

Fatal Spontaneous Bilateral Vertebral Artery Dissection in Giant Cell Arteritis (GCA)

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Introduction

Giant cell arteritis (GCA) is a common immune-mediated inflammatory disease of unknown etiology which typically occurs in patients older than 50 years of age and affects medium and large sized arteries. Different medical subspecialties are frequently involved in the management of patients with GCA. Therefore, this report shall highlight the potential life-threatening complications of GCA and raise awareness of those in the different subspecialties treating these patients.

In this case, we report the medical history of a 77 year old woman, which presented with a four-day history of blurred vision of the right eye in our clinic. Further symptoms were bi-temporal headache and intermittent jaw claudication. Her past medical history was unremarkable except of sufficiently controlled arterial hypertension.

Physical examination revealed diminished pulses of the superficial temporal arteries. Funduscopy showed paleness of the right optic disc, suspicious of anterior ischaemic optic neuropathy. C-reactive protein was 5.9 mg/l (reference range: <5.0 mg/l) and the erythrocyte sedimentation rate was 60 mm/h (reference range: <30 mm/hour). Cranial giant cell arteritis (GCA) was suspected and treatment with intravenous prednisolone 1 g per day was started. Under this therapy the patient showed exacerbation of the known arterial hypertension. This was sufficiently treated with intensified anti-hypertensive medication.

Three days after treatment initiation, the patient experienced complete vision loss of her right eye, despite normalisation of the laboratory inflammatory parameters under treatment. In order to prevent bilateral vision loss, high dose intravenous corticosteroid treatment was continued for a total of 7 days, and low-dose aspirin was commenced. Diagnosis of GCA was confirmed by temporal artery biopsy (Figure 1D).

On day 8 of corticosteroid treatment, after reduction to a prednisolone dose of 1 mg/kg bw/day, the patient reported acute visual deterioration of her left eye. Subsequently, the patient collapsed and exhibited tetraparesis, dysarthria, and conjugate gaze palsy. A few minutes after symptom onset, the patient lost consciousness. As protective reflexes were absent, endotracheal intubation was required. Cerebral computed tomography with CT-angiography was immediately performed and demonstrated occlusions of the V2- and V3-segments of both vertebral arteries. Intravenous systemic

thrombolysis with rt-PA was started 1 hour and 40 minutes after symptom onset. After completion of thrombolysis, follow-up CT-angiography documented persistent occlusion of the vertebral arteries. Therefore, endovascular treatment was attempted. Intra-arterial angiography suggested bilateral, long segment vertebral artery dissection, with the intracranial segments of the vertebral arteries and the basilar artery appearing patent (Figure 1A). Percutaneous transluminal angioplasty of the left vertebral artery was performed with procedural success (Figure 1B). Following the intervention, the patient was brought to the intensive care unit and intravenous dose-adjusted unfractionated heparin was administered. Unfortunately, left-sided hemiplegia persisted and the patient's condition gradually deteriorated during the following days with eventual loss of brainstem-reflexes. Repeated cranial CT documented extensive bilateral ischaemic infarction of the thalamus, cerebellum and brain-stem (Figure 1C).

The patient died six days after onset of neurological symptoms due to central dysregulation caused by cerebellar tonsillar herniation. Necropsy confirmed the diagnosis of bilateral vertebral artery dissection and uncovered Vasculitis involvement of the vertebral arteries (Figure 1E).

Cranial ischemic complications are observed in about 20% of patients with GCA [1], but ischemic stroke is a rare disease manifestation (2 to 3% of patients) [1,2]. The vertebrobasilar circulation is affected most frequently [1-3]. The reported cases were similar in the clinical presentation with typical clinical symptoms of GCA (age >50 years, new onset of headache, elevated inflammatory parameters and some with jaw claudication and/or visual deterioration) and a subsequent neurological worsening [3-5]. The majority of events occur before or early after initiation of corticosteroid treatment [2,3]. Vice versa, bilateral vertebral artery occlusion should always raise suspicion of underlying GCA [3]. Treatment is challenging and, in addition to established medical treatment strategies of GCA and ischemic stroke, may necessitate rescue endovascular treatment. While clinically not successful in our patient, neurological improvement after angioplasty and stenting has been reported in a similar clinical situation [4].

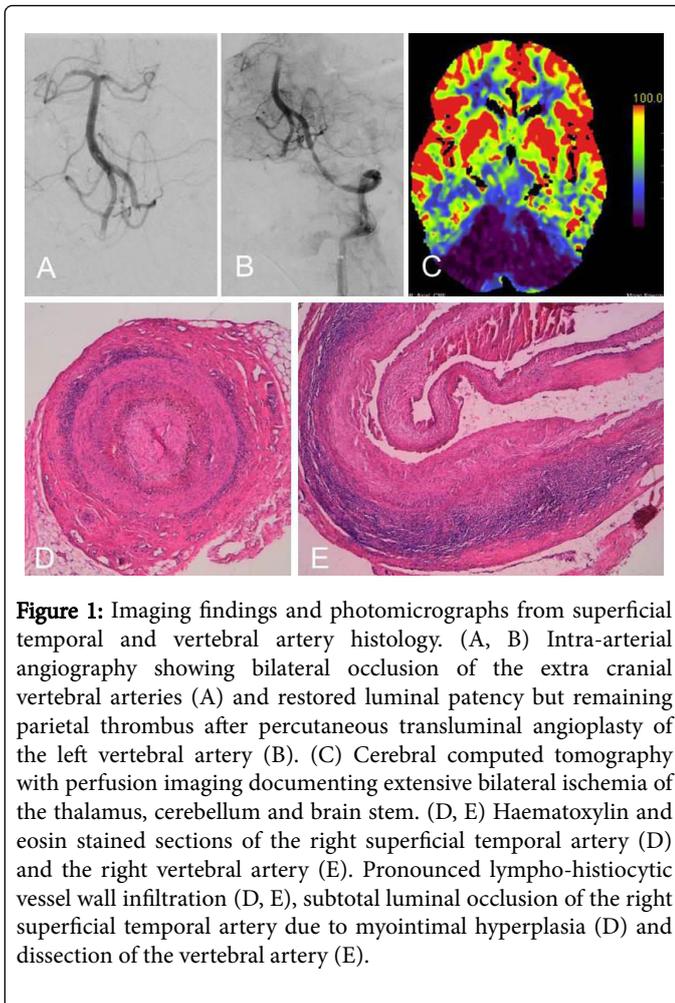


Figure 1: Imaging findings and photomicrographs from superficial temporal and vertebral artery histology. (A, B) Intra-arterial angiography showing bilateral occlusion of the extra cranial vertebral arteries (A) and restored luminal patency but remaining parietal thrombus after percutaneous transluminal angioplasty of the left vertebral artery (B). (C) Cerebral computed tomography with perfusion imaging documenting extensive bilateral ischemia of the thalamus, cerebellum and brain stem. (D, E) Haematoxylin and eosin stained sections of the right superficial temporal artery (D) and the right vertebral artery (E). Pronounced lympho-histiocytic vessel wall infiltration (D, E), subtotal luminal occlusion of the right superficial temporal artery due to myointimal hyperplasia (D) and dissection of the vertebral artery (E).

Arterial dissection was found to be causative for acute bilateral vertebral artery occlusion in our patient. Although it is well known that GCA carries a risk for aortic dissection in the mid and long term, dissections of the supraaortic branches including the vertebral arteries have been reported only occasionally [5].

Medical subspecialties involved in the management of patients with GCA should be aware of the potentially catastrophic complications of bilateral vertebral occlusions resulting in leading to major stroke in the posterior circulation.

Key Message

Awareness of a fatal bilateral-vertebral-artery dissection due to large vessel Vasculitis should be raised.

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