

# Concurrence of multiple sclerosis and vascular malformation of the brainstem: case report

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**ABSTRACT** - Multiple sclerosis (MS) is a demyelinating disease of the central nervous system. In most of cases, the disease follows a relapsing-remitting pattern. Vascular malformations are described in differential diagnosis of MS because of the similarity of clinical presentation, especially when hemodynamic changes resemble the relapsing-remitting course of MS. Advances in diagnostic procedures, particularly magnetic resonance imaging (MRI) of neural axis and cerebral angiography have facilitated differential diagnosis between these two entities. However, simultaneous occurrence of asymptomatic vascular malformations and MS is rare. We present a case report of a patient with these two entities, i.e. MS diagnosed on the basis of clinical findings, supporting evidence from laboratory tests and MRI, and basilar artery aneurysm.

**Key words:** multiple sclerosis, aneurysm, basilar artery

## INTRODUCTION

Multiple sclerosis (MS) is an immune-mediated inflammatory disease that attacks myelinated axons in the central nervous system (CNS), which occurs mostly in young adults. In most cases, the disease follows a relapsing-remitting pattern. As first presentation prominent are sensory loss (40%) and eye symptoms (35%), less motor strength (21%), spinal cord symptoms (16%) and cerebellar symptoms (15%) (1). In some patients, neurologic deficit does not resolve completely and they enter the progressive phase of the disease (secondary progression). A minority of patients experience

steadily progressive neurologic deterioration (primary progressive MS). On diagnosing MS, most important are clinical findings, symptomatic episodes “separated in time and space”, but other courses of similar clinical findings must be excluded.

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ed (2). Differential diagnosis includes other diseases having relapsing-remitting course in young adults. It could even be expansive processes of the central nervous system (CNS), especially arteriovenous malformations (AVM), angiomas, but also meningiomas, gliomas of the brainstem, etc. (3,4). Sometimes, tumors have a relapsing-remitting course, similar to MS, but accurate diagnosis is made by magnetic resonance imaging (MRI) of neural axis. MS may not be recognized in the initial years, despite obviousness. MRI is the diagnostic procedure of choice for diagnosing MS. There is no laboratory or imaging test pathognomonic for MS (5). Diagnosis of MS or less common diseases includes series of diagnostic procedures (6,7). MS, however, can occur simultaneously with other diseases. Coexistence of MS and intracranial vascular malformation (VM) is very rarely described in the literature.

## CASE REPORT

A 29-year-old woman presented with vision disturbance characterized as optic neuritis. Symptoms were preceded by smallpox vaccination one month before. Eighteen months later, in spring, there were signs of pyramidal and brainstem lesion, with vestibular disturbances. In the autumn of the same year, she had a relapse in the form of motor weakness of lower extremities together with bulbar symptomatology, upper limb paresthesias, cerebellar manifestation, and optic nerve atrophy. A year later, she experienced a relapse again, followed by marked spastic paraparesis. Other exacerbations occurred at the age of 35, 45 and 50 years. Clinical presentation was predominated by spastic paraparesis, ataxia, dysarthria and sensory disturbances.

Considering that symptoms were “separated in time and space”, we performed additional procedures. Serology for *Toxoplasma gondii* and *Borrelia burgdorferi* was negative, while vitamin B12 and folate findings were normal. HLA phenotype was A2, B5, B40, DR1, DR3, DQ1, DQ2. Anti-tissue antibodies, hepatitis markers, HIV and Wright’s reaction were negative. Plasma cells in the cerebrospinal fluid (CSF) were reported. CSF analysis confirmed the presence of oligoclonal bands. Color Doppler and transcranial color Doppler revealed high circulatory resistance at the irrigation of basilar artery and right vertebral artery.

In relapses, the patient was treated with corticosteroid therapy, which led to remission.

Three years after the last exacerbation, at the age of 53, right sided peripheral facial palsy and impair-

ment of bulbar movements were evident. MRI of the CNS indicated demyelinating lesions characteristic for MS (Fig. 1). According to clinical presentation and color Doppler findings, cerebral digital subtraction angiography (DSA) was also performed (Fig. 2). It confirmed basilar artery aneurysm, parasagittal on the right side. Based on clinical findings, “separated in time and space”, MRI of the CNS and finding of oligoclonal bands

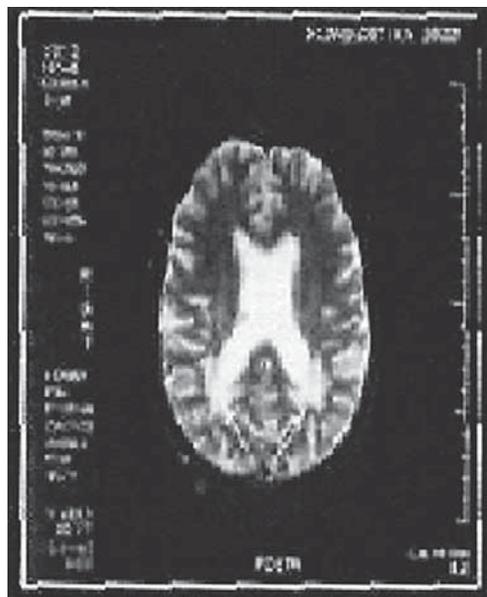


Fig. 1. (SE TR 2500, TE 25/110, TR 500, TE20) Cerebral MRI: cerebral atrophy, multiple hyperintensive lesions in the white matter, especially periventricularly, typical of MS lesions.



Fig. 2. Cerebral DSA: aneurysm of the upper part of basilar artery, partially thrombosed, aneurysm diameter 20 mm.

on CSF examination, definitive diagnosis of MS and intracranial aneurysm of the basilar artery was made. For now, we decided to prescribe conservative treatment with monitoring of the aneurysm.

## DISCUSSION

Differential diagnosis of MS, among others, includes cerebrovascular disease, intracranial neoplasms, arteriovenous malformations, especially of the spinal cord (7). De Stefano *et al.* describe the difference between isolated demyelination and astrocytoma (8). Ernst *et al.* (9) report on isolated damage appearing as annular amplification surrounded by edema, which showed good response to corticosteroid treatment and was eventually confirmed as MS on cerebral angiography. Sometimes, arteriovenous malformation can mimic the clinical presentation of MS, particularly primary and secondary progressive MS, but may also have a relapsing-remitting course because of hemodynamic changes (10,11). Thanks to the development of diagnostic procedures, particularly MRI and cerebral angiography, differentiation of these two entities is easier today. Arteriovenous malformations, which mostly occur at spinal cord and brainstem, often have fluctuating symptoms that are not revealed in the initial stage of the disease (12-14). Dhopes and Weinstein (15) and Ritzenthaler *et al.* (16) point to the importance of early detection of vascular malformation, thus enabling timely operative treatment and prevention of neurologic deterioration.

Simultaneous occurrence of MS and other diseases is possible. Concurrence of MS and vascular malformation of the CNS is rarely reported. Ho and Wolfe report on 20 patients with MS and simultaneous occurrence of glioma and 9 patients with MS and other neoplasms recorded in the literature. In their study, there was only one patient with MS and arteriovenous malformation. It was a 63-year-old patient with a 25-year history of MS. Arteriovenous malformation was revealed on autopsy. This was the first case of simultaneous occurrence of MS and arteriovenous malformation described (17). Only a few similar cases are found in later literature. Our patient is therefore a rare example of simultaneous occurrence of MS and basilar artery aneurysms, verified by MRI and cerebral angiography. The case presented also points to the importance of extra examinations besides the usual diagnostic procedures in the algorithm for a specific disease in case of clinical suspicion and aberration.

## CONCLUSION

Cerebrovascular disease, intracranial neoplasms, arteriovenous malformation, particularly of the spinal cord and meninges, may be coexisting with MS. Sometimes, arteriovenous malformation can mimic the clinical presentation of MS, having a relapsing-remitting course because of hemodynamic changes. Thanks to the development of diagnostic procedures, particularly MRI and cerebral angiography, differentiation of these two entities is easier today. Asymptomatic and symptomatic vascular malformation can occur simultaneously with MS, but it is rarely described in the literature. Our case is an example of these two entities coexisting in one patient, who had clinically defined MS and basilar artery aneurysm.

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## Združenost multiple skleroze i vaskularne malformacije moždanog debla: prikaz bolesnika

**SAŽETAK** - Multipla skleroza (MS) je demijelinizacijska bolest središnjega živčanog sustava, najčešće relapsno-remitirajućeg tijeka. Vaskularne malformacije opisane su u diferencijalnoj dijagnozi MS zbog sličnosti kliničke slike, osobito u slučaju relapsno-remitirajućeg tijeka, zbog hemodinamskih promjena. Zahvaljujući razvoju dijagnostičkih postupaka, osobito magnetske rezonancije (MR) živčane osi i cerebralne angiografije, razlikovanje ovih dvaju nozoloških entiteta danas je jednostavnije. Rijetka je, međutim, istodobna pojavnost asimptomatske vaskularne malformacije i MS. Prikazujemo bolesnicu s oba klinička entiteta, tj. s klinički sigurnom MS potvrđenom laboratorijskim testovima i pomoću MR, i aneurizmom bazilarne arterije.

**Ključne riječi:** multipla skleroza, aneurizma, bazilarna arterija