

Ischemic monomelic neuropathy: an underappreciated cause of pain and disability following vascular access surgery

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ABSTRACT: The establishment and maintenance of a functioning arteriovenous access site is essential for the care of the hemodialysis (HD) patient. Ischemic monomelic neuropathy (IMN) represents an underappreciated complication and an unrecognized source of short- and long-term disability because of its protean manifestations in the post-operative period. Delayed diagnosis of IMN greatly reduces the chance of avoiding irreversible disability. Prompt diagnosis and early management of IMN can avoid morbidity for the patient, diminish healthcare costs for the system and avoid medical liability for care providers. A multidisciplinary awareness of the symptoms of IMN is essential to permit prompt diagnosis and management. (J Vasc Access 2010; 11: 00-00)

Key words: Ischemic monomelic neuropathy, Hemodialysis access, Steal syndrome, Vascular access, End-stage renal disease

INTRODUCTION

The steady increase in the incidence of treated end-stage renal disease (ESRD) and the associated costs are an urgent national public health concern. The number of new cases of ESRD in the United States is projected to be 650,000 by 2010, with accompanying Medicare expenditures of \$28 billion (1).

The successful establishment and maintenance of a functioning vascular access (VA) in the hemodialysis (HD) patient is essential to minimize patient morbidity and to control the healthcare costs associated with caring for patients who suffer from this chronic medical condition. Recognized complications associated with chronic VA sites include infection, access thromboses, aneurysm formation and VA associated tissue ischemia.

I present a case of ischemic monomelic neuropathy (IMN). An infrequently reported, and likely underappreciated cause of nerve injury, which may result in both short- and long-term patient disability. Beyond the immediate issues of prevention and patient care, the potential for a delayed or missed diagnosis of IMN raises concerns about medical liability. Better education and awareness of this underappreciated disorder among the multidisciplinary team of caretakers provides the best opportunity for early diagnosis and prompt management of IMN.

CASE

A 57-year-old female with long standing diabetes, hypertension and a prior VA history of an abandoned prosthetic upper arm straight graft in the left extremity.

Pre-operative work up of the right arm consisted of a vein mapping which demonstrated a patent basilic vein measuring 3-5 mm. No cephalic vein was identified. Venography revealed patent central veins. Brachial artery pressures demonstrate no arm to arm discrepancy. On examination, she had a palpable radial pulse. Neurologic examination of the extremity was remarkable for normal sensation and intact motor function.

A single-stage upper arm transposed basilic vein fistula was created. The procedure was uneventful and the patient was discharged the same day per our access surgery care pathway. At her initial 2 week post-operative appointment following surgery, she complained of forearm stiffness and numbness of the fingers with diminished grip strength. Her forearm was remarkable for generalized soft tissue swelling with no evidence of a hematoma within the upper arm wound. The symptoms appeared to be mild and likely related to the residual forearm swelling. The fistula had a strong thrill. Her radial pulse was palpable. Because of her palpable pulse - concerns about severe vascular steal syndrome (VSS) were minimal.

She was seen a week later with persistent and worsening complaints of distal forearm, hand and digit pain and numbness; despite the improvement of soft tissue swelling and no evidence of wound hematoma. She demonstrated diminished grip strength and altered sensation along the first, second and third digits. Her radial pulse remained palpable. Non-invasive pulse volume recordings of the extremity demonstrated mild steal physiology. The wrist brachial index was 1.04 on the affected extremity and 1.08 on the contralateral side. Digits 1 through 5 had pressures of 55, 53, 52, 54 and 54 mmHg respectively with the

access occluded. Pressures decreased marginally to 46, 47, 46, 49 and 43, respectively, with the VA open.

Neurologic consultation was obtained as part of her evaluation. Findings were consistent with underlying bilateral peripheral polyneuropathy probably secondary to long standing diabetes. Nerve conduction studies revealed axonal loss and reduced motor and sensory nerve conduction velocities in the ulnar and median nerves. These findings were present bilaterally; however, they were more profound on the affected, right extremity. Electromyogram demonstrated severely diminished muscle contractility in the median and ulnar innervated muscles on the right hand.

Because of her profound symptoms and the suspicion of IMN, fistula ligation was agreed upon and performed. Despite fistula ligation, her symptoms transiently worsened for 4 weeks after ligation requiring several hospitalizations for pain management. Her hand function was limited with impaired grip strength and paresthesias. The pain management service was consulted. Pain directed management of her symptoms included gabapentin and noratriptaline. Her symptoms have gradually abated over a 6-month interval of intensive physical therapy and she currently has baseline strength with minimal numbness. She has refused any new attempts at alternative access locations.

DISCUSSION

The differential diagnosis of hand and digit pain following VA surgery includes: local wound hematoma with nerve compression, carpal tunnel syndrome exacerbated by surgery, soft tissue swelling, IMN and VSS.

Opinions differ as to whether IMN represents a distinct entity from VSS. A subset of patients diagnosed with IMN will demonstrate no evidence of VSS. Conversely all patients with VSS do not suffer from IMN. IMN results from ischemic injury to the sensory and motor nerves distal to a brachial artery based VA. Ischemic neuropathy of upper limb nerves after dialysis surgery was first reported by Bolton et al in 1979 (2). A more detailed description of the condition and coining of the term IMN came in 1983 (3). The descriptions of IMN are largely confined to the surgical and neurologic literature although some reports exist in the renal literature (4, 5).

The incidence of IMN is not precisely described, probably because the entity may easily be misdiagnosed for other vascular and neurologic disorders. IMN has been most commonly observed following VA in diabetic patients who suffer from pre-existing neuropathy in association with peripheral arterial occlusive disease or extensive vascular calcific disease (6). Women appear to be more vulnerable to IMN. Raheb et al described

12 patients diagnosed with IMN after forearm prosthetic bridge grafts based off the brachial artery. All were women (7). Miles described IMN symptoms as being immediate in onset with dominant neurologic signs and symptoms (5).

In general, sensory complaints are more prominent than motor. Symptoms occur along the distribution of the radial, ulnar and median nerve resulting in poor wrist extension and reduced thumb apposition. The forearm muscles may be relatively spared compared to the intrinsic hand muscles resulting in a profound loss of grip, ie a "claw hand" deformity. Subjectively, the hand is warm and the radial pulse is commonly present (8).

Prompt identification of IMN may be challenging as a result of the protean manifestations in the post-operative period when incisional discomfort is not uncommon. Various authors report the incidence of symptomatic VSS to range from 8-40% (9). In most cases, mild symptoms of VSS will stabilize. For moderate to severe symptoms of VSS, aggressive diagnostic and therapeutic measures should be applied.

Paradoxically, the early post-operative window may represent the best and only chance to arrest the progression or reverse the symptoms of IMN. The author advocates for early neurologic evaluation in patients who have symptoms consistent with IMN. Classically, patient complaints that appear to be out of proportion combined with a lack of evidence for VSS, should raise one's suspicion for IMN.

In IMN, non-invasive digital pressures may be normal or mildly reduced. Non-invasive vascular studies must be performed to obtain digital values with the access circuit both opened and temporarily occluded. As a point of reference, non-invasive vascular studies which reveal digital pressures greater than 50 mmHg and a digital to brachial pressure index greater than 0.3 (10) are less consistent with VSS. As a provocative maneuver, the augmentation of digital pressures with occlusion of the upper arm VA is conclusive for the diagnosis of VSS. IMN has only been described with VA constructed at the brachial artery level or proximally. IMN has been described for both prosthetic bridge graft and autogenous fistula constructions. Perhaps the most difficult patient management dilemma is the finding of mild to moderate VSS associated with nerve conduction study abnormalities.

The underlying pathogenesis of IMN appears to be an incremental decrease in perfusion which damages the more sensitive sensory motor nerve tissue. Nerve conduction studies show axonal loss and reduced sensory and motor nerve conduction velocities of median, radial and ulnar nerves. Kaku et al reported a predilection for earlier and more severe (but not isolated) median nerve involvement (11). Paralysis of a single nerve

in the setting of vascular access surgery should prompt a search for local nerve compression secondary to hematoma, aneurysm or abscess (12). Case reports have also documented the initial manifestation or exacerbation of carpal tunnel syndrome following dialysis access creation. The proposed mechanism is venous hypertension in the region of the flexor retinaculum.

The exact mechanism of IMN is poorly understood; however, ischemic injury to susceptible peripheral nerves seems to explain the common history of pre-existing diabetic neuropathy in individuals who develop IMN subsequent to VA surgery. Furthermore, transient ischemia during the intra-operative clamping of the arterial tree may explain why some patients have no objective findings of VSS at the time of the post-operative examination.

No proven reason exists to explain why all patients with VSS do not develop IMN. The explanation is probably related to a complex interplay between the degree and abruptness of the vascular ischemia and the degree of susceptibility caused by the pre-existing peripheral nerve damage long standing diabetes [AQ: correct or missing text]. Kelly et al (13) demonstrated underfilling of the microcirculation of the vasa nervorum with electron microscopy after acute large vessel ligation in their experimental model. It is possible that a nutrient vessel watershed area of the forearm nerve plexus in some patients is more vulnerable to acute ischemia, which may explain the almost universal association of IMN with brachial artery based HD access configurations.

As the discussion demonstrates, early diagnosis and prompt treatment of this process is challenging due to the vague symptoms and frequent delay in the definitive diagnosis. There are no pre-operative findings or diagnostic tests that appear accurately to predict vulnerability to the development of IMN. All patients undergoing HD access creation seem to be at risk for the process. However, there appears to be a relatively higher risk in patients in whom there is a baseline presence of arterial insufficiency or diabetic neuropathy.

A more global awareness among the multidisciplinary team, which often includes nephrologists, vascular surgeons, interventional radiologist, nurses and dialysis care technicians, is necessary to advocate for the early consideration of IMN. Diligent follow-up in the immediate post-operative period following surgical reconstruction can facilitate earlier diagnosis and potential correction. The diagnostic evaluation of the differential diagnosis of hand and digit pain following HD access creation includes tests which may unavoidably contribute to a delay in diagnosis. Especially, since much of the HD surgery is conducted in outpatient settings. In my practice, the average time to return the patient to the operating room for AV [AQ: correct?] access ligation is 17 days. For a patient with the most severe symptoms

of VSS upon awakening, indistinguishable from IMN, careful consideration should be given to an immediate return to the operating theater for ligation of the HD vascular construction.

Perhaps most discouraging with regards to IMN, is the dismal prognosis for complete recovery following closure of the HD access. Because of the propensity for chronic, disabling neurologic symptoms when IMN is not managed aggressively, Miles advocates for immediate closure of the access (5). Despite attempts at earlier closure, several published reports suggest that even under this aggressive management, IMN symptoms may be permanent or only partially reversible (14-16). As a practical matter, adding to the management dilemma is the concern that many patients may have few alternative sites for future VA construction. The decision to close a functioning access requires careful deliberation. The recognition that the same complications may occur in other locations given the fact that the predisposing systemic factors in the patient are unchanged. [AQ: sentence correct or missing text?] Importantly, the subgroup with the best chance for recovery appears to be those patients suffering from IMN with a component of correctable VSS.

IMN represents a significant diagnostic dilemma. Improved awareness of the presentation of this infrequent access related disorder and a high level of clinical suspicion, provides the clinician with the greatest chance of a timely diagnosis with an optimal patient outcome.

Conflict of interest: None
No financial support

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