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*Case report*

# SUSPECTED RHOMBENCEPHALITIS WITH VENTRICULITIS IN AN ELDERLY PATIENT WITH PREVIOUS MENINGITIS AND VENTRICULOPERITONEAL SHUNT: A CASE REPORT

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## Abstract

Rhombencephalitis is a rare inflammatory disorder involving the brainstem and cerebellum, with a broad differential diagnosis that includes infectious, autoimmune, paraneoplastic, and post-infectious causes. Diagnostic evaluation may be particularly challenging in elderly patients with previous central nervous system pathology and cerebrospinal fluid diversion devices.

We report the case of a 75-year-old man admitted with progressive neurological deterioration manifested by vertigo, gait instability, somnolence, confusion, and dysphagia. Brain magnetic resonance imaging revealed inflammatory changes involving the medulla oblongata and cerebellar peduncle, accompanied by ependymal and ventricular wall enhancement consistent with rhombencephalitis and ventriculitis. Cerebrospinal fluid analysis demonstrated elevated protein levels and blood–cerebrospinal fluid barrier dysfunction with minimal pleocytosis. Extensive microbiological, molecular, and immunological investigations, including polymerase chain reaction testing for neurotropic viruses and Mycobacterium tuberculosis, were negative. The patient had a history of meningitis six years earlier, complicated by hydrocephalus requiring ventriculoperitoneal shunt placement.

The patient was treated with anti-edematous, empirical antimicrobial, and supportive therapy, resulting in partial clinical improvement. This case highlights the diagnostic complexity of rhombencephalitis in elderly patients with prior central nervous system disease and emphasizes the importance of a multidisciplinary approach and careful exclusion of infectious etiologies.

***Keywords:*** rhombencephalitis, ventriculitis, brainstem encephalitis, ventriculoperitoneal shunt, magnetic resonance imaging

## Case report

A 75-year-old right-handed male was admitted to the Neurology Clinic due to progressive neurological deterioration. The patient was retired, married, and had higher

education. His presenting symptoms included progressive vertigo, nausea and vomiting, gait instability, daytime somnolence, confusion, and dysphagia.

The medical history was significant for meningitis treated abroad in 2019, which was complicated by increased intracranial pressure and required placement of a ventriculoperitoneal (VP) shunt. Additional comorbidities included hyperlipidemia, a history of alcohol use (ceased 5-6 years prior), and chronic smoking (approximately four cigarettes per day). Regular medications on admission included rivaroxaban 20 mg once daily, cilostazol 100 mg twice daily, atorvastatin 20 mg once daily, betahistine once daily, and cinnarizine 75 mg once daily.

On admission, the patient was afebrile, with a blood pressure of 110/60 mmHg and a regular heart rate of 74 beats per minute. General examination revealed limited mobility. Neurological examination showed a conscious but confused patient, disoriented to time and place but oriented to person. Horizontal gaze palsy with coarse nystagmus on left gaze was present. Coughing during liquid swallowing suggested bulbar involvement. Mild left-sided limb weakness was observed, along with an ataxic, broad-based gait and hyperreflexia in the lower extremities. Plantar reflexes were flexor bilaterally, and sphincter control was preserved.

Laboratory investigations revealed leukocytosis ($16.06 \times 10^9$/L), markedly elevated C-reactive protein (148 mg/L), mild hyponatremia (132 mmol/L), and a procalcitonin level of 0.56 ng/mL. Cerebrospinal fluid analysis demonstrated mild pleocytosis (13 cells), elevated total protein (0.63 g/L), increased albumin and IgG index, and evidence of blood–cerebrospinal fluid barrier dysfunction, without oligoclonal bands. Polymerase chain reaction testing for neurotropic viruses and Mycobacterium tuberculosis was negative.

Brain magnetic resonance imaging (picture 1 a, b, c, d) with and without contrast showed hyperintense T2/FLAIR lesions involving medulla oblongata and left cerebellar peduncle, with vasogenic edema and contrast enhancement of the ependyma and ventricular walls, findings consistent with rhombencephalitis and ventriculitis. No diffusion restriction was observed. The VP shunt was visualized in the right frontal region, without ventricular dilatation. Electroencephalography showed normal background activity without epileptiform discharges.

The patient received anti-edematous therapy, empirical antiviral and antibiotic treatment, anticoagulant therapy, anxiolytic and antipsychotic medications, and supportive care. During hospitalization, partial somatic and neurological improvement was observed. The patient was discharged in a stabilized condition, requiring assistance for daily activities, physical rehabilitation, and mobility aids. Follow-up at the Neurology Clinic was scheduled one month after discharge.

**Discussion**

Rhombencephalitis is a rare inflammatory disorder involving the brainstem and cerebellum, characterized by a broad etiological spectrum that includes infectious, autoimmune, paraneoplastic, post-infectious, and vascular causes. Owing to its heterogeneous clinical presentation and often nonspecific laboratory findings, diagnosis may be delayed, particularly in elderly patients and in those with pre-existing central nervous system pathology or cerebrospinal fluid diversion devices.

Early descriptions of rhombencephalitis emphasized its association with infectious etiologies, most notably Listeria monocytogenes. In a large review of 97 cases, Armstrong and Fung demonstrated that brainstem encephalitis frequently presents with subacute onset of vertigo, cranial nerve palsies, ataxia, and altered mental status, often in the absence of marked cerebrospinal fluid pleocytosis[1]. Subsequent studies confirmed that cerebrospinal fluid findings may be subtle or atypical, contributing to diagnostic uncertainty[2,3].

Magnetic resonance imaging plays a central role in the diagnostic evaluation of rhombencephalitis. Typical findings include T2/FLAIR hyperintensities involving the

brainstem and cerebellar peduncles, with variable contrast enhancement. Moragas *et al*. reported that ependymal enhancement and ventricular wall involvement may occur, particularly in cases with associated ventriculitis, and should raise suspicion for inflammatory or infectious processes affecting the hindbrain[3]. In the present case, MRI findings were highly suggestive of rhombencephalitis with ventriculitis, despite the absence of diffusion restriction and overt signs of abscess formation.

Although Listeria monocytogenes remains the most frequently reported infectious cause of rhombencephalitis in elderly and immunocompromised patients, microbiological confirmation is not always achieved. Brouwer and van de Beek highlighted that delayed diagnosis and empiric treatment are associated with worse outcomes in listerial CNS infections, underscoring the importance of early recognition and broad initial diagnostic workup[4]. In our patient, extensive microbiological and molecular investigations, including polymerase chain reaction testing for neurotropic viruses and Mycobacterium tuberculosis, were negative.

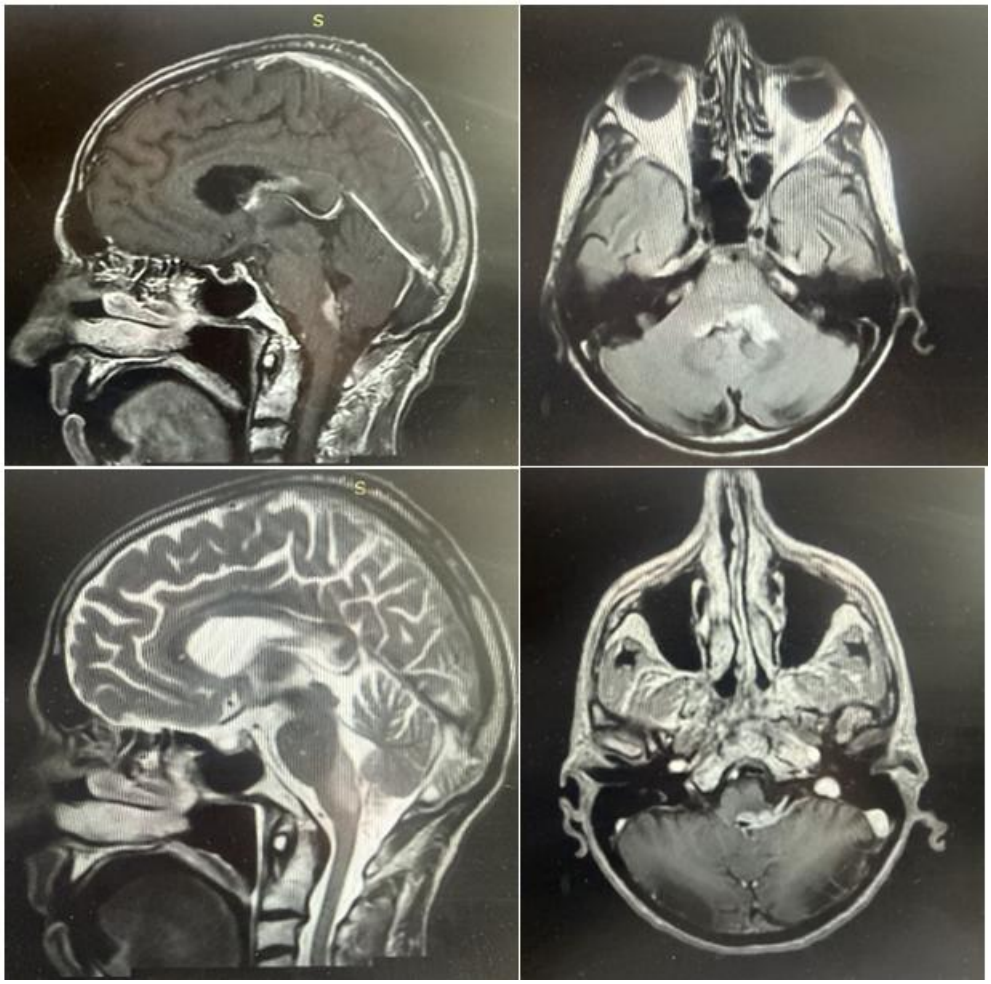

**Fig. 1.** Brain magnetic resonance imaging (a, b, c, d) with and without contrast showed hyperintense T2/FLAIR lesions involving medulla oblongata and left cerebellar peduncle, with vasogenic edema and contrast enhancement of the ependyma and ventricular walls, findings consistent with rhombencephalitis and ventriculitis.

The cerebrospinal fluid profile in this case, characterized by elevated protein concentration, increased IgG index, and blood–cerebrospinal fluid barrier dysfunction with minimal pleocytosis, favors an inflammatory rather than an acute bacterial infectious process. Such findings have been reported in post-infectious and autoimmune brainstem encephalitis,

as well as in inflammatory conditions occurring in the context of prior CNS injury or foreign material, such as ventriculoperitoneal shunts[2].

Recent literature has expanded the spectrum of rhombencephalitis to include post-infectious and immune-mediated mechanisms, including cases associated with systemic viral infections such as SARS-CoV-2. Ellul *et al*. emphasized that immune-mediated neurological syndromes may present with brainstem involvement and mimic infectious encephalitis, particularly in older patients with comorbidities[5]. Although no definitive autoimmune marker was identified in the present case, the clinical course and partial response to supportive and anti-edematous therapy support a non-bacterial inflammatory etiology.

This case highlights the diagnostic complexity of rhombencephalitis in elderly patients with previous CNS pathology and ventriculoperitoneal shunt placement. It underscores the necessity of a multidisciplinary diagnostic approach, repeated neuroimaging, and careful exclusion of infectious causes. Recognition of atypical laboratory profiles and imaging patterns is essential to guide appropriate management and avoid unnecessary delays in treatment.

## Conclusion

This case illustrates the diagnostic challenges of rhombencephalitis in an elderly patient with a history of previous meningitis and ventriculoperitoneal shunt placement. Despite radiological findings strongly suggestive of rhombencephalitis with associated ventriculitis, extensive microbiological and molecular investigations failed to identify a definitive infectious etiology. Cerebrospinal fluid findings and clinical evolution supported an inflammatory rather than acute infectious process. Early recognition, comprehensive diagnostic evaluation, and multidisciplinary management are essential for optimizing outcomes in complex cases of rhombencephalitis.

*Conflict of interest statement.* None declared.

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