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Successful use of combined corticosteroids and rituximab in a patient with refractory cutaneous polyarteritis nodosa

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Abstract

Cutaneous polyarteritis nodosa is a rare subtype of polyarteritis nodosa that lacks significant internal organ involvement. It has a relapsing remitting nature and usually is less responsive to conventional treatments. We report a case of refractory cutaneous polyarteritis nodosa who failed three immunosuppressive therapies and three different biological agents. He was successfully treated with two rituximab 1000 mg infusions with a good efficacy and tolerance. This case demonstrates the safety and efficacy of rituximab in treatment of refractory cutaneous polyarteritis nodosa.

Keywords: Rituximab; Refractory; Cutaneous; Polyarteritis nodosa

1. Introduction

Cutaneous polyarteritis nodosa (cPAN) is a subtype of polyarteritis nodosa (PAN) characterized by different cutaneous clinical features such as livedoid lesions, necrotic ulcers or painful nodules without any evidence of visceral involvement (Morgan and Schwartz, 2010). It is associated with relapsing remitting lesions that were less responsive to conventional treatments. There are no recommendations or guidelines available to date to manage refractory cPAN. Herein, we report a case of a refractory cPAN who was successfully treated with rituximab.

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2. Case report

A 25-year-old man presented with recurrent intermittent episodes of painful ulcerative lesions (Fig. 1) over both legs of 4 years duration. He denied any systemic complaints. There were no clinical features that suggested inflammatory bowel diseases. Clinical examination revealed multiple irregular ulcers of various sizes ranging 2–15 cm over both legs but more in the right leg. Investigations revealed raised acute-phase reactants (erythrocyte sedimentation rate 83 mm/h and C-reactive protein 40.2 mg/L). Complete blood counts (CBC) and chemistry were normal. Rheumatoid factor, antinuclear antibody and anti-neutrophilic cytoplasmic antibody (ANCA), anti-phospholipid antibodies, cryoglobulins and viral markers (HIV/HBsAg/anti-HCV) were negative. A magnetic resonance arteriography/venography (MRA/MRV) of thoracic, abdominal and extremities were normal. A skin biopsy of the ulcer showed necrotizing transmural vasculitis of small- and medium-sized vessels consistent with cPAN. No evidence of granulomatous inflammation or fungal elements was noticed. He was managed initially with prednisolone

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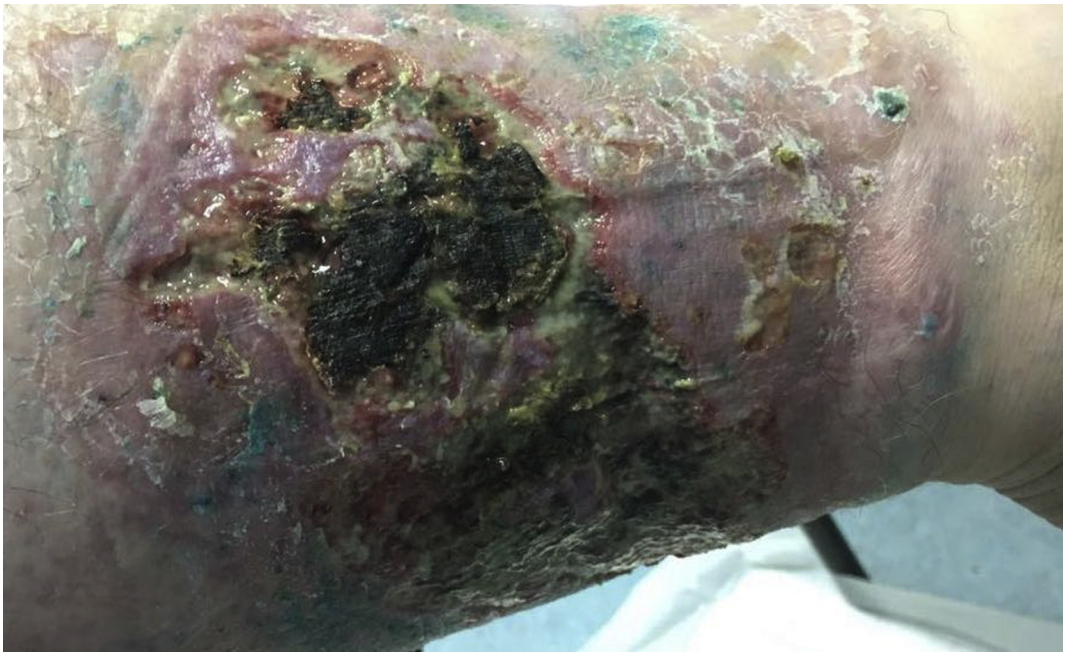

Figure 1. Painful initial ulcer at the lateral aspect of the right leg.

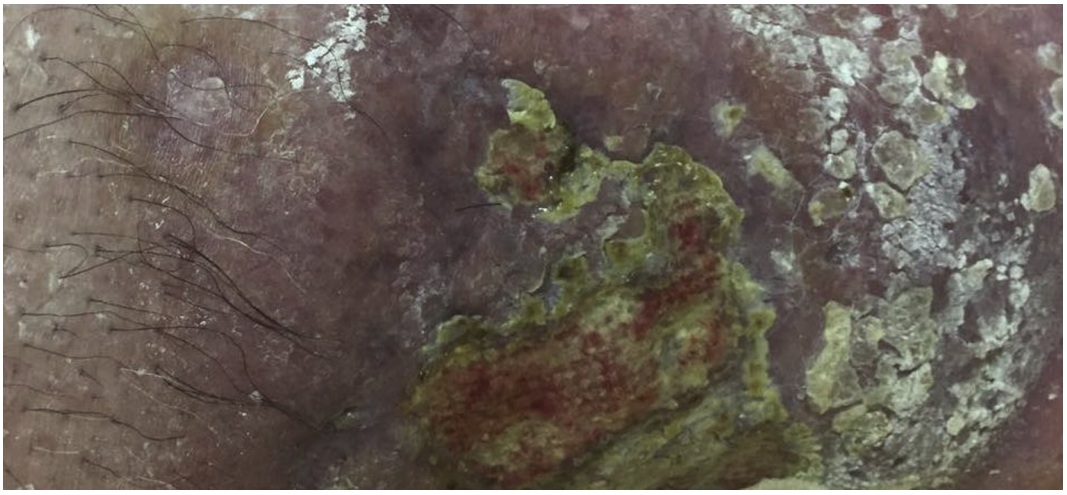

Figure 2. Healed ulcer at the lateral aspect of the right leg.

(1 mg/kg) for 4 weeks with tapering doses. However, steroid tapering beyond 20 mg daily without relapses can't be achieved. Therefore, different conventional therapies tried including azathioprine100 mg/day, mycophenolate mofetil 1500 mg twice a day and methotrexate 15 mg/weekly but without a significant improvement. Subsequently, three different anti-tumor necrosis factor-α (TNF-α) agents, etanercept 50 mg/week for 6 months, adalimumab 40 mg biweekly for 6 months and infliximab 5 mg/kg/dose for total 10 doses, were administered but with little benefit.

In view of refractory recurrent cPAN, three pulses of 250 mg of methylprednisolone followed by oral prednisolone at 1 mg/kg/day and two doses of rituximab (1000 mg) at 2-weekly intervals were initiated. After two weeks, the patient became pain free and able to walk. Indeed, a significant reduction in the size of vasculitic ulcers was noted (Fig. 2), along with resolution of raised acute-phase reactants and the ability to taper prednisolone doses without recurrence.

3. Discussion

To date, very few cases of adult with refractory cPAN treated successfully with rituximab are described (Krishnan et al., 2012; Sonomoto et al., 2008). Our case illustrates the difficulty of management of cPAN. Few data are available regarding the management of refractory cPAN and not based on clinical guidelines. Most of time

conventional immunosuppressive therapies are used before biological agents. Other drug-regimens included anti-TNF agents (Valor et al., 2014). However, our patient had a refractory disease to all commonly used immunosuppressive therapies and thee different anti TNF- α agents. Rituximab is a chimeric monoclonal antibody against the B cell-specific antigen CD20. Rituximab is used in treatment of rheumatoid arthritis, ANCA-associated vasculitis and variable autoimmune diseases. Rituximab has been well tolerated and effective in this case, since the pain disappeared and the size of ulcers reduced significantly and the healing process was obvious within 2 weeks. Thus, rituximab could be used for treatment of refractory cutaneous polyarteritis nodosa.

Disclosure of interest

The authors declare that they have no conflicts of interest

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