Case Report

The vascular armamentarium explored-the ultimate goal achieved

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INTRODUCTION

Peripheral arterial disease (PAD) due to diabetes as a risk factor and atherosclerosis as an aetiology has been evaluated, studied and well documented, PAD due to vasculitis as an aetiology is usually underdiagnosed and overlooked. Limb loss can be the result of either trauma, disease, congenital anomalies, malignancy, PAD with PAD resulting in chronic threatening limb ischemia (CTLI) being the most common cause of limb loss.1 Due to the slow progression of symptoms, the transition to critical ischemia in PAD can appear to “come as a surprise.” Notable risk factors include advanced age, nicotine abuse, diabetes mellitus, hypercholesterolemia, and hypertension.2 Rate of dysvascular amputation is 8 times greater than that of trauma related amputations, risk of amputation is higher when PAD and diabetes coexist compared to PAD and other risk factors.3 74% undergoing dysvascular amputations are diabetic and 55% of these will have an amputation in their contralateral limb 2-3 years after their initial amputation.4 Amputation is the last treatment option for critical limb ischemia after unsuccessful vascular or endovascular treatment.5 The objective was to implement all available modalities to salvage the Limb of a patient who presented to us with CTLI due to medium vessel vasculitis rather than most common presentation of Peripheral arterial disease with atherosclerosis and coexisting diabetes mellitus resulting in CTLI and to emphasize on the need and benefit of being on regular follow up.

CASE REPORT

45 years old female presented with severe rest pain over left leg and forefoot gangrene with past history of right
below knee amputation done one month ago. A brief past history narrating her events will throw light to our timely attempts to save her limb. Diagnosed with nephrotic syndrome and treated with steroids, immunosuppressants and was treated by dermatologist for mottled extremity skin, itching (01/2018). Had blackish discoloration of left great toe for 6 months (dry gangrene), non healing ulcer over dorsum right foot for 3 months with absent peripheral pulses and cold extremities with CTLI of right lower limb, CT angiogram showed total occlusion of bilateral lower limb axial arteries, underwent right below knee amputation and minor amputation of left great toe (05/2018). Presented with severe rest pain over left leg with forefoot gangrene (vital limb), was evaluated further, had grossly elevated CRP, ESR; ANCA-negative, ACR 1990 -4/10 positive was diagnosed with polyarteritis nodosa (medium vessel vasculitis being the aetiology for PAD), was started on pulsed cyclophosphamide, steroids; antibiotics as per culture and sensitivity were given (Table 1).

Catheter directed thrombolyis was done with urokinase for 48 hours adjusted according to fibrinogen levels following which pain reduced, flow improved on Doppler, gangrene started demarcating (07/2018). Presented with left foot gangrene and underwent left forefoot amputation (08/2018). Was receiving pulsed cyclophosphamide, wound was healthy (09/2018); presented with rest pain over left leg, CT angiogram showed flush occlusion of bilateral common femoral arteries with collaterals distally resulting in reformation, underwent left lower external iliac to profunda bypass (10/2018) (Figure 1). Presented with non-healing ulcer, underwent seven sessions hyperbaric oxygen therapy followed by Split skin grafting (11/2018) (Figure 2, 3A and B). On follow up graft had taken up well and limb was saved (12/2018) (Figure 3C). Finally we had an ambulant patient with intact prosthesis who was able to carry out her daily chores without any support thereby achieving our goal of limb salvage (02/2019) (Figure 4).

Table 1: American College of Rheumatology criteria 1990 for polyarteritis nodosa.

<table>
<thead>
<tr>
<th>Classification criteria</th>
<th>Case fulfilment</th>
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</thead>
<tbody>
<tr>
<td>Weight loss &gt;4 kg</td>
<td>+</td>
</tr>
<tr>
<td>Livedo reticularis</td>
<td>+</td>
</tr>
<tr>
<td>Testicular pain</td>
<td>NA</td>
</tr>
<tr>
<td>Myalgia, weakness, leg tenderness</td>
<td>+</td>
</tr>
<tr>
<td>Mono/polyneuropathy</td>
<td>-</td>
</tr>
<tr>
<td>DBP &gt;90 mmHg</td>
<td>-</td>
</tr>
<tr>
<td>Elevated blood urea, creatinine</td>
<td>-</td>
</tr>
<tr>
<td>Hepatitis B</td>
<td>-</td>
</tr>
<tr>
<td>Arteriographic abnormality</td>
<td>+</td>
</tr>
<tr>
<td>Biopsy showing polymorphonuclear cells</td>
<td>-</td>
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</tbody>
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Figure 1: CT Angiogram showing bilateral common femoral artery occlusion.

Figure 2: Pre and post hyperbaric oxygen therapy.

Figure 3: (A) After SSG, (B) 48 hours after SSG and (C) on follow up.
By exploring the armamentarium available and aptly using the right modality at the right time we were able to ultimately achieve our goal of limb salvage.

DISCUSSION

Vessels of any size or location can be affected by vasculitis causing inflammation of vessel wall, resulting in vascular damage and a wide variety of clinical signs and symptoms. Classified based on the predominant type of vessel involved (referred to as large- medium- or small- vessel vasculitis. Polyarteritis nodosa (PAN) is a form of necrotizing vasculitis, predominantly involving medium-sized arteries. Primary (idiopathic) and secondary form is (related to hepatitis b virus [HBV] infection). 1990 ACR criteria for the diagnosis of PAN with 3 or more of 10 criteria distinguishes PAN from other forms of vasculitis with a sensitivity of 82.2% and specificity of 86.6%. Pathological hallmarks are focal segmental necrotizing vasculitis, arterial wall inflammation characterized by fibrinoid necrosis of the media and a cellular infiltrate primarily composed of neutrophils and lymphocytes. Almost any organ/organ system can be affected (kidneys, liver, spleen, lower extremities, skin, nervous system, gastrointestinal system). Constitutional symptoms (fever, weight loss) are also common but not very specific. PAN typically does not affect the lungs a feature that is often helpful in distinguishing from the anca-associated vasculitides, which often involve the pulmonary capillaries. There is no diagnostic laboratory test for PAN, and anca tests are typically negative, ESR and CRP are elevated. Computed Tomography of the abdomen can be useful in detecting organ infarcts and/or bowel-wall thickening. Characteristic findings on conventional angiography such as multiple microaneurysms of the celiac, mesenteric, and renal artery branches are often diagnostic of PAN, stenosis and occlusion of arteries is also common and in one study 98% angiograms demonstrated occlusive lesions, which is even more sensitive than renal microaneurysms, which have been reported in 70% patients. Depending on the pattern of clinical involvement, muscle, nerve, or deep- skin biopsies for histopathologic confirmation of vasculitis may be required in some patients. 50% of PAN cases can be cured with glucocorticoid treatment alone, appropriate for patients with mild disease and avoids the risks associated with more intense immunosuppressive therapy. Poor prognostic indicators (e.g., renal insufficiency or gastrointestinal, cardiac, or neurologic involvement) require glucocorticoids combined with cyclophosphamide to induce disease remission. Surgical modalities like bypass are well documented and extensively evaluated for limb salvage in CTLI patients. Procedures like minor amputations, split skin grafting are adjuncts for wound healing. Physiotherapists play a major role in psychosocial recovery of patients with long duration of hospital stay, long treatment time and are a major support for patients to carry out their activities of daily living (ADL). Hyperbaric oxygen therapy augments wound healing in almost any form of ulcer by providing optimal oxygen which plays a major role in wound healing.

CONCLUSION

With the availability of multiple treatment modalities, identifying correct aetiology is the key for salvage of a critical limb despite factors such as patients’ general condition and other co morbidities. We as vascular specialists should make an attempt to implement all available modalities at the right place and time to ultimately achieve the goal of limb salvage.

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REFERENCES


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