

Dysarthria, difficulty in walking and dizziness

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CASE REPORT

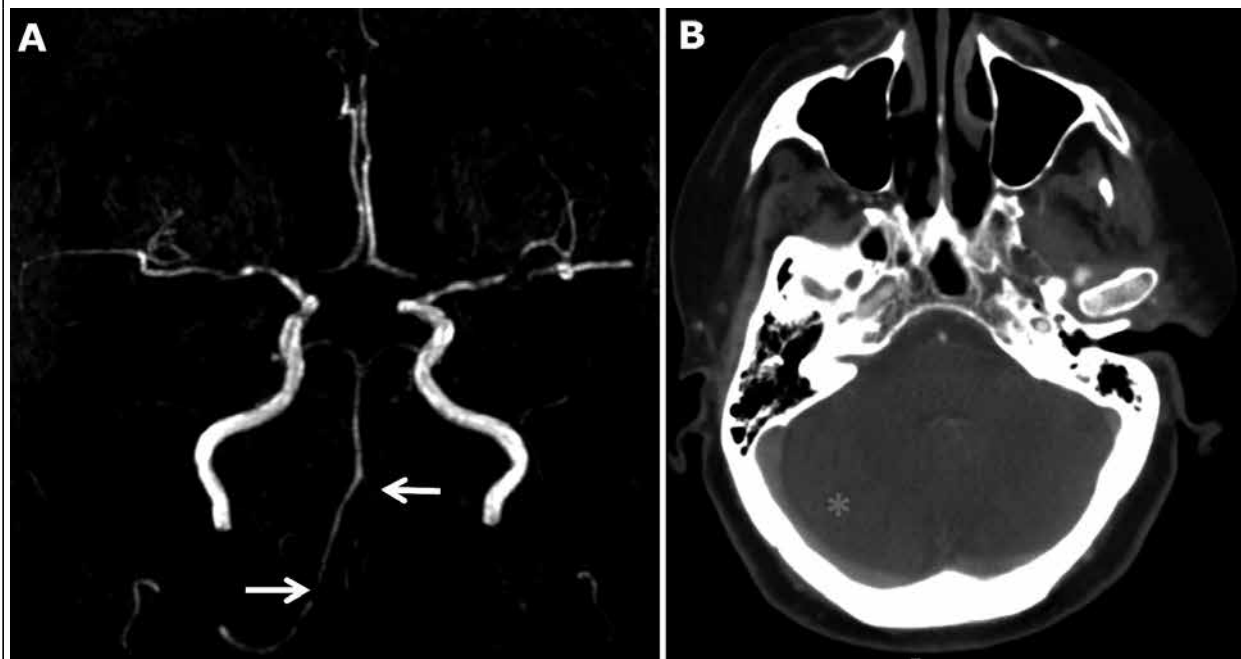
A 75-year-old Dutch woman presented with dysarthria, difficulty in walking, vertigo, headache and vomiting. Until recently she was an active and relatively healthy woman. In a few weeks, she had become bedridden by her vertigo. She had been diagnosed with polymyalgia rheumatica by her general practitioner five months earlier for which she used a prednisolone weaning schedule (7.5 mg once daily at presentation). On physical examination a dysarthria, nystagmus, broad-based gait and word-finding difficulty

were observed. Laboratory results showed an erythrocyte sedimentation rate (ESR) of 110 mm/h and C-reactive protein (CRP) level of 36 mg/l.

WHAT IS YOUR DIAGNOSIS?

See page 379 for the answer to this photo quiz.

Figure 1. Brain magnetic resonance angiography showing a bilateral occlusion of the vertebral arteries with retrograde filling of the right vertebral artery (A) with fresh ischaemia bilaterally in the cerebellum and occipital lobe (B, asterisk)



DIAGNOSIS

Magnetic resonance angiography revealed a bilateral occlusion of the vertebral arteries (*figure 1A*) with retrograde filling of the right vertebral artery, collateral formation and a divergent circle of Willis with fresh ischaemia bilaterally in the cerebellum and occipital lobe (*figure 1B*). A temporal artery biopsy confirmed giant cell arteritis (GCA). A diagnosis of bilateral cerebellar and occipital ischaemia as the consequence of bilateral vertebral arterial occlusion (BVAO) resulting from GCA was made. Treatment with high-dose pulse methylprednisolone was started after which the symptoms completely disappeared and the ESR and CRP level normalised.

BVAO is a rare complication of GCA and has a high mortality (75-80%).¹ 3% of GCA patients develop a (considered GCA related) cerebrovascular accident, of which 40-60% involves the vertebrobasilar area. Conversely, among 118 reported cases of non-traumatic BVAO, five cases (4.2%) were attributed to GCA.¹ The intracranial vertebrobasilar arteries are almost always spared from GCA (except the proximal 5 mm after the passage through the dura mater), possibly due to the significant rarefaction of the inner elastic layer.¹ The recognition of GCA as the underlying cause of BVAO may be difficult in part due to the relatively high prevalence of atherosclerosis in this older population. In our case, the patient had recently been diagnosed with polymyalgia rheumatica. Previous cases have been reported in which BVAO with associated neurological signs and symptoms were the first clinical manifestations of GCA.^{1 (patient 1),2} Clinically, BVAO resulting from GCA differs from BVAO

of arteriosclerotic origin by the much higher mortality rate (75% vs 19%, respectively), the presence of headache (100% vs 22%), fever (50% vs 0%) and an elevated ESR and not by neurological signs.¹ The slower evolution over time observed in arteriosclerotic BVAO allows the formation of collaterals supplying the otherwise perfusion-deprived vertebrobasilar territory. In GCA, the much more accelerated progression impedes the timely formation of collaterals which may itself contribute to the remarkably higher mortality of BVAO.³ The spectrum of clinical manifestations in BVAO is wide and reflects the territories supplied by these arteries and includes visual disturbances, cranial nerve palsies, affection of the pyramidal and sensory tracts, cerebellar signs and altered consciousness. Due to the rareness of BVAO caused by GCA, therapy remains empiric and consists of prompt administration of high-dose corticosteroids. Additionally, anticoagulation and other immunosuppressive (as cyclophosphamide) therapy should be considered per individual patient in this highly fatal disease.

REFERENCES

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