Extradural, retro-orbital, arteriovenous malformation as the cause of a long-term history of syncope

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DESCRIPTION

In May 2012, a 73-year-old man presented to us with a long-term history of syncope occurring during daily activities, characterised by a rapid onset and spontaneous recovery. He had a history of hypertension and previous coronary stent placement. Technetium-99m sestamibi cardiac scintigraphy and neurological and vestibular assessments were all negative. A head-up tilt test (HTT) and cerebral MRI were then performed. During the HTT, his HR, blood pressure (BP) and cardiac output (CO, l/min) were measured using a Finometer (Finapres, The Netherlands). When the patient was tilted to 60° for 20 min without giving any medication, his CO decreased (3.2 vs 4.8 l/min), but his HR (59 vs 62 bpm) and BP (122/74 vs 116/63 mm Hg) did not. After sublingual administration of nitroglycerin 0.3 mg, the tilting was interrupted because of nausea, blurred vision and dizziness with a minimal further CO reduction and decrease in BP to 62/43 mm Hg. A brain MRI revealed the presence of a left retro-orbital, extradural, high-flow arterio-venous malformation (AVM).2

In AVMs, the arteries connect directly to veins without a capillary bed in between. This creates a high-pressure shunt. AVMs thus ‘steal’ blood from the surrounding brain normal tissue (figure 1). This condition of cerebral hypo-perfusion may abruptly worsen and cause syncope in the presence of severe hypotension, which may occur after a HTT or when recovering from some exercise activities. In this patient, AVM treatment options, such as microsurgery, endovascular therapy and radiosurgery,3 should be considered.

Learning points

▸ Extramural retro-orbital arterio-venous malformation is a rare congenital developmental vascular lesion.
▸ This anomaly may cause haemorrhagic stroke, epilepsy, chronic headache or focal neurological deficits.
▸ The substantial differences between surgical, endovascular, radiosurgical and multimodality treatments are impacted by age and haemorrhage status.

Figure 1 Cerebral MRI. Left retro-orbital, high-flow arterio-venous malformation. (A) Coronal, T2-weighted images show a signal void (blue arrows in A and B) in a vein located in the middle cranial fossa adjacent to the left fronto-temporal lobe. (B) Three-dimensional time of flight-MR angiography images show a central nidus (red asterisk) located posterior to the left orbit that determines a shunt between a small feeding artery originating from the left carotid artery at the level of the cavernous sinus tract (red arrows in B) and the superficial venous drainage.
REFERENCES

