Relief of sympathetically maintained pain associated with a venous malformation of the hand using intravenous regional guanethidine

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RA Hopper, K Anderson. Relief of sympathetically maintained pain associated with a venous malformation of the hand using intravenous regional guanethidine. Can J Plast Surg 1994;1(4):188-190. A 28-year-old woman with a venous malformation of the hand, unresponsive to previous surgical therapy, experienced significant relief from her incapacitating symptoms after a series of four intravenous regional guanethidine blocks. The effects of the blocks lasted an average of six weeks each. Continuation of the treatments was delayed following an unexpected hypersensitivity reaction to the last block. The case would suggest a sympathetic component to this malformation-associated pain. Further assessment of the effect of this relatively simple, easily repeated treatment on other malformations is indicated.

Key Words: Guanethidine block, Sympathetically maintained pain, Venous malformation

Soulagement d’une douleur sympathomimétique persistante associée à une malformation veineuse de la main à l’aide de guanéthidine intraveineuse locale

RÉSUMÉ : Une femme de 28 ans, atteinte d’une malformation veineuse à la main, qui n’avait pas répondu à la chirurgie, a ressenti un soulagement important de ses symptômes invalidants après une série de quatre blocs intraveineux locaux, à l’aide de guanéthidine. L’effet de ces blocs a persisté en moyenne six semaines pour chacun. La poursuite des traitements a été retardée après une réaction allergique imprévisible au dernier bloc. Ce cas donne à penser qu’il y aurait une composante sympathomimétique à cette douleur associée à une malformation. Il y a lieu d’approfondir l’évaluation de l’effet de ce traitement relativement simple et facile à répéter sur d’autres types de malformation.

Vascular malformations are always present at birth, grow commensurately with the individual, never involute, and are characterized histologically by normal endothelial cell turnover (1,2). A result of defective embryogenesis, they may contain one or a combination of vascular elements: capillary, venous, arterial or lymphatic. Of these, the venous type is the most common in the upper limb (3).

Cutaneous manifestations of venous malformations range from large saccular lesions to a simple blue discoloration of the skin (4). Swelling is frequently evident (4,5). The most common clinical symptom is pain in the affected limb (4); however, a sense of heaviness which is worse with dependency and localized hyperhidrosis are also often present (3).

It is unclear whether the localized pain of malformations is due to intravascular coagulation, localized phlebothrombosis or some other cause (6). Initial treatment should be conservative, including ASA, elevation and elastic support (3,4). Large diffuse masses causing chronic pain cannot be entirely resected, but can be debulked with subtotal resection (6,7); recurrences, however, are frequent (8). Intractable pain, functional loss of the extremity and failed previous operations may result in amputation being the only remaining treatment option (3).

Intravenous (iv) regional noradrenergic neuron blocks with guanethidine have been used successfully in relieving sympathetically maintained pain (9) in a number of conditions involving the extremities (10-12). They are simple to perform and can easily be repeated at regular intervals (12). A literature search covering CIBA-Geigy’s data base on Ismelin, the only guanethidine marketed in Canada, was conducted. Medline references dating back to Hannington-Kiff’s first description of the iv regional block in 1974, as well as prominent textbooks on pain control (12) and on vascular malformations (3), failed to report that the pain from vascular malformations can be relieved by guanethidine blocks. If successful in alleviating cases of intractable pain from venous malformations, this relatively conservative treatment could prevent otherwise inevitable amputation of affected extremities.

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CASE PRESENTATION

A 28-year-old white female was seen in clinic in July 1992 with a bluish discoloured swollen ulnar aspect of the ring finger of her right dominant hand (Figure 1). There was associated constant throbbing pain, hyperesthesia and increased swelling on dependency. All symptoms were localized to the affected region. She had been followed by our service since 1988 and three months prior had undergone amputation of the little finger and denervation of the ulnar half of the ring finger of the same hand for attempted relief of identical symptoms. Intraoperatively, the lesion was seen to extend to the ulnar aspect of the ring finger (Figure 2).

The lesion had been present since birth, increasing in size during puberty and a recent pregnancy. Conservative therapy as well as five staged subtotal resections of the lesion had been ineffective in relieving the associated pain for longer than two months. Preamputation arteriography had revealed quick arterial flow, no arteriovenous (AV) shunting and a minimal vascular blush over the right little finger and ulnar aspect of the right ring finger. Radiographs had demonstrated decreased bone density in the fourth and fifth phalanges, as well as the third, fourth and fifth metacarpals. Histological examination of the amputated digit had shown dilated vascular channels with abnormal anastomoses, yet no endothelial atypia. In the months following amputation of the little finger, cutaneous manifestations of the previous lesion with similar associated symptoms began to increase on the dorsum of the ulnar aspect of the right ring finger.

At the time of the clinic visit, the right hand was nonfunctional due to the intensifying localized throbbing pain and hyperesthesia which had been associated with the spreading malformation for the preceding five years. The pain was decreased on elevation of the limb and increased on dependency. The appearance and clinical examination of the uninvolved area of the hand was normal. Amputation of the ring finger was requested by the patient and considered inevitable. The pain clinic was consulted concerning the appropriateness of a guanethidine block. Although not known to be effective under these circumstances, the block was performed using standard technique (13).

There was immediate and total recovery of the hyperesthesia. The throbbing pain diminished to a level that was described as “comfortable” by the patient. For the first time the patient could recall, she was able to use her right hand without pain for writing and other basic tasks. The swelling and discoloration were not affected by the block. Relief of symptoms lasted for five to six weeks, with two successive blocks being equally effective. During the fourth block, the patient experienced an unexpected apparent anaphylactic reaction with mild hypotension and generalized pruritic urticaria. She responded quickly to standard resuscitative measures, and the beneficial effect of the block lasted for six weeks. Due to this reaction an alternate sympathetic inhibitor, bretylium, was substituted with minimal effect. Options being considered at present are retrying bretylium or continuing with guanethidine using prophylaxis in anticipation of a repeat hypersensitivity reaction. There has been one documented report of apparent central nervous system sensitivity to guanethidine involving a postblock apneic episode (14); however none have mentioned an allergic reaction such as the one experienced by this patient.

DISCUSSION

The presenting signs and symptoms of this patient, both before and after the amputation, were characteristic of a vascular malformation. Presence of the lesion at birth, as well as the lack of endothelial atypia on histology, would rule out a hemangioma. From the histology, the malformation was considered to be primarily of the venous type. The exacerbation during puberty and pregnancy (3), the increased swelling on dependency, and the radiographic evidence of bone hypoplasia and demineralization (15) would support this classification. The extension of the lesion following subtotal resection is not uncommon (8,16).

The patient’s preblock pain was identical in nature to that experienced before her amputation and denervation. This
would decrease the possibility that the cause was a postdene-
ervation neuralgia. There have been cases of post traumatic
sympathetic dystrophy following dissection of major
neurovascular structures in very unstable high-flow vascular
malformations (3); however, this is not a documented com-
pliation with low-flow anomalies. There was no history of
trauma nor any clinical indication that reflex sympathetic
dystrophy could have been the source of the patient’s pain.
The localized symptoms would also tend to exclude a central
source of the pain.

Guanethidine depletes nerve terminal noradrenaline stores and
inhibits the release of any remaining neurotransmitter,
thereby producing a pharmacological sympathectomy (9,11).
Sympathetically maintained pain is characterized by a burn-
ing dysesthetic quality and hyperesthesia, as well as trophic
and vasomotor disturbances. Tissue injury involving periph-
eral nerves with sustained noxious input is a critical
element to its development (17).

Although the pain experienced by this patient did not bear
the classical burning quality of sympathetically maintained
pain, the rapid and consistent response to guanethidine blocks
would imply a sympathetic component. The noxious input
stimulus from the malformation, secondary to nerve compres-
sion or ischemic changes, may constitute the afferent arm
of a spinal pain arc ending in the peripheral release of norad-
renaline lowering the pain threshold to the initial stimulus
(9,18).

We suggest that the relatively simple, easily repeated iv
regional guanethidine block may be considered in the symp-
tomatic treatment of vascular malformation associated pain.

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