Case report

Transient prosopagnosia resulting from a cerebral gas embolism while diving

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Key words
Cerebral arterial gas embolism (CAGE), arterial gas embolism, decompression illness, prosopagnosia or visual agnosia, treatment, case reports

Abstract
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A 33-year-old male was diving in excess of 50 metres of water in a marine tunnel at a remote location off the west coast of Scotland when he ran out of air. He made an emergency ascent and was recovered unconscious on the surface; he was not breathing, had no pulse and had missed a significant amount of decompression. Following resuscitation he was transferred by helicopter to the Dunstaffnage Hyperbaric Unit where he received hyperbaric oxygen therapy for a cerebral arterial gas embolism. He made a quick recovery but during the treatments he demonstrated a number of neurological abnormalities including visual disturbances which were diagnosed as prosopagnosia, the inability to recognise faces. The effects were transient and the patient went on to make a full recovery. Prosopagnosia is a rare affliction and this is thought to be the first reported case that appears to have occurred as a direct result of a diving accident. The case is described in detail and prosopagnosia is reviewed.

Introduction

Arterial gas embolism (AGE) is a recognised serious problem related to diving, caused when gas passes through the alveolar walls and into the pulmonary vasculature.[1] There are a wide number of signs and symptoms related to AGE depending where the bubbles are distributed in the brain.[1] Prosopagnosia is a rare condition in which individuals are unable to recognise faces. More unusually transient prosopagnosia has been reported following ischaemic cerebral vascular accidents, as part of normal pressure hydrocephalus and following temporal surgery for epilepsy.[2–4] It does not appear to have been reported previously in relation to diving. This report gives details of a rare case of transient prosopagnosia following a cerebral gas embolism while diving.

Case report

JP was a 33-year-old television engineer who was an experienced sports scuba diver for 16 years, with no past medical history of note or history of any diving-related medical problems. He was taking part in a liveaboard expedition which had departed from Oban (on the west coast of Scotland) to the isolated volcanic archipelago of St Kilda, lying in the Atlantic 45 miles to the west of the Outer Hebrides, 170 miles from Oban (see postscript).

The diving expedition started with a "shake down" dive on the first afternoon to a maximum depth of 30 metres' sea water (msw) for a total duration of 18 minutes. Following that dive, the liveaboard vessel began the 170 mile journey to the St Kilda islands; the boat arrived there late in the evening of the following day. On the day following arrival, JP carried out a single dive to a maximum depth of 61.4 msw with a total dive time of 34 minutes.

After a surface interval of 22 hours, he carried out his third dive, the incident dive, at 1112 hr to a maximum depth of 51.8 msw for about 30 minutes. He entered the water with twin air tanks of unknown capacity and descended on his own. He entered a cave at about 50 msw and prepared to wait for his companions with his camera. While he waited he began exploring, on his own, one of the tunnels and swam along to its end. During this initial exploration he dipped into a couple of dead-end holes. Still at the end of the tunnel he was aware that he still had not been joined by his companions and so he decided to return to the entrance. However, on his way back he encountered a strong current and remembered having to swim with considerable effort, finning hard and pulling himself along the cave wall. At the same time he remembered that he was "dragging" on his demand valve. He also remembers being relieved to have made it back to the entrance and seeing the sun filtering down through the water. Unfortunately because of the excessive effort used, JP had run out of air and he was still at a depth of approximately 50 msw. He had no recollection of surfacing but he had appeared to have inflated his buoyancy jacket and surfaced rapidly. He had some vague recollection of being in a helicopter but really remembered nothing for the next three hours until he was being examined at the hyperbaric unit at Dunstaffnage, near Oban.

According to the divers on the scene he was seen floating on the surface at 1145 hr. He was recovered into a dinghy, when he was noted to not be breathing and have no pulse, so they commenced cardiopulmonary resuscitation, carrying it out for 5–10 minutes as he was taken back to the dive boat. His twin tanks were noted to be empty as his gear was removed; his drysuit was cut off and he was placed in the recovery position as he was by now breathing and had a pulse.

He was taken ashore to the Ministry of Defence facilities on St Kilda and was attended by an army medic. The emergency services were contacted and Stornoway Coastguard tasked their helicopter for urgent evacuation from St Kilda to Dunstaffnage. He remained deeply unconscious for a further 30 to 40 minutes, was given high-flow oxygen via closed re-breathing trauma mask and the medic started an intravenous infusion. During the helicopter transfer oxygen and IV fluids were continued as his conscious level started to improve; he arrived at the Dunstaffnage Hyperbaric Unit at 1430 hr.

Examination on admission found him confused, with poor recollection of recent or past events, and with disorientation in time and place. Finger–nose coordination was poor and he was noted to have dysdiadochokinesia. He had generalised bilateral weakness of the thigh and calf muscles (grade 3). Reflexes were brisk in both knees and ankles with sustained clonus in both ankles. Clonus was also triggered by having his reflexes tested. He had been incontinent of faeces and possibly of urine. His cranial nerves were intact.

Recompression commenced at 1445 hr to 18 metres (283 kPa) following the Royal Navy treatment table 62 protocol. At the first air break he was less confused. At the second air break he was able to stand unsupported, carry out a heel-shin slide while standing and carry out accurate serial sevens. As he reported to be feeling "absolutely normal" and examination showed no abnormality the treatment table was continued without extensions, surfacing at 1930 hr. He later admitted, however, having difficulty in distinguishing or recognising the faces of those who were treating him.

On completion of the treatment, he was admitted to the local hospital for post-treatment monitoring. He kept asking as to what had happened to him and if he had inflated his buoyancy jacket. He also now reported some problem with his vision. Though he had a history of an amblyopic left eye since childhood, his near and distance vision were normal, he was able to recognise objects, and could see people but had difficulty in recognising the faces of those who had been closely involved in his recompression.

He slept well overnight with further recollections of the previous day's events. Full examination the next day was normal except for the 'visual disturbance' with continuing problems in "seeing faces as to who they were". As there was incomplete resolution of his symptoms he was recompressed using a Comex 12 hyperbaric oxygen (HBO) table (150 minutes at a maximum pressure of 222 kPa, 12 msw depth). Although apparently more relaxed after this he described faces as being indistinct around the mouth, nose and eyes, appearing "cartoon-like", and found recognising people by their faces as being difficult. He realised that he was recognising people by their voices or the glasses they were wearing rather than by their faces.

The following day his condition remained unchanged and so he was referred to the specialist neurology unit in Glasgow for assessment and CT scanning. Prior to transfer he underwent a further Comex 12 HBO treatment, with seemingly complete resolution of his ongoing problem. Clinical assessment by the neurologist found him to be completely normal and likewise the CT scan they carried out was normal. He remained well and was discharged the following day to his home in Glasgow. He returned the following day to the pier in Oban to welcome back the dive boat after completion of the trip, showing his colleagues that he had recovered completely and thanking them for their life-saving help.

JP was contacted seven years after his incident and was found to be in good health with no ongoing problems and without any return of his prosopagnosia. He had returned to diving two months after his incident, though he had restricted his diving to 30 metres or less. There had been no further episodes of decompression illness and he remained a happy man following this near-death experience.

In summary, following a rapid ascent from depth with omitted decompression, this man suffered a near-fatal arterial gas embolism (AGE) with cardiorespiratory arrest, after running out of air. He had neurological abnormalities as a result, which all resolved with recompression, including a visual problem diagnosed as prosopagnosia.

Discussion

Anyone suffering an AGE will have a rapid onset with variation in symptoms. Five per cent of patients suffering a catastrophic AGE will experience apnoea, unconsciousness and cardiac arrest immediately and many are unresponsive to CPR.[5] Having survived the initial event, with successful CPR in this case, the most appropriate assistance was sought with intravenous fluid resuscitation and high-flow oxygen and rapid evacuation. Recompression was commenced three hours after surfacing with apparent rapid resolution of symptoms. The resultant visual problem of prosopagnosia became apparent only during the time following the patient's first treatment, but completely resolved after two further HBO treatments.

Agnosia is "*the loss of the ability to recognise objects, persons, sounds, shapes or smells while the specific sense is not defective, nor is there a specific memory loss*." Usually agnosia is associated with brain injury or neurological illness, particularly after damage to the temporal lobe. Specifically

there is a group of visual agnosias, with prosopagnosia being the inability to recognise faces. The name originates from the Greek '*prosopon*' – 'face' and '*agnosia*' – 'non-knowledge'. It was described by both Jackson and Charcot in the 19th century but the term was first used by Bodamer in 1947.[6]

Patients suffering from prosopagnosia have the ability to recognise all objects except faces, including their own when viewed in a mirror. The patient recognises people instead by their clothing, hair colour, body shape, voice, spectacles, etc. There have been some peculiar cases of prosopagnosia reported: for example, a dairy farmer who developed prosopagnosia lost the ability to recognise the individual cows in his herd and a prosopagnosic bird-watcher reported "all birds look the same".[7] Evidence shows that prosopagnosia is usually associated with extensive damage to the temporal and occipital lobes, particularly in the region of the right occipitotemporal or fusiform gyrus.[8] Recent research, however, suggests that there are individuals who may also have a congenital or heritable cause.[9]

Early researchers of agnosias theorised that visual agnosia was the result of a reduction in low-level visual processing with impairments to mental abilities. This was often termed the sensory-deficit account.[10] There were, however, problems with this theory and the more recent "peppery mask" account offers a better explanation.[11] This theorises that the presence of random visual noise is caused by obstructing air bubbles circulating in the blood or the presence of blood clots in an intact blood vessel. Such impairment makes it difficult to organise information in its totality. It is highly probable that, in this case, the bubbles created by the embolism and the resultant tissue reaction have affected the area of the brain that supports the function of face recognition. This supposition is supported by the resolution of the visual disturbance following the third HBO treatment, after which it might be expected that any remaining bubbles would have been significantly reduced in size or eradicated and the tissue reaction improved or resolved.

Oliver Sacks, the eminent neurologist, wrote a short story entitled "*The man who mistook his wife for a hat*" about a music professor with a visual agnosia and prosopagnosia.[12]

"*Sometimes a student would present himself, and Dr P would not recognise his face. The moment the student spoke, he would recognise his voice … For not only did Dr P increasingly fail to see faces, but saw faces when there were no faces to see; genially, Magoo-like, when in the street, he might pat the heads of hydrants and parking meters, taking these to be the heads of children; he would amiably address carved knobs on furniture and be astounded when they did not reply.*

He also appeared to have decided that the examination was over and started to look round for his hat. He reached out his hand and took hold of his wife's head, tried to lift it off, to put it on his own head. He had apparently mistaken his wife for a hat! His wife looked as if she was used to such things."

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Postscript concerning St Kilda

St Kilda is currently one of the few double World Heritage sites in the world.[1] The fragile community that inhabited these islands for 4,000–5,000 years lived by a system of what was described as feudal communism. The community was sustained predominantly by obtaining food by scaling the cliffs and sea stacks and collecting seabirds and their eggs. The inhabitants had little or no contact with the outside world until the 18th century when curious visitors made their way there, unfortunately bringing diseases unknown to the local inhabitants. In 1724 smallpox reduced the population of St Kilda from 124 to 30. Between 1830 and 1843, 80% of the babies born on the islands died of neonatal tetanus. In 1852, 36 of the islanders emigrated to Australia and, although 20 died on the voyage, the suburb of St Kilda in Melbourne was established. On St Kilda, no-one paid taxes, no-one was registered to vote and no-one was called up to the armed services; they were left to their own devices. After petitioning the UK government in 1930, the remaining 36 islanders were evacuated to the Scottish mainland, as life had become unsustainable on St Kilda.[2]

Today, St Kilda is owned by the National Trust for Scotland (NTS), who maintain a small presence there in the summer months. St Kilda has one of the most extensive groups of vernacular building remains in Britain.[3] The layout of the 19th-century village remains to this day and over 1,400 stone-built cleitean for storing food and fuel are scattered all over the islands, and even on the sea stacks. The Ministry of Defence lease part of the islands and maintain a missile tracking station there. The islands have few visitors, from yachtsmen, diving groups and volunteer work parties for the NTS. Diving at St Kilda is considered to be the finest in the UK with clear water of 50–60 metre visibility, sea caves and arches, and interesting marine life, though the diving there is challenging because of the depths of the dives and the near continuous sea swell and currents.

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