Case report

Atypical distally distributed cutis marmorata decompression sickness associated with unconventional use of thermal protection in a diver with persistent foramen ovale

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Abstract

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Cutis marmorata is a mottled, marbling, livedoid rash caused by vascular inflammation and congestion in cutaneous decompression sickness. It may occur during or after ascent due to the formation of bubbles from dissolved nitrogen accumulated throughout the dive. It is strongly associated with the presence of right to left shunts, particularly persistent (patent) foramen ovale (PFO). We report a case of cutis marmorata decompression sickness of an unusual pattern associated with unconventional use of thermal protection (a 'shorty' wetsuit worn over full suit) by a diver with a PFO. The patient also had neurological manifestations of decompression sickness. The distal lower limb pattern of involvement favours the hypothesis that cutis marmorata in humans is likely to be due to bubbles in the skin itself and/or adjacent tissues rather than cerebrally mediated.

Introduction

Scuba and deep-water divers are at risk of decompression sickness (DCS) during or after ascent, due to the formation of bubbles from dissolved nitrogen accumulated throughout the dive. The presence of a persistent (patent) foramen ovale (PFO) allows paradoxical gas embolism, whereby bubbles cross from the venous circulation directly into the arterial circulation, bypassing lung filtration. These bubbles then grow upon reaching vulnerable supersaturated tissues. Intravascular bubbles cause vascular inflammation and congestion which manifests as a myriad of symptoms and signs, one being a rash characterised by the terms cutis marmorata or livedo racemosa;[1–3] a cutaneous discolouration with characteristic persistent, erythematous or violaceous change, in a broken net-like pattern. The typical mottled, marbling, livedoid rash of cutis marmorata often crosses the midline and is seen in areas rich in subcutaneous fat, namely the chest and abdomen,[4] bilateral thighs,[5] back, buttocks, breast and upper arm.[1]

While there is consensus that bubble formation secondary to dissolved inert gas is involved in the pathogenesis of cutis marmorata DCS, further details pertaining to the pathophysiology remain a bone of contention amongst researchers. There are three main hypotheses for the formation of cutis marmorata:

1. Paradoxical embolism of venous bubbles across a right-to-left shunt followed by augmentation in bubble size on arrival at supersaturated skin via the arterial circulation.[3]
2. *In situ* bubble formation at the skin itself or within its circulation.[1,6]
3. Entry of bubbles to the arterial cerebral or brainstem circulation resulting in a sympathetically mediated vasomotor response.[7]

Many case reports of cutaneous DCS involving the legs state that the typical rash does not extend below the level of the patella.[4,8–10] To our knowledge, it is rare and/or underreported that a cutaneous DCS rash involves the shins. In this case there was an unusual distal rash distribution associated with an unconventional pattern of thermal protection worn by the diver. This may give further insight into the pathophysiology of DCS, in particular cutaneous DCS.

Case report

The patient gave written consent for her case details to be reported.

A 34-year-old previously healthy lady with a history of 35 uneventful dives performed the following two dives on open circuit air. During both dives she wore a 3 mm 'shorty' wetsuit over a 5 mm full suit.

1. Dive 1: maximum depth 38.1 metres of sea water (msw), absolute bottom time (ABT) 12 min and total time of dive (TTD) 32 min followed by a surface interval (SI) of 1 hour 43 min. The water temperature was 17ºC.
2. Dive 2: maximum depth 37.8 msw, ABT 12 min and TTD 44 min, water temperature 17ºC. She noted an erythematous, lacy rash present below the knees bilaterally which improved on surface oxygen. She therefore did not seek medical advice.

The following day she performed another two dives on open circuit air using the same wetsuit configuration.

1. Dive 1: maximum depth 31.3 msw, ABT 27 min and TTD 56 min followed by a SI of 1 hour 31 min, water temperature 20ºC.
2. Dive 2: maximum depth 30.4 msw, ABT 19 min and TTD 46 min, water temperature 24ºC.

All dive profile information (Figure 1) was sourced from the diver's Ratio iDive dive computer (Buhlmann ZHL-16 B decompression algorithm). The dive profile analysis included the computation of the surfacing gradient factor (GF) values while taking into account residual gas loadings, which for the four consecutive dives yielded: 0.64, 0.60, 0.88, and 0.83 respectively.

For all dives, the diver used a 5 mm full wetsuit. On top of the full suit she also wore a 3 mm 'shorty' which terminated just above her knees and elbows. Five minutes after surfacing from the second dive she claimed to have rushed to get an oxygen cylinder for another diver after which she complained of sudden bilateral thigh pain which progressed to both arms. She also noted lethargy and according to her dive buddy an erythematous, lacy rash was present on both legs below the lower third of the thigh and lower back.

Upon review at the emergency department, the diver had a good volume regular pulse at 70 beats per minute and a blood pressure of 110/52 mmHg. She had a normal cardiorespiratory examination as well as normal power and reflexes in both upper and lower limbs. Gait was normal, however the patient complained of dizziness and nausea on walking. No pronator drift, nystagmus or cerebellar signs were observed but a Romberg's test was positive. The rash was noted on both shins and over the extensor surface of the right knee and the distal right thigh. It consisted of flame-shaped erythematous, blanching, papular lesions (Figures 2 and 3). It did not involve the proximal thighs, abdomen, breasts or trunk. Blood investigations showed no abnormalities apart from neutrophilia (18.11 cells x10^{9}·L^{-1}).

Figure 1

Dive profile data uploaded from Ratio iDive diving computer. A: Dive 1 on day 1; B: Dive 2 on day 1; C: Dive 1 on Day 2; D: Dive 2 on Day 2. msw: metres sea water

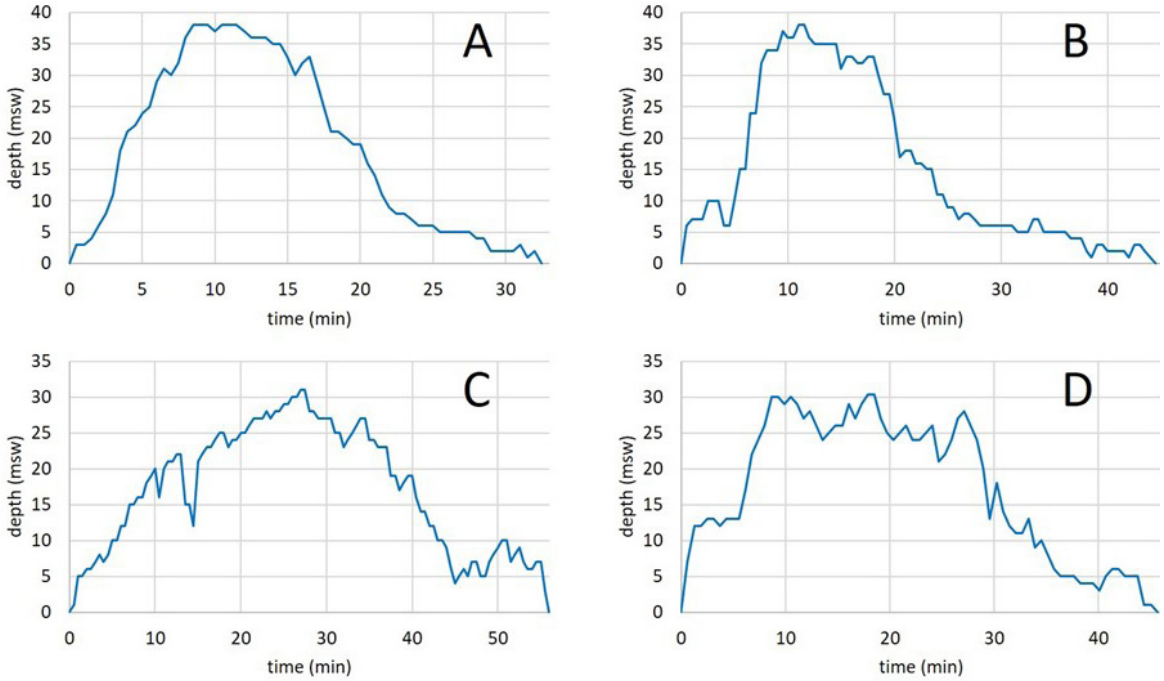


Figure 2
Pre-treatment; right distal thigh and knee with cutis marmorata rash visible

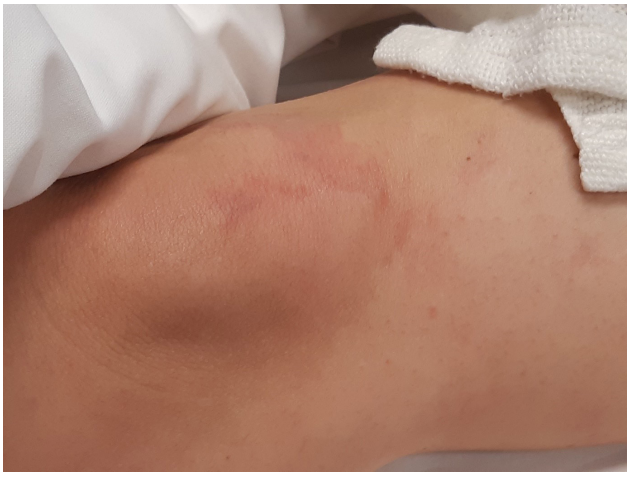


Figure 3
Pre-treatment; left shin bruise and cutis marmorata rash visible

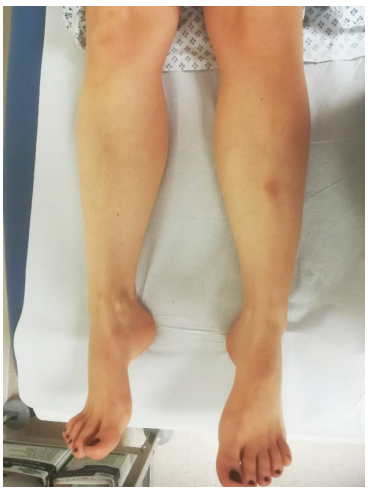
Hartmann's solution and normal saline were given, together with 100% oxygen via demand-valve mask. The patient was treated on US Navy treatment table 6 (USN TT6), initiated on oxygen at 284 kPa. After around 15 minutes, the rash improved and the patient was able to walk around the chamber during the second air break without any dizziness or nausea. A negative sharpened Romberg test was elicited.

The patient was sent home, where she later noted a return of the bilateral thigh pain and lethargy. On examination the following morning, a sharpened Romberg test was positive. The left shin manifested an oval shaped, erythematous, blanching, macular lesion measuring approximately 4 cm by 2 cm (Figure 4). She received follow-up hyperbaric treatment (Royal Navy 60).

Upon review a day later, the patient reported feeling much improved. She reported mild generalised aches and pains in both thighs, left shoulder and upper arm not requiring analgesia. Her gait was normal, a sharpened Romberg test was negative, and she had full power in both arms and legs.

A further follow-up hyperbaric treatment was given (US Navy treatment table 9) with further improvement of symptoms. The residual left shin oval-shaped macular lesion showed skin colour changes consistent with that of a bruise. A transthoracic echocardiogram with agitated saline bubble contrast study was carried out four weeks later. Right–to-left shunting of bubbles was observed both at rest as well as after a Valsalva manoeuvre, suggestive of a PFO. The examination was otherwise within normal limits.

Figure 4
Post-treatment; left shin bruise still visible, no further cutis marmorata rash



Discussion

We describe a case of cutis marmorata DCS demonstrating an atypical distal distribution of the irregular, mottled, erythematous rash in a patient with a right-to-left shunt. The rash was observed over both shins and the extensor surfaces of the right distal thigh and knee. The thermal protection employed by the diver consisted of a 3 mm 'shorty' neoprene wetsuit worn over a 5 mm neoprene full wetsuit. Analysis of the diving profiles indicated that the dives on the day of the incident were provocative, reaching a surfacing GF of 0.88. A recent study analysing Divers Alert Network (DAN) database diving data reported maximum GF-values of 0.70–0.90 in the majority of DCS cases.[11]

Published studies demonstrate that the diving wetsuit exerts various effects on human body physiology. In a non-immersed study, wetsuit wearing was associated with decreases of heart rate and cardiac output, vital capacity and expiratory reserve volume, possibly secondary to the compressive effect exerted on the chest.[12] A tight-fitting wetsuit increases mean arterial pressure and affects heart rate variability resulting in a decrease in the LF/HF ratio

(low and high frequency ratio).[13] It may also compress deep limb veins and peripheral blood vessels, resulting in central pooling of venous blood and an increase in cardiac preload.[14] Wetsuit use has been suggested as a predisposing factor for swimming induced pulmonary oedema[15] and it influences hydromineral homeostasis resulting in increased urine output both during scuba diving and in dry conditions.[16]

To our knowledge, there are no studies linking patterns of wetsuit use to specific forms of DCS. We hypothesise that in our diver, at some point during the scuba dives, the shorty neoprene wetsuit exerted a tourniquet effect at the level of the distal third of the thighs. It appears plausible that the distribution of the rash was a result of impaired off-gassing of inert gas due to reduced perfusion in the distal thigh and leg tissues during ascent.

The tourniquet effect may have been established the moment the wetsuits were worn on the surface. However, as a diver descends, bubbles in the neoprene rubber are compressed, resulting in a looser wetsuit during the deep part of the dive and probable reversing of the tourniquet effect. During experimental research, Bühlmann observed that if limb circulation, for example of an arm, is compromised by the cuff of a sphygmomanometer during decompression that is critical for the skin, red spots and swelling may develop on the arm.[17] In this patient, reversal of the tourniquet effect in the early phase of the dive due to neoprene compression may have permitted sufficient on-gassing of distal thigh and leg tissues. During ascent, a return of the tourniquet effect in the shallows may have impaired off-gassing of the distal lower limb tissues. Moreover, enhanced levels of vasoconstriction due to cold exposure towards the latter stages of the dive may have further hampered off-gassing of inert gas during ascent. Cold exposure during diving is one of the mechanisms which contributes to the dive reflex and results in peripheral vasoconstriction.[18,19]

It is well-established that cutis marmorata DCS is associated with an intra-cardiac right-to-left shunt.[2,3,6] Several mechanisms for its pathophysiology have been proposed and have been mentioned above. There has been debate on whether the rash could also be cerebrally mediated following a study in swine. Internal carotid artery gas injection in anaesthetised pigs was followed by the appearance of a cutis marmorata mottled rash similar in appearance to cutis marmorata / livedo racemosa.[7] Notably, the 30–40 kg pigs were injected with 0.25 and 1 ml·kg^{-1} air producing a sympathetic surge similar to that noted in subarachnoid haemorrhage and other catastrophic brain injuries. Most human patients with cutaneous DCS have no neurological symptoms and if they do, they are rarely catastrophic.[20] Moreover, a recent human study using ultrasound technique confirmed the presence of bubbles in skin microcirculation affected by cutis marmorata.[1] This adds more weight to the mechanism of cutis marmorata in human cases being non-cerebrally mediated.

The case described in this paper illustrates that bubble amplification is likely occurring in the skin and surrounding tissues themselves, rather than the rash being cerebrally mediated. It is thought that the unusual use of thermal protection resulted in supersaturated tissues distal to the level of the shorty wetsuit. This resulted in either in situ bubble formation or local augmentation of paradoxical gas emboli arriving in the distal lower limbs.

Of note, this diver also complained of dizziness, observed at the emergency department shortly after gait assessment. While this symptom may have been due to non-documented postural hypotension, the Romberg's test was positive indicating likely vestibular DCS which is also strongly associated with a PFO.[21] These findings are further in-keeping with paradoxical air embolism with subsequent bubble enlargement at the legs as the most likely mechanism of cutis marmorata formation in this case.

Conclusions

Cutaneous DCS is a well-documented condition strongly associated with the presence of a right-to-left shunt. The distal lower limb pattern of involvement and other circumstances of this case favour the hypothesis that cutis marmorata in humans is likely to be due to bubbles in the skin itself and/or adjacent tissues rather than cerebrally mediated. It illustrates how thermal protection employed by the diver may influence the distribution of a cutis marmorata rash in a diver with a right-to-left shunt.

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