Behavior Changes and Gait Unsteadiness: The Value of Imaging and Prompt Neurosurgical Intervention

Alteração do Comportamento e Instabilidade da Marcha: O Valor da Avaliação Neuroimagiológica e da Intervenção Neurocirúrgica Precoces

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ABSTRACT

Cavernous angiomas are central nervous system malformations. Most common manifestations are seizures and acute focal neurological deficits. We present a case report of a seventy-one year-old man with a two-month history of behavior changes, attention deficit and indifference followed by gait unsteadiness. Neuropsychological evaluation showed severe cognitive impairment and executive dysfunction. Head computed tomography depicted a supraventricular hydrocephaly. Magnetic resonance imaging revealed a small hemorrhage, contiguous to a mesencephalic cavernous angioma, obstructing the Sylvius aqueduct, causing secondary hydrocephalus. Four months after endoscopic ventriculocisternostomy, neuropsychological evaluation showed improvement and the patient regained autonomy. Parenchyma cavernous angiomas causing direct hemorrhage and subsequent obstruction of the Sylvian aqueduct are uncommon. Sub-acute behavior and mental state abnormalities are rare first manifestations of cavernous angioma and requires high clinical suspicion for its correct diagnosis. Magnetic resonance imaging evaluation is crucial in the detection of such patients as prompt neurosurgical intervention may substantially improve cognitive function.

Keywords: Cognition Disorders; Gait Disorders, Neurologic; Hemangioma, Cavernous; Magnetic Resonance Imaging; Neurosurgical Procedures; Tomography, X-Ray Computed; Ventriculostomy

RESUMO

Os angiomas cavernosos são malformações do sistema nervoso central cujas manifestações mais comuns são crises epiléticas e défices neurológicos agudos. Apresentamos o caso clínico de um doente de 71 anos com uma história com dois meses de evolução de alteração do comportamento, défice de atenção e apatia, sucedidas por marcada instabilidade da marcha. A avaliação neuropsicológica revelava alterações cognitivas e disfunção executiva marcadas. Na Tomografia Computorizada Cerebral observava-se hidrocefalia supraventricular enquanto na Ressonância Magnética cerebral era possível observar uma pequena hemorragia que obstruía o aqueduto de Sylvius localizada contiguamente a um angioma cavernoso mesencefálico. Quatro meses após o tratamento com ventriculocisternostomia endoscópica, o doente apresentava melhoria significativa na avaliação neuropsicológica tendo voltado a adquirir a autonomia prévia. A apresentação de um angioma cavernoso parenquimatoso como uma obstrução do aqueduto de Sylvius secundária a hemorragia é invulgar. Rara é também a sua apresentação como alterações do comportamento e do estado mental subagudas. Uma avaliação neuroimagiológica e uma intervenção neurocirúrgica precoces foram essenciais para a melhoria cognitiva observada.

Palavras-chave: Hemagnioma Cavernoso; Perturbações da Cognição; Perturbações Neurológicas da Marcha; Procedimentos Neurocirúrgicos; Ressonância Magnética; Tomografia Computorizada; Ventriculostomia

INTRODUCTION

Cavernous angiomas are hamartomatous vascular malformations,[1] with an estimated prevalence of 0.4% - 0.6%.[2] These comprise 10 to 15% of all central nervous system vascular malformations and mostly occur in the supratentorial territory.[3] Brainstem cavernous angiomas are much less common, comprising 18% of all cavernomas. The mean presentation age is 30.6 years, usually with seizures and acute focal deficits due to hemorrhage.[4] We present a case report illustrative of an unusual sub-acute clinical presentation of a brainstem cavernous angioma.

CASE REPORT

A seventy-one year-old male, upper secondary education level, with no relevant previous clinical history, was admitted due to recent behavior changes and gait unsteadiness. For two months, he had been losing initiative and interest in domestic tasks he used to pleasurably perform. In the previous two weeks his wife noticed he walked differently, being unable to climb down stairs and walk long distances. Initial neurological examination revealed psychomotor slowing, attentional deficit, temporal disorientation and lack of initiative. His gait was wide-based and tandem gait was impossible. Neuropsychological evaluation exposed cognitive impairment as assessed with Mini-mental State Examination (MMSE, 22 out of 30) and executive dysfunction in the frontal assessment battery (9 out of 18). Head computed tomography (CT) showed supratentorial ventricular enlargement. Brain magnetic resonance imaging (MRI) depicted an intra-aquedutal micro-hemorrhage contiguous to a mesencephalic cavernous angioma

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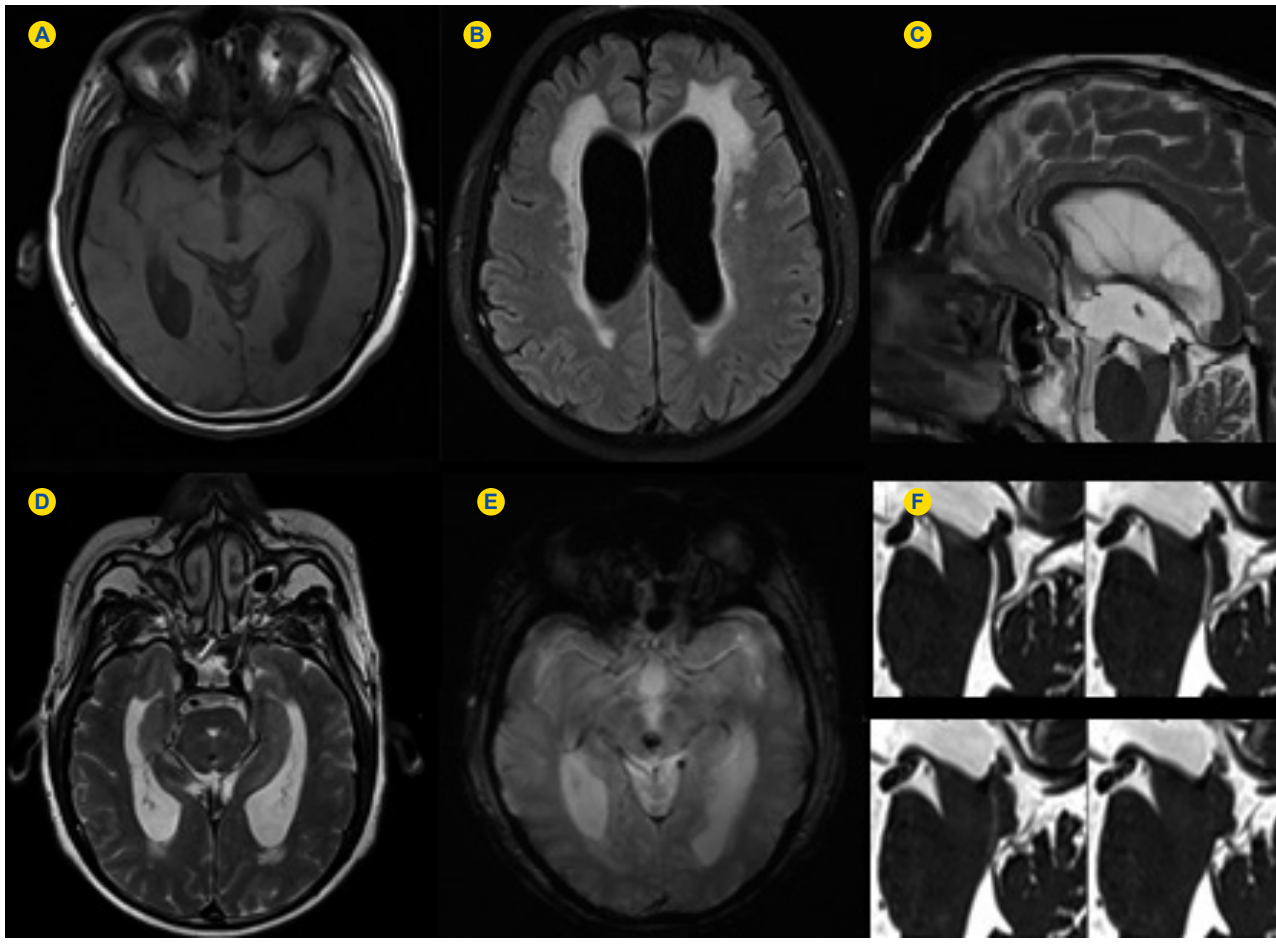

Figure 1 - Pre-operative MRI: A,B,D,E, axial (A - T1 SE; B - T2 FLAIR; D - T2 TSE; E - T2 gradient echo); C, Sagittal T2 TSE; F, Sagittal 3D CISS reconstruction, MRI. Obstructive supratentorial hydrocephalus with interstitial edema, due to periaquedutal cavernous angioma with recent hemorrhage (T1 hyperintensity) to the aqueduct of Sylvius.

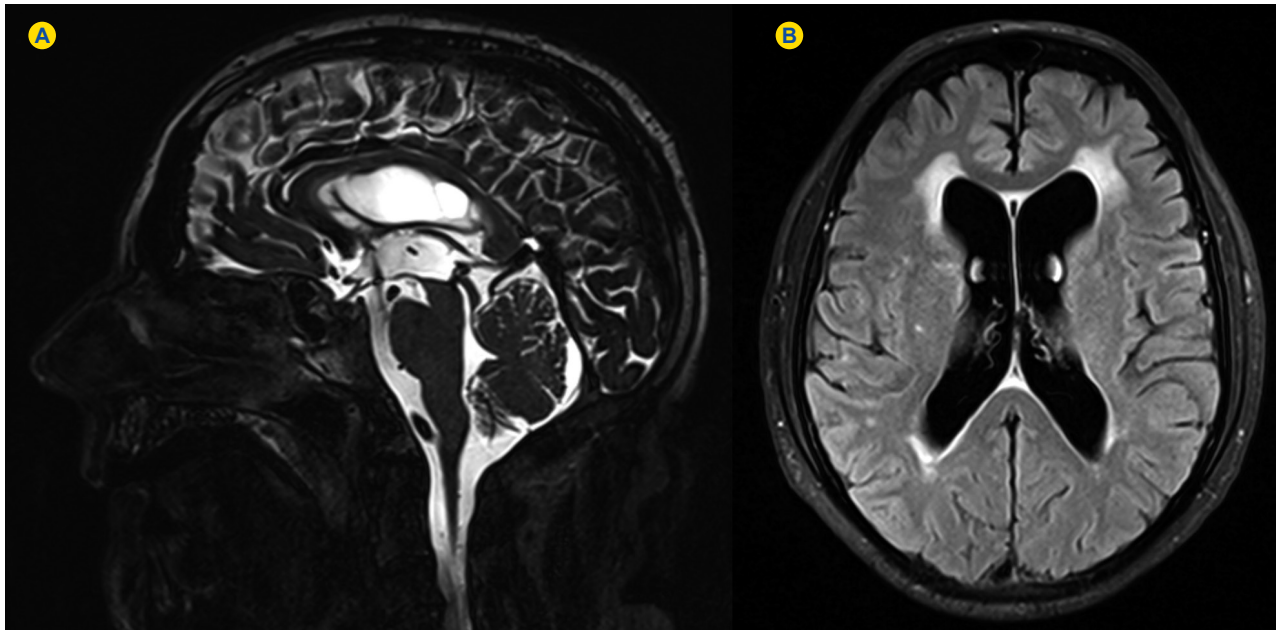

Figure 2 - Post-operative MRI three weeks after neurosurgery. A, Sagittal 3D-CISS reconstruction. Linear flow-void sign crossing the floor of the third ventricle, consistent with patency of ventriculostomy; B, axial T2 FLAIR. Decrease of ventricular size and periventricular interstitial edema.

(Fig. 1). An endoscopic ventriculocisternostomy was performed (Fig. 2). Four months later the patient had returned to his ordinary life, presenting an unremarkable gait. A second neuropsychological evaluation proved only a mild cognitive impairment in MMSE (26 out of 30) and no signs of frontal dysfunction in frontal batteries (14 out of 18).

DISCUSSION

This case report aims to illustrate an unusual subacute cavernous angioma presentation occurring due to hemorrhage and consequent obstructive hydrocephalus.

Numerous central nervous system malformations are described to cause obstructive hydrocephalus.[5,6] Reports concerning cavernous angiomas are more frequently associated with intraventricular location, causing obstruction due to direct hemorrhage[7-9] or parenchymal location, causing a mass effect on the cerebrospinal fluid drainage system. A comprehensive literature review had shown very scarce reports of parenchyma cavernous angioma causing direct hemorrhage and subsequent obstruction of the Sylvian aqueduct.[10] Furthermore, in this context, the presence of solely sub-acute mental state disturbance and behavior changes is fairly uncommon, since most patients present with headache and nausea owing to intracranial hypertension.

Rapidly progressive behavioral changes and mental state disturbance associated with unsteady gait should prompt immediate search for treatable causes of dementia. Brain MRI was crucial for the final diagnosis as well as for the neurosurgical management that led to considerable cognitive improvement.

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CONFLICTS OF INTEREST

The authors declare that there are no conflicts of interest.

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