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CASE REPORT

Vasculitis masquerading as neurologic decompression illness

P. J. BENTON and R. W. SMITH

Institute of Naval Medicine, Alverstoke, Gosport, Hampshire PO12 2DL, and Royal Naval Hospital Haslar, Gosport, Hampshire PO12 2AA, United Kingdom

Benton PJ, Smith RW. Vasculitis masquerading as neurologic decompression illness. Undersea Hyperbaric Med 1996; 23(3): 189–191.—An adult male diver developed limb pains and peripheral neurologic signs following a series of dives. He was treated for acute neurologic decompression illness (DCI) and responded well, but subsequently he deteriorated and developed features of a progressive multisystem disease. Investigation confirmed a clinical diagnosis of polyarteritis nodosa complicated by a mononeuritis multiplex. Vasculitis is uncommon and its masquerade as neurologic DCI may be unique. However, this case empha-sizes the importance of careful clinical assessment and illustrates the potentially wide differential diagnosis of DCI.

polyarteritis nodosa, vasculitis, decompression illness, hyperbaric oxygen therapy

A 27-yr-old military diver carried out a routine series of three dives, each of 8- to 10-min duration over a 2-h period, in a diving tank 15-m deep, water temperature 21°C,. The dives, breathing air, were uneventful and included a controlled ascent on each occasion. Approximately 30 min after completion of the final dive, the diver noticed paresthesia affecting the ring and little fingers of his right hand which he attributed to a tight wrist seal. However, despite removal of his suit these symptoms persisted and began to extend proximally. Following the development of pain and weakness in his right arm and hand, medical advice was sought.

RESULTS

Four hours after the last dive, examination revealed marked weakness affecting the ulnar innervated intrinsic muscles of his right hand (2/5) and an area of reduced sensation approximating to the right C8 dermatome. Equivocal weakness of other muscle groups of the right arm was difficult to interpret in the presence of severe wrist and elbow pain. The painful joints had a full range of movement, were not tender on palpation, and the intensity of pain was not affected by position. The only other positive finding was elevated blood pressure (160/100). Past medical history was unremarkable. The dive profiles were of low risk but, in view of the timing and pattern of the clinical features, a diagnosis of acute neurologic decompression illness (DCI) could not be discounted, and urgent transfer to a recompression facility was arranged.

Hyperbaric oxygen (HBO_2) treatment, commencing with a Pi_{O_2} of 2.8 atm abs (RN table 62, USN table 6) (1), was associated with a dramatic and rapid symptomatic improvement. On completion of HBO_2, residual abnormal signs were limited to the right hand, with altered sensation affecting the ring and little fingers and slight weakness (4+/5) of the ulnar innervated intrinsic muscles. However, 4 h after completion of HBO_2, some 14 h post dive, he became aware of paresthesia affecting both feet and recurrent symptoms affecting his right hand. Simultaneously, he developed dull central abdominal pain. Examination revealed no features to suggest an acute abdomen but did confirm a recurrence of the initial symptoms, weakness (2/5) of the intrinsic muscles of the right hand and reduced sensation in the ulnar nerve distribution, as well as reduced sensation of a stocking pattern affecting both feet. Equivocal weakness of the muscle groups of the lower limbs was difficult to interpret in the presence of the abdominal pain. Recurrence of neurologic DCI could not be excluded and so further HBO_2 was organized. On this occasion there was no symptomatic response or change in neurologic signs after two 25-min O_2 periods at 2.8 atm abs (RN table 60, modified Royal Adelaide table), and thus further HBO_2 was deemed inappropriate.

Over the ensuing 5 days he continued to experience infre-

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quent episodes of central abdominal pain, although repeated clinical assessments revealed only poorly localized tenderness. Results of preliminary investigations indicated an acute inflammatory response (white blood count = 31.3×10^9/liter, C-reactive protein = 100 mg/liter) and an unexplained eosinophilia (8.3×10^9/liter); an intensive search for infection was negative. During this period he became increasingly unwell and addi-tional problems evolved: low grade pyrexia, general malaise, anorexia, increasing hypertension (170/115), severe pharyn-gitis, myalgia, weight loss, subungual splinter hemorrhages, a fine erythematous rash, glove and stocking anesthesia, and left foot drop. Together, these multisystem manifestations suggested a clinical diagnosis of vasculitis, supported by findings at visceral angiography which demonstrated aneu-rysms of medium-sized arteries typical of polyarteritis nodosa (PAN) (2).

Neurophysiologic studies revealed a mononeuritis multiplex. Marked, predominantly motor changes affected the right median nerve in the forearm in a pattern suggesting mixed axonal and demyelinating pathology, while axonal pathology was evident affecting left lateral popliteal and both ulnar nerves. This patchy and widespread distribution of asynchronous lesions, although not specific to any one disease, is well recognized in vasculitic involvement of the peripheral nervous system.

Despite early and aggressive immunosuppressive therapy, fulminant complications ensued, including a near catastrophic intestinal hemorrhage. At laparotomy an inflamed and necrotic segment of large bowel was identified and resected, histologic examination of which demonstrated a necrotizing vasculitis of medium and small arteries characteristic of PAN (3). Over the course of the following 2 yr he has made a remarkable and sustained recovery on immunosuppressive drugs, gradually reduced. Unfortunately there is a definite though diminishing risk that his disease will recur, and therefore he must at present be considered unfit for both diving and full military duties.

DISCUSSION

The term vasculitis is used to describe a group of conditions (2) characterized by focal and segmental inflammation of blood vessel walls often involving multiple organs, asynchronously (3). Pathologic changes include aneurysm formation, vessel wall rupture, and thrombosis in situ, resulting in local ischemia or infarction or both, often with dramatic clinical consequences (4–6). These varied manifestations and the associated systemic disturbance lack diagnostic specificity, and early identification thus requires a high index of suspicion. Current classification systems have evolved from the early work of Zeek (7) and are based largely on the size and distribution of the blood vessels affected. Precise diagnosis, in the appropriate clinical context, is determined from pathologic or angiographic study or both, and is supported by laboratory results. Polyarteritis nodosa involves medium-sized arteries predominantly and may affect a wide range of organs, the prognosis being especially poor in cases with intestinal involvement (8). Unlike many types of vasculitis, and excluding that associated with endemic hepatitis B infection, polyarteritis nodosa is becoming increasingly rare (9). Nevertheless, prompt diagnosis and aggressive therapeutic intervention are essential, and in this respect similarities exist with DCI (1).

It is well accepted that the onset of acute neurologic dysfunction following hyperbaric exposure is, until proven otherwise, due to DCI and that response to recompression therapy provides supporting evidence for the diagnosis. This case serves to illustrate that other conditions, although very uncommon, may on occasion closely mimic DCI, even in their response to HBO_2. In this case, the search for an alternative diagnosis was prompted by the atypical progression of neurologic symptoms and the development of other features incompatible with DCI. It may be that the initial symptoms were indeed due to DCI caused by abnormal inert gas transfer in tissues already hemodynamically compromised as a result of local vasculitis. The mechanism for the initial, almost complete resolution of symptoms following HBO_2 in this case is speculative. We postulate that local tissue hypoxia may have been ameliorated by the increased tissue P_{O_2} per se. Alternatively, an effect may have been due to the anti-inflammatory action of HBO_2 or, although unlikely in view of the timing of events, this may reflect the fluctuating nature of vasculitic disorders.

Vasculitis is uncommon and its masquerade as neurologic DCI may be unique. However, this case emphasizes the importance of careful ongoing clinical assessment and illustrates the potentially wide differential diagnosis of DCI.

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