

## Case Report

## Diagnosing of Small Vessel Vasculitis Might be a Challenge – A Rare Case Report

\*Shahin MA<sup>1</sup>, Khan MM<sup>2</sup>, Shrestha S<sup>3</sup>, Abdal SJ<sup>4</sup>, Islam MA<sup>5</sup>, Choudhury MR<sup>6</sup>**Abstract**

The term vasculitis refers to the inflammation of vessel walls. It may range in severity from a self-limited disorder in one single organ to a life-threatening disease due to multiple organ failure. Most patients with small vessel vasculitis present with constitutional symptoms like fever, malaise, weakness, fatigue and weight loss. To diagnose small vessel vasculitis, serology like ANCA, serum cryoglobulin and biopsy play an important role. Despite the serology and biopsy, diagnosing small vessel vasculitis occasionally remains challenging in resource constraint countries. Here we are reporting a case of a 26-year-old female who presented with purpura and neuropathy. The patient lacks clinical features like constitutional symptoms, renal involvement, upper airway involvement and her ANCA was negative. Depending on biopsy finding and skin and neurologic involvement, we diagnosed her as a case of small vessel vasculitis (unclassified). The patient improved with IV methylprednisolone followed by oral glucocorticoid treatment along with methotrexate. Although small vessel vasculitis has some typical features, diagnosis may often remain challenging even after biopsy.

**Keywords:** Small vessel vasculitis, unclassified purpura, neuropathy

1. \*Dr. Md. Abu Shahin, Associate Professor, Department of Rheumatology Bangabandhu Sheikh Mujib Medical University (BSMMU), Shabagh, Dhaka. Email: abushahinrh@gmail.com
2. Dr. Mohammad Mamun Khan, Phase B Resident, Department of Rheumatology, BSMMU, Shabagh, Dhaka.
3. Dr. Swikriti Shrestha, Phase B Resident, Department of Rheumatology, BSMMU, Shabagh, Dhaka.
4. Dr. Syed Jamil Abdal, Medical Officer, Department of Rheumatology, BSMMU, Shabagh, Dhaka.
5. Dr. Md. Ariful Islam, Assistant Professor, Department of Rheumatology, BSMMU, Shabagh, Dhaka.
6. Dr. Minhaj Rahim Choudhury, Professor, Department of Rheumatology, BSMMU, Shabagh, Dhaka.

\*For correspondence

**INTRODUCTION**

Small vessel vasculitis typically involve small blood vessel. It is of two types, ANCA associated and not ANCA associated. ANCA associated vasculitis are microscopic polyangitis, granulomatosis with polyangitis and eosinophilic granulomatosis with polyangitis.<sup>2</sup> There is also some small vessel vasculitis with ANCA negative but immune complex mediated. These vasculitides are cryoglobulinaemic vasculitis, IgA vasculitis, hypocomplementaemic vasculitis and urticarial vasculitis etc.<sup>3</sup>

ANCA associated vasculitides typically involve skin, kidney, lung and nervous system and upper airway.<sup>4</sup> The most common presentation is palpable purpura. Upper airway involvement is the most common for GPA whereas renal involvement is mostly for MPA and neuropathy is most common presentation for EGPA and MPA.<sup>5</sup> New onset asthma or worsening of asthma is common for EGPA.<sup>6</sup> Mononeuritis multiplex is the common neurological manifestation. About 80-90 % of patients with GPA or MPA are ANCA positive whereas few patients may be ANCA negative.<sup>7</sup>

Immune complex mediated vasculitides are ANCA negative. These patients may have purpura, joint pain, abdominal pain and most patients are younger aged consistent with IgA vasculitis.<sup>8</sup> Purpura, Reynaud's, joint pain, renal disease, peripheral neuropathy, weakness are typical features of cryoglobulinaemic vasculitis.<sup>9</sup> Serology like ANCA, serum cryoglobulin and biopsy may be conclusive for small vessel vasculitis. But sometimes it is difficult to categorize small vessel vasculitis in a resource constraint country.

Here we present a case of small vessel vasculitis but the patient had ANCA negative, no immune deposit on skin biopsy and serum cryoglobulin cannot be performed due to lack of facilities. So categorizing small vessel vasculitis is difficult for this patient.

**CASE PRESENTATION**

A 26-year-old female came to the hospital with history of recurrent purpuric skin lesions in both lower limbs for the last 5 years. Each episode lasted for 10-15 days then resolve spontaneously. She had no history of fever, joint pain,

abdominal pain, cough, shortness of breath, nasal crusting, tingling or numbness, high colored frothy urine. Her past history and family history was insignificant.



**Figure 1:** Purpura in both lower limbs (before treatment)

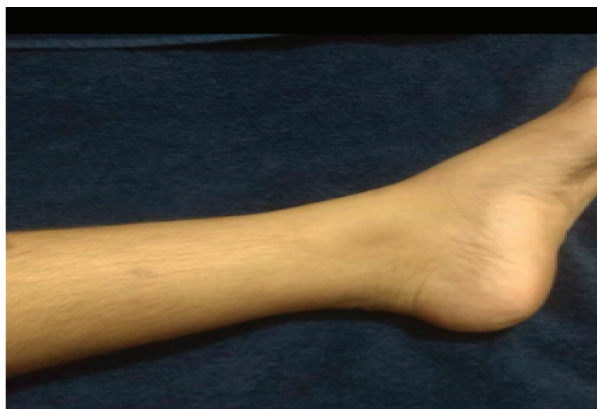
On examination, her vitals were stable. There was maculopapular rash in both lower limbs up to the buttocks which were erythematous, various shapes and sizes, some were coalesced, some were palpable and some were non palpable and non-tender (Fig-1). On neurological examination, there was wasting of thigh muscles, thenar and hypothenar muscle. All jerks were absent in both upper and lower limbs. All other systemic examinations were unremarkable.

Investigation reports revealed- Hb was 12.2 gm/dl, total leucocyte count was  $8 \times 10^9 / \mu\text{L}$ , platelet was  $320 \times 10^3 / \mu\text{L}$  and RBC count was  $4.76 \times 10^6 / \mu\text{L}$ . Her ESR was 33 mm in 1st hour, CRP was 21.7 mg/dl, SGPT was 13 U/L, S. creatinine was 0.59 mg/dl. Lipid profile revealed - total cholesterol- 168 mg/dl, HDL - 44 mg/dl, LDL - 96.2 mg/d and TG was 139 mg/dl. Urine RME showed pus cells 0-1/HPF. RA test, ANA, Anti-ds-DNA, P-ANCA and C-ANCA were negative. C3 was 1.21 g/l (0.9-1.8g/l); C4 was 0.45g/l (0.1-0.4g/l). HBsAg and Anti HCV were negative.

Biopsy from the skin showed mild hyperkeratosis with follicular plugging and thinning of epidermis. The dermis showed mild endothelial swelling with infiltration of chronic inflammatory cells in the vessel wall and increased collagen deposition. Cryostat sections of skin did not show any deposition of IgG, IgM, C3, and fibrinogen. NCS was suggestive of mixed sensory motor demyelinating and axonal polyneuropathy.

The patient was considered to be a case of small vessel vasculitis (unclassified). She had purpura and sensorimotor

axonal and demyelinating neuropathy. There was no renal involvement or constitutional symptoms and this makes the diagnosis challenging.



**Figure 02:** Improvement of purpura (after 3 months of treatment)

The patient was put on IV methylprednisolone 1 gm for 3 days followed by steroid 1mg/kg and methotrexate. The skin manifestation improved significantly three month of treatment (Fig-02).

## DISCUSSION

Vasculitis refers to a heterogeneous group of disorders in which there is inflammation and damage in blood vessel walls, leading to tissue necrosis.<sup>10</sup> It can result in different degrees of stenosis or damage to the vessels and ischemic damage to the innervated tissues or organs.

Small-vessel vasculitis is responsible for a wide variety of diseases that affect vascular structures such as venules, capillaries, arteries and arterioles with classic inflammation. The ANCA associated vasculitides include Wegener's granulomatosis; microscopic polyangiitis and its renal limited form, idiopathic necrotizing crescentic glomerulonephritis; and Churg-Strauss syndrome.<sup>11</sup> There is also some small vessel vasculitis with ANCA negative but immune complex mediated which include cryoglobulinaemic vasculitis, IgA vasculitis and hypocomplementaemic urticarial vasculitis

Immune complex mediated vasculitis is associated with immune complex deposition in the vessel wall and is usually ANCA negative. These patients may have purpura, joint pain, Reynaud's, renal disease and peripheral neuropathy. Serology like ANCA, serum cryoglobulin and biopsy may be conclusive for small vessel vasculitis. But sometimes it is difficult to categorize small vessel vasculitis in a resource constraint country.

Here we have presented a case of small vessel vasculitis but the patient was ANCA negative, no immune deposit on skin biopsy. Serum cryoglobulin cannot be performed due to lack of facilities. So categorizing small vessel vasculitis was a challenge for that particular case.

The patient had purpura and sensorimotor axonal and demyelinating neuropathy. Nerve biopsy and serum cryoglobulin could not be done due to lack of facilities. There was no renal involvement or constitutional symptoms and this makes the diagnosis challenging. The patient was considered to be a case of small vessel vasculitis (unclassified).

### CONCLUSIONS

Patients with vasculitis may present with atypical clinical findings. Several times clinical findings do not correlate with laboratory findings. Sometimes routine laboratory test as well as skin biopsy may be inconclusive. Strong clinical suspicion, rare diagnostic tests and specific drug therapies may be helpful to reach the diagnosis.

### Conflict of interest

The authors have no conflicts of interest to declare.

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