamethoxazole. The patient was admitted to the intensive care unit for 15 days before passing away from granulomatous amoebic encephalitis 5 weeks after the onset of neurological symptoms.

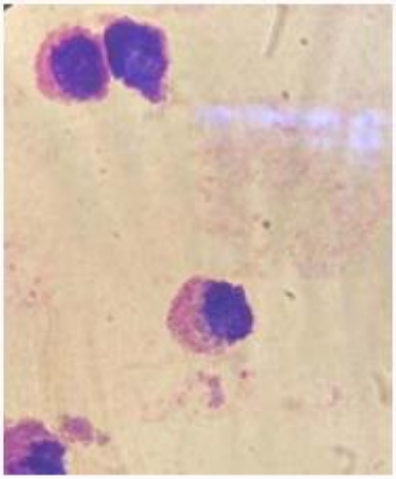

Figure 2. Microscopic analysis of CSF following MMG staining reveals amoeboid bacteria.

Discussion:

GAE is a rare fatal infection that affects the central nervous system, with a fatality rate of 97% to 98%. In the present case, heavy doses of Trimethoprim-sulfamethoxazole and Fluconazole were used to treat the patient. After being diagnosed, the patient remained comatose for 20 days until passing away. This case report suggests that a combination of fluconazole, trimethoprim-sulfamethoxazole, miltefosine, and rifampicin could be an effective treatment regimen.[1] A previous study showed that Trimethoprim-sulfamethoxazole is the most often used antibiotic, with 47% of patients surviving. Fluconazole was the most

often used azole in GAE treatment, resulting in a 40% survival rate. Ketoconazole had the highest effectiveness rate at 46%.[3]

Conclusion:

This case report shows the challenges that clinicians face when managing this condition due to its non-specific clinical and radiological presentation, as well as the lack of defined therapeutic guidelines following diagnosis. These findings highlight the critical need for precise diagnostic criteria and thorough treatment strategies to enhance patient outcomes.

References:

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4. Case report on Cardiac Myxoma mimicking Cerebral Vasculitis

Introduction:

Despite their benign nature, cardiac myxomas can cause significant morbidity and mortality in the general population, especially in young adults.

Cardiac myxoma, the most common kind of heart tumor, is typically considered benign due to its slow growth rate and lack of metastasis.[1] The World Health Organization (WHO) defines it as a neoplasm made up of mesenchymal cells that are cytologically bland, stellate to plump, and surrounded by a myxoid stroma.[2] Patients typically suffer from embolic consequences, which occur when tumor fragments or thrombi detach and travel through the bloodstream, resulting in serious and sometimes fatal diseases such as stroke, systemic embolization, or vital organ infarction.[1] Around 25% of all ischemic strokes are cardioembolic ischemic strokes, making them one of the world's major causes of morbidity and mortality. However, these strokes can be prevented by identifying their cause early and treating them quickly. Moreover, left atrial myxoma is uncommon.[2] For patients with cardiac myxoma, the only available treatment option at this time is surgical resection.[1] Cardiac myxoma has been associated with several neurological problems, including pseudovasculitis.[2] This case involves a 31-year-old man who had several ischemic strokes because of atrial myxoma. At first, it was believed that vasculitis was the cause of his previous strokes.

Case Presentation:

A 31-year-old man who had suffered an ischemic stroke nine months before presented to the facility complaining of extreme dizziness and unbalance along with a fever. Before his presentation, he experienced instances of blue discoloration on the tips of his fingers, which may have been caused by Raynaud's phenomenon and was linked to subjective fever episodes that went away on their own. The examination revealed no significant findings, except for an ataxic gait and a positive Romberg's test. His erythrocyte sedimentation rate (ESR) was slightly elevated to 54 mm/hr at the time of his initial presentation, and he tested positive for COVID-19 on the PCR test. Multiple bilateral cerebral, cerebellar, and brainstem areas of aberrant intensity were visible on his magnetic resonance imaging (MRI) scans. There were low/isointense signals at T1WI and high signals at FLAIR/T2-WI, with some of them displaying patchy postcontrast enhancement and restricted diffusion. At the time, there was a suspicion of cerebral vasculitis.

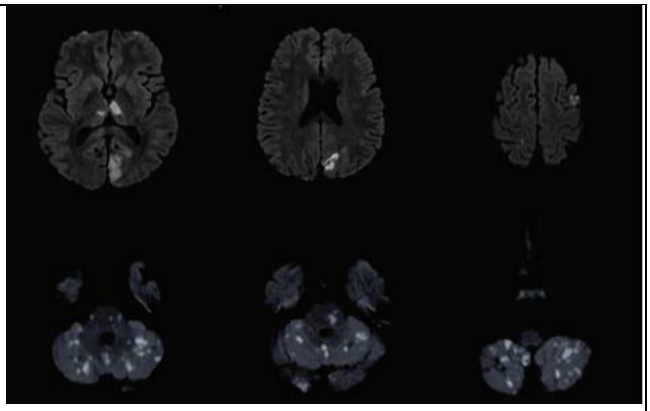

Figure 1. MRI diffusion-weighted scans indicate many distributed sites of diffusion limitation, consistent with acute ischemia.

The patient's documents and reports were gathered from their initial treatment in a non-tertiary facility. Nevertheless, the patient lost follow-up after discharge on ASA and atorvastatin along with outpatient physical therapy. Nine months after the first incident, he was discovered unconscious with vomit on his clothes and brought to emergency room. His neurological examination revealed moderate dysarthria, persistent hiccups, right homonymous hemianopia, restricted right eye movement in all directions, horizontal nystagmus on left eye abduction, right upper motor neuron facial nerve palsy, and dysmetria in both his upper and lower limbs. Examination results for rheumatology, pulmonary conditions, and cardiovascular disease were normal. An instantaneous computed tomography (CT) scan revealed a hypodense lesion in the right cerebellar lobe and left caudate head, which was associated with his previous infarctions. A lengthy segment blockage of the intracranial right vertebral artery was discovered during a brain CT angiography (CTA). His MRI brain revealed several dispersed regions of diffusion restriction in the posterior circulation territory, consistent with acute ischemia affecting the brainstem, thalami, cerebellum, and bilateral occipital lobes (Figure 1). Additional dispersed ischemic foci were observed in the frontal and parietal lobes, as well as the left corona radiata, which included the left caudate head. His lab results revealed a positive COVID-19 PCR test, a high procalcitonin level (9.51 ng/mL), and an elevated ESR level of 72 mm/hr. Immediate echocardiography revealed a thick tumor in the left atrium, extending from the mitral valve, indicating an atrial myxoma (Figure 2). A cardiac MRI revealed a left atrial tumor with two heads, the longest measuring 35 mm and projecting through the mitral valve into the left ventricle. The mass was linked to the interatrial septum via a stalk and shows no enhancement on delayed pictures, confirming the diagnosis of a left atrial myxoma. After a previous diagnosis of cerebral vasculitis at another center, a cerebral angiogram revealed normal arteries with no stenosis, occlusion, or aneurysmal dilatation in the left internal carotid, middle, anterior, and cortical arteries.

Minor irregularities were observed in the distal branches of the left MCA, perhaps due to embolic events. The patient's symptoms and study findings indicate that his clinical presentation is due to an atrial myxoma. The patient was sent to a nearby cardiac center and successfully underwent tumor excision without complications.

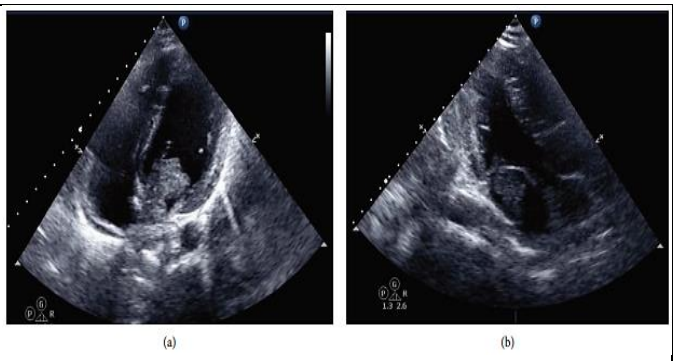

Figure 2. Atrial myxoma-like dense mass in the left atrium that is extremely mobile and protrudes from the mitral valve in the shape of a finger.

Discussion:

Diagnosing cardiac myxoma has been challenging due to its similarity to vasculitis, particularly in the absence of cardiac symptoms. The most effective treatment for atrial myxomas is the surgical removal of the tumor, which can be curative. Anticoagulation therapy can be used as a bridge therapy before surgery or as secondary prophylaxis after surgery.[2] Corticosteroid treatment was commonly employed in early similar cases in which vasculitis was the presumptive diagnosis.[3] The patient in this instance did not show any symptoms of vasculitis during his current presentation, and all of his serological markers came back negative during his current presentation, therefore corticosteroid therapy was not considered.[2]

Conclusion:

In the general population, cardiac myxomas can cause significant morbidity and mortality, especially in young adults, despite their benign nature. Tumors are regarded as an emergency and require immediate surgical resection, which is usually curative.

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