Review

Acute loss of vision—an uncommon presentation of decompression sickness (DCS). A case report and review of literature

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Abstract

Decompression Sickness (DCS) results from gas bubble formation in the tissues of individuals who undergo a reduction in ambient pressure without adequate time to eliminate excess inert gas from body. These gas bubbles cause vascular/cellular insufficiency which is responsible for all types of clinical manifestations. DCS occurs in divers and also in those who work in compressed-air as in caissons and tunnels. It can also result from a reduction of normal barometric pressure, such as breathing in an unpressurized aircraft at high altitude. DCS has been classified as Type I, Type II and Type III named as Arterial Gas Embolism (AGE); in Type I, Limb and joints pain is present; while in Type II, systemic symptoms or signs, caused by the involvement of the CNS or Cardiopulmonary systems are dominant. Divers usually present with Type I DCS which can be easily recognized because of the symptom of joints pain while the neurological symptoms of Type II DCS mostly are not recognized early, while in case AGE patient have almost similar symptoms as in Type II DCS but these symptoms are more severe and rapid in onset. Neurological manifestations of DCS comprise symptoms from Cerebral hemisphere, the spinal cord, vestibular, retinal/optic nerve involvement. Retinal or optic nerve involvement results in monocular loss of vision, homonymous hemianopia, nystagmus, papilledema and rarely loss of vision. But fortunately with Hyperbaric oxygen therapy (HBOT) the recovery is rapid and complete.

Keywords: Acute vision loss, Hyperbaric Oxygen Therapy (HBOT), Decompression Sickness (DCS), Central Retinal Artery Occlusion.

INTRODUCTION

Since 4500 BCE, humans have engaged in free (breath-hold) diving to obtain food and substances from shallow ocean floors at depths of 100 ft or more. The 2007 record-setting breath-hold unlimited dive of Herbert Nitsch to 702 ft (214 m) attests to this human feat (AIDA International. World Records, 2014). However, as a testament to physical limitations, in 2012, when he tried to break his own record by diving to 819 ft (250 m), he suffered a narcosis blackout on ascent, causing a violation of his safety and decompression plan. As a result, he suffered a severe Type II neurologic DCS that ended his record-breaking career (John and Herbert, 2014).

Humans began experimenting with crude diving bells
as early as 330 BCE. These bells were submerged containing only air. In 1690, the first diving bell with a replenishing air supply was tested. The first crude underwater suit dates back to 1837, and helium was first used in place of nitrogen in 1939 (Stephen and Joe, 2014).

All these early diving methods required a physical connection to a support platform or boat. The Aqua-Lung, developed by Cousteau and Gagnon, and the submarine escape appliances, developed by Momsen and Davis, in the 1930s, were forerunners of the self-contained underwater breathing apparatus (SCUBA), which frees divers from the limitations of tethering.

The increasing popularity of scuba diving and the growth of commercial diving have increased the frequency of deep-pressure injuries (Stephen and Joe, 2014).

DCS does not occur in divers alone rather the workers who work in compressed air as in caissons and tunnels can also be affected. It can also result from a reduction of normal barometric pressure, such as in hypobare chamber and in aircraft at altitudes in excess of 5000 meters even when oxygen is breathed (Jains, 2006). Emergency physicians worldwide should know the physiologic effects and management of decompression sickness (Stephen and Joe, 2014).

Pathophysiology

Decompression sickness (DCS), is one form of dysbarism, which is a general term applied to all pathological changes that occur in the body secondary to rapid alteration in environmental pressure (Jains, 2006).

Changes in pressure affect only compressible substances in the body. The human body is made primarily of water, which is noncompressible; however, the gases of hollow spaces and viscous organs, and those dissolved in the blood, are subject to pressure changes. The principal component is most usually nitrogen (Stephen and Joe, 2014).

At sea level almost 1 Litre of Nitrogen is dissolved in the body. A little less than half of this is dissolved in water and a little more than one half in the fat, which constitutes only 15% of the normal male body. Nitrogen is five times more soluble in fat than in water (Jains, 2006). So as the individual descends under water, an increasing amount of nitrogen dissolves in the blood. Over time, increasing amounts of nitrogen dissolve and accumulate in the lipid component of tissues. When a critical amount of nitrogen is dissolved in the tissues, ascending too quickly causes the dissolved nitrogen to return to its gas form while still in the blood or tissues, causing bubbles to form (Stephen and Joe, 2014).

If the bubbles are still in the tissue, they can cause local problems; if they are in the blood, embolization may result. Children, pets, and even scuba divers watch and play with bubbles. However, when bubbles are inside, such as a trapped gas bubble in the intestine or stomach, the results are uncomfortable. This is even truer for divers (Stephen and Joe, 2014).

Not only does the quantity and size of the bubbles matter, but the type of reactions these bubbles cause is important as well. Location is also important. If bubbles end up in the lung and are not too large, they simply get filtered and exhaled. However, if a right to left shunt is present, such as from a patent foramen ovale, they bypass the natural filtering effect of the lungs and continue on to the brain or other organs. Nitrogen bubbles are believed to start as minute gas nuclei, present before the dive, rather than from supersaturation of the blood and tissues that acts as the seed for large bubble formation (Blatteau et al., 2006). All divers have bubbles (Thom et al., 2012). However, few divers that develop DCS. Thus, more than bubbles have to be involved. The presence of bubbles alone does not increase DCS (Møllerløkken et al., 2011). Microbubbles precede larger venous gas emboli (Swan et al., 2014). These emboli can occlude blood flow in smaller vessels and cause direct ischemia and damage. Bubbles have also been found to alter vascular endothelium through adhesion-molecule-mediated endothelial activation, in addition to activating platelets. In neurological tissue, this leads to focal ischemia. The TREK-1 potassium channel mediates this effect in a neuroprotective manner (Vallee et al., 2012; Bao et al., 2012).

Microparticles derived from vascular walls have been found to increase 3-4 times with dives and decompression stress. They appear to activate neutrophils and interact with platelet membranes (Thom et al., 2012; Thom et al., 2013). Endothelial cells, blood platelets, or leukocytes shed microparticles upon activation and cell apoptosis (normal programmed cell death). In particular, the release of platelet microparticles could reflect bubble-induced platelet aggregation. This could be the cause of coagulation and thrombosis, thus interfering with blood flow (Pontier et al., 2012). Once the bubbles form, they create a foreign body interface to which platelets then adhere (Philp et al., 1975). In severe DCS, significant decreases in platelet count have been documented. These decreases may someday be used as a marker for severity of injury (Pontier et al., 2008). Endothelial nitric oxide synthesizes produces nitric oxide through the combination of arginine and oxygen. It is a powerful vasodilator which, through relaxation of smooth muscles, inhibits platelet aggregation and inhibits inflammation. The combination contributes to blood vessel homeostasis. The presence of nitric oxide may reduce bubble formation (Blatteau et al., 2013; Taylor, 2014). However, the increasing partial pressure of oxygen at depth drives the reaction towards nitric oxide. Once the body’s natural processes for dealing with
oxidizers, which this is, are overwhelmed, it yields an excess of oxidative, excitatory neurotransmitters (Taylor, 2014). Nitrogen dioxide, a nascent gas nucleation site synthesized in some microparticles, initiates decompression inflammatory injury (Thom et al., 2013). It is also an oxidizer that exists in equilibrium with dinitrogen tetroxide (Taylor, 2014).

There appears to be a relationship among bubbles, microparticles, platelet-neutrophil interactions, and neutrophil activation. However, exactly what that relationship is still remains obscure (Blatteau et al., 2013; Lemaitre et al., 2009).

Clinical manifestations of DCS

Results from the effects of these bubbles on organ systems. Clinical manifestations of DCS are mostly due to skin, musculoskeletal and central nervous system(brain and spinal cord) involvement. The initial symptoms started within 6 hours of surfacing in 99% of cases with an overall mean delay to onset of 62 minutes. The shorter the time to onset, the more serious the symptoms (Xu et al., 2012).

DCS may be divided into 3 categories: (1) type I (mild), (2) type II (serious), and (3) arterial gas embolization (AGE).

Type I decompression sickness

Type I DCS is characterized by one or a combination of the following: (1) mild pains that begin to resolve within 10 minutes of onset (niggles); (2) pruritus, or "skin bends," that causes itching or burning sensations of the skin; and (3) cutis marmorata, a skin rash which generally is widespread mottling and/or marbling of the skin, or a papular or plaque-like violaceous rash (Oode et al., 2013).

Pain (the bends) occurs in most (70-85%) patients with type I DCS. Pain is the most common symptom of this mild type of DCS and is often described as a dull, deep, throbbing, toothache-type pain, usually in a joint or tendon area but also in tissue. The shoulder is the most commonly affected joint. The pain is initially mild and slowly becomes more intense. Because of this, many divers attribute early DCS symptoms to overexertion or a pulled muscle. Upper limbs are affected about 3 times as often as lower limbs (Stephen and Joe, 2014).

Type II decompression sickness

Type II DCS is characterized by the following: (1) pulmonary symptoms, (2) hypovolemic shock, or (3) nervous system involvement. Pain is reported in only about 30% of cases. Because of the anatomic complexity of the central and peripheral nervous systems, signs and symptoms are variable and diverse. Symptom onset is usually immediate but may be delayed as long as 36 hours (Stephen and Joe, 2014).

Nervous system

The spinal cord is the most common site affected by type II DCS; symptoms mimic spinal cord trauma. Low back pain may start within a few minutes to hours after the dive and may progress to paresis, paralysis, paresthesia, loss of sphincter control, and girdle pain of the lower trunk (Hennedige et al., 2012).

Eye

When DCS affects the brain, many symptoms can result. Negative scotomata, devoid of any lights or shapes, are the earliest symptom. Negative scotomata become positive after a few minutes. Other common symptoms include headaches or visual disturbances, diplopia, photophobia, optic neuropathy, branch retinal artery occlusion, and loss of vision (Latham et al., 2008).

Ear

Labyrinthine DCS (the staggers) causes a combination of nausea, vomiting, vertigo, and nystagmus, in addition to tinnitus and partial deafness (Huang et al., 2003). In inner ear DCS (IEDCS), vertigo was the major presenting complaint in 77-100%. Hearing loss occurred in 6-40% and a combination of both in 18%. Additional skin and neurologic symptoms were present in 15%. Symptoms occurred within 120 minutes of surfacing with a median delay of 20 minutes (Klingmann, 2012; Gempp and Louge, 2013).

Lungs

Pulmonary DCS (the chokes) is characterized by the following: (1) burning substernal discomfort on inspiration, (2) nonproductive coughing that can become paroxysmal, and (3) severe respiratory distress. This occurs in about 2% of all DCS cases and can cause death. Symptoms can start up to 12 hours after a dive and persist for 12-48 hours (Stephen and Joe, 2014).

Arterial gas embolism

Pulmonary over pressurization/pulmonary barotrauma can cause large gas emboli while a rupture into the
pulmonary vein allows alveolar gas to enter systemic circulation (Rudge, 1992). Gas emboli can lodge in coronary, cerebral, and other systemic arterioles. These gas bubbles continue to expand as ascending pressure decreases, thus increasing the severity of clinical signs. Symptoms and signs depend on where the emboli travel. Coronary artery embolization can lead to myocardial infarction or dysrhythmia. Cerebral artery emboli can cause stroke or seizures (Stephen and Joe, 2014).

Differentiating cerebral AGE from type II neurologic DCS is usually based on the suddenness of symptoms. AGE symptoms typically occur within 10-20 minutes after surfacing. Multiple systems may be involved. Clinical features may occur suddenly or gradually, beginning with dizziness, headache, and profound anxiousness. More severe symptoms, such as unresponsiveness, shock, and seizures, can quickly occur. Neurologic symptoms vary, and death can result. DCS of the CNS is clinically similar to AGE (Huang et al., 2003).

Patients with mild type I DCS probably do not require treatment other than breathing pure oxygen at sea level for a short time. Divers with type I DCS symptoms do however, require close observation, as symptoms may portend the onset of more serious problems requiring hyperbaric recompression. Consult a diving medicine or HBO specialist for all diving-related injuries. The only effective treatment for gas embolism is recompression; other treatments are merely for symptoms (Stephen and Joe, 2014).

The basic theory behind HBO therapy is, first, to repressurize the patient to simulate a depth where the bubbles from nitrogen or air are redissolved into the body tissues and fluids. Then by breathing intermittently higher concentrations of oxygen, a larger diffusion gradient is established. The patient is taken slowly back to surface atmospheric pressure. This allows gases to diffuse gradually out of the lungs and body. The addition of helium to oxygen has been shown to yield an advantage over oxygen alone even in severe neurologic DCS or treatment-refractory DCS (Goldenberg et al., 1996; Shupak et al., 1997).

Acute DCS is a purely clinical diagnosis that requires a fair amount of clinical suspicion. When the slightest suspicion or possibility is noted, timely transfer with 100% oxygen for HBO should be pursued (Hennedige et al., 2012).

**Reported case**

18 years-old male new diving cadet from Marine diving school, dived in sea using air at a depth of 25 feet sea water at 8:05 a.m. with bottom time 5 minutes and total dive time 7 minutes. After surfacing (ascent with control) he complained of severe pain and tearing in both eyes. He was treated by eye drops as immediate remedy. At 6:00 p.m. this driver was scheduled to dive again at same depth (25 fsw for 30 minutes). He dived but just able to make it to 12 fsw. He ascended again upon experiencing the same severe pain in his eyes and just finished the 30 minutes swimming time on sea surface. When he reached ashore, he could see at a distance of approximately 10 meters only (more or less). After about 10 minutes, he told that he cannot see at all which prompted us to bring him to the hospital.

He was assessed by ER physician, ophthalmologist and Diving Medical Specialist and it was found that he had 0/6 vision in both eyes with no pupillary reflex to light while ophthalmoscopic examination was done without dilating the pupils and that was insignificant.

The only suspicion was DCS Type II or Arterial Gas Embolism. So It was then decided by Diving Medical Specialist to recompress him starting at USN TT6 (United State Navy Treatment Table 6). Reassessment was again made upon reaching bottom within 10 minutes but he had still no improvement. He was recompressed further to USN TT 6A at that time, in which upon reaching the bottom 165 fsw he started to regain his vision in no time. The table was followed until completed without extension. Patient vision almost completely recovered. He was also assessed by the ophthalmologist and his visual acuity was 6/6 with normal Retina and normal color vision. He was admitted for 24hrs and discharged after reassessment from Ophthalmologist.

So it was proved that Diver was having Type II DCS/ Cerebral Arterial Gas Embolism (CAGE) which responded to Hyperbaric/Recompression Therapy.

**DISCUSSION**

DCS is rare unless patient has been exposed to pressures greater than 2 ATA although cases have been described from long exposures to lesser pressures. In surface decompression procedures Type II DCS is more common and on deep dives presents more frequently than Type I symptoms. Type I DCS is easy to recognize because of joint pain while symptoms of Type II DCS involving CNS can be easily missed (Jains, 2006).

Neurologic manifestations of DCS comprise symptoms from cerebral hemisphere, the spinal cord, vestibular system as well as visual disturbances. Ocular symptoms may occur alone or in combination with other signs and symptoms. Rivera noted visual disturbances in 6.8% of DCS patients in his study of 935 cases and were the sole presenting symptom in 1.4% of cases (Rivera, 1964).

Vision loss in Type II DCS or in case of Arterial Gas Embolism (AGE) occurs as a result of Central Retinal Artery occlusion or Cerebral Ischemia. Visual symptoms associated with AGE include monocular loss of vision (typically from Central Retinal Artery Occlusion), homonymous hemianopia, nystagmus, papilledema, and
cortical blindness (Butler, 1995). In one report, ten of 91 cases of cerebral arterial gas embolism (CAGE) displayed visual abnormalities (Brooks et al., 1986). Gorman reported a similar 11.2% of cases of AGE displaying visual signs or symptoms (Gorman, 1978).

The Retina has dual blood supply that is from Central Retinal Artery (CRA) and the posterior Ciliary/choroidal Arteries. Both are the branches of ophthalmic Artery which in turn is the branch of the Internal Carotid Artery. The long posterior ciliary arteries provide blood to the choroid and the outer layers of the retina while the retinal artery supplies the inner layers of the retina. Normally, the choroidal circulation supplies the majority of the oxygen to the retina, and only the inner layers of the retina are oxygenated from the retinal circulation. Under normoxic conditions, approximately 60% of the retina’s oxygen supply comes from the choroidal circulation (Butler et al., 2008).

The sign and symptoms of CRA occlusion include painless loss of vision in the affected eye. The ophthalmoscopic findings with Central Retinal Artery occlusion include a pale disc with attenuated arterioles and veins, a cloudy retina and a “cherry red” macula. Even though the vision is lost in seconds the retinal cells are resistant to ischemic death and may survive for as long as 22min if both the retinal and the choroidal circulations are occluded. This is due to the ocular reservoir of oxygen (vitreous) and the high rate of anaerobic glycolysis that occur in the visual cells. Conventional therapy of CRA occlusion include:

a) Anterior Chamber paracentesis
b) Retrobulbar anaesthetic block
c) Anticoagulation

But it was found that none of these methods are satisfactory, till 1984 when 2 patients were treated with HBOT and they showed excellent recovery even after 6 hrs of occlusion (Jains, 2006). Healthyprimate can tolerate vascular shutdown for approx. 90 minutes before permanent damage occurs. Institution of HBOT in this interval may allow preservation of inner retinal function as manifested by maintenance of vision and normal retinogram. If the choroid can supply oxygen to the retina the HBOT should be useful in the treatment of patient with the occlusion of CRA (Jains, 2006).

The therapeutic benefit of HBOT in AGE is believed to result from one or more of the following effects:

1) mechanical reduction of the volume of the gas emboli; 2) reduction of the partial pressure of nitrogen in the blood, thereby increasing the resolution speed of the gas bubble; 3) increasing the oxygenation of hypoxic neural tissue; and 4) decreasing postembolic cerebral edema (Bitterman and Melamed, 1993).

Emergent HBOT as outlined in the U.S. Navy Diving Manual is recommended for AGE and has been shown to be effective in reversing the visual manifestations of AGE when undertaken promptly (Mitchell et al., 2000; Pao and Hayden, 1996). Treatment for presumed AGE should be administered even when there are significant delays to treatment.

The case which we reported, had acute loss of vision of both eyes, which is not the common presentation mentioned in literature and the bilateral loss of vision is rarely seen (Rivera, 1964). The point in favor of diagnosis of DCS/AGE was acute vision loss after diving and the diver had no illness before. Ophthalmologist in short time did eye examination but there was no specific finding related to DCS/AGE. So instead of wasting time in further advance investigations HBOT was started immediately because Recompression has been advocated as a definitive test for DCS provided it is undertaken immediately (Jains, 2006). That resulted in complete recovery and return of vision.

CONCLUSION

The diagnosis of DCS/AGE can be made on the basis of history and clinical features and it is essential to stress that if a significant dive has been undertaken then the presumption must be made that the symptoms are due to decompression sickness not natural disease and early Recompression Therapy is indicated.

REFERENCES


