Acute ophthalmic artery occlusion in decompression illness with underlying anterior cerebral artery A1 segment hypoplasia

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Abstract

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A diver presented with total loss of vision in the left eye and right hemiparesis following a routine no-stop scuba dive to 20 metres' depth. A diagnosis of decompression illness (DCI) with acute ophthalmic artery air embolism and left carotid artery insult causing acute anterior circulatory ischaemia was made. He underwent seven hyperbaric treatments leading to a full recovery. Magnetic resonance angiography revealed an underlying left anterior cerebral artery A1 segment hypoplasia. Making a prompt diagnosis and early hyperbaric oxygen treatment are crucial to halt further tissue damage from ischaemia in central nervous system DCI. In this case, the finding of a left A1 anterior cerebral artery segment hypoplasia variant may have increased the severity of DCI due to deficient collateral circulation.

Introduction

Breathing compressed air under pressure results in increased dissolved inert gases in tissues and reduction in ambient pressure may lead to dysbaric diseases, commonly referred as decompression illness (DCI).[1] A common feature of DCI is the embolization of gas bubbles in the venous and/or arterial circulation. This causes ischaemia in surrounding tissues, which is responsible for the large variation of presenting symptoms.

The congenital variation of hypoplastic A1 segment anterior cerebral artery (ACA) of the circle of Willis is reported in 1–10% of the population, based on angiographic and autopsy studies.[2] This preexisting variation may impair collateral blood flow through the circle of Willis and is a recognised risk factor for ischaemic stroke.[2,3] We report a diver with DCI in whom this congenital defect may have contributed to an ipsilateral ophthalmic artery occlusion and carotid artery insult causing anterior circulatory ischaemia.

Case report

A 28-year-old man presented four hours after surfacing from scuba diving with painless, acute total loss of vision in the left eye. He was a smoker with a five-year-pack history with no other significant medical history. He had three years of diving experience, completing 68 uneventful dives. This particular dive was a single dive to 20 metres' depth and a 30 minutes bottom time. He had good buoyancy control, maintained a safe ascent rate and completed a safety stop. He developed sudden blurring of vision and floaters in his left eye a few minutes after surfacing and within four hours his left eye vision was totally lost with no light perception (NLP). This was associated with bilateral fronto-orbital headache and right-sided hemiparesis. He immediately sought treatment at a nearby hospital. Upon arrival, his right visual acuity was 20/20 and the left eye was NLP in all quadrants. The relative afferent pupillary defect sign (RAPD) was positive, indicating absence of retinal signal passing through the optic nerve from the left side. Anterior segment examinations were unremarkable. Posterior segment finding and disc appearance of both eyes were normal. There was no 'cherry red spot' sign, no visible venous pulsation or focal arterial narrowing. Neurological assessment revealed a right-sided sensorimotor deficit.

The diagnosis of left ophthalmic artery occlusion secondary to arterial gas embolism (AGE) was made in view of total visual loss and positive RAPD as well as absence of a 'cherry red spot' (due to lack of blood flow to both the retinal and the choroidal circulation).The contralateral motor-sensory deficit was the result of a vascular insult to the left carotid artery, causing acute anterior circulatory ischaemia with lacunar syndrome (from probable occlusion of one of the penetrating arteries providing blood to deep brain structures).

He was admitted and hyperbaric oxygen therapy (HBOT) was commenced according to the Royal Navy Treatment Table 62.[4] He received a total of seven HBOT sessions at

the end of which left visual acuity was 20/20 with no RAPD and the right hemiparesis had recovered fully. Magnetic resonance angiography (MRA) showed hypoplasia/absence of the A1 segment of the left anterior cerebral artery (ACA). Transthoracic echocardiography (TTE) was performed and there was no evidence of a persistent foramen ovale (PFO).

The patient was referred to a diving medicine specialist who certified him fit to return to diving. However, he was advised to dive within the no-decompression limits, to optimise his fitness and to dive in areas where HBOT was readily accessible.

Discussion

Arterial gas embolism (AGE) is often due to the pulmonary over-inflation syndrome, caused by the expansion of trapped gas in the lung during ascent.[5] It is especially associated with rapid or uncontrolled ascent. Alveolar air may enter the pulmonary venous circulation, thence to the systemic arterial circulation blocking the lumen and damaging the endothelium of small distal arteries, causing ischaemic tissue damage. The brain is especially vulnerable because it obtains a high proportion of cardiac output.[6,7] In this diver's case, neither a rapid ascent nor exceeding recommended dive times appear to have been contributing factors.

The incidence of ocular symptoms in patients with DCI has been reported to be 7% and 12% in two large study series.[8,9] Ocular features described include nystagmus, diplopia, visual field defects, cortical blindness, periocular pain, convergence insufficiency, optic neuropathy and central retinal artery occlusion.[8,9] Delay in therapeutic recompression will cause tissue hypoxia, subsequently leading to permanent ischaemic injury and poor recovery.[9]

The diver was shown to have ACA A1 segment hypoplasia on MRA imaging. The A1 segment of the ACA is a principal supplier of anterior collateral blood flow. Symptoms of neurological deficit in this patient were consistent with a study in which the majority of patients with A1 hypoplasia-related stroke had lacunar infarcts with a contralateral hemiparesis, resulting from occlusion of the penetrating arteries which supply the deep structures of the brain.[3]

TTE was within normal parameters, with no evidence of a PFO. The presence of a PFO is reported to increase the risk of AGE in divers. High-resolution computed tomography (HRCT) of the lungs was not done because the patient defaulted his appointment. Due to our limited resources, we were unable to proceed with pulmonary angiography. HRCT and pulmonary angiography are important investigations in order to look for known risks of AGE such as small airway disease and pulmonary bullae.

Issuing a diving certification or permitting hyperbaric exposure in workers with known ACA A1 segment hypoplasia must take into consideration the absence of collateral circulation in that brain area and the risk must be evaluated thoroughly because the condition has both medical and safety implications. If a fit-to-dive certification is issued, the hyperbaric hazards and risks of a higher morbidity in case of DCI should be fully explained to he diver who should be cautioned to avoid diving in locations lacking rapid accessibility to HBOT.

Conclusion

The developmental variant of ACA A1 segment hypoplasia in the circle of Willis was a major contributing factor in the severity of the ophthalmic artery and anterior circulatory arterial occlusion in a diver with DCI. Early recognition and prompt therapeutic recompression with well-established protocols are crucial to prevent ischaemic tissue injury and permanent disability. Any decision regarding fitness to dive or other hyperbaric exposure for people with this variation should be made taking into account medical safety, economics and medico-legal concerns.

References

1 Bove AA. Diving medicine. Am J Respir Crit Care Med. 2014;189:1479–86.
2 Mäurer J, Mäurer E, Perneczky A. Surgically verified variations in the A1 segment of the anterior cerebral artery. Report of two cases. J Neurosurg. 1991;75:950–3.
3 Chuang YM, Liu CY, Pan PJ, Lin CP. Anterior cerebral artery A1 segment hypoplasia may contribute to A1 hypoplasia syndrome. European Neurology. 2007;57:208–11.
4 BR2806 Royal Navy diving manual. London: Her Majesty's Stationery Office; 1972.
5 Hall J. The risks of scuba diving: a focus on decompression illness. Hawaii J Med Public Health. 2014;73(Suppl.2):13–6.
6 Moon RE, Vann RD, Bennett PB. The physiology of decompression illness. Scientific American. 1995;272:70–7.
7 Russi W. Diving and the risk of barotrauma. Thorax. 1998;53(Suppl.2):S20–S24.
8 Summitt JK, Berghage TE. Review of diving accident reports 1968. Washington, DC: US Navy Experimental Diving Unit Research Report 11-70. 1970;11–70. [cited 2018 April 22]. Available from: http://www.dtic.mil/dtic/tr/fulltext/u2/723175.pdf.
9 Rivera JC. Decompression sickness among divers – an analysis of 935 cases. Military Med. 1964;129:314–34.

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