

Case Report**Small vessel vasculitis**¹Narani A, ²Abiramy A¹Teaching Hospital, Jaffna**Abstract**

Vasculitis is inflammation of vessel walls. Different types of vasculitis preferentially affect different sizes of blood vessels. Small vessel vasculitis is a rare immunological disease that affects arterioles, capillaries and venules. The inflammation can restrict blood flow and damage vital organs and tissues.

Keywords

Small vessel vasculitis, ANCA negative

Introduction

The SVV are divided into those conditions associated with circulating ANCA, and those not associated with ANCA. Those conditions that are not associated with ANCA, are associated with immune-complex deposition.

Case report

A 15-year-old girl, presented with 25 days of high-grade fever associated with chills and bilateral knee joint pain which is preceded by sore throat. After 10 days she developed a reddish rash on the bilateral lower limb (feet) later it progressed to the bilateral upper limb. She complained of numbness of bilateral hands and feet, then it progressed to gangrene formation. In due course, she also complained of abdominal pain but no history of malena, hematuria, oliguria, and breathlessness. Also, she had a history of hair fall, dry eyes, and dry mouth but no history of rashes, oral ulcers and Raynauds. She started with intravenous methylprednisolone pulse and continued with high-dose steroids. However, while on treatment she defaulted.

She had no significant past medical and surgical history, but she had an unknown drug allergy. On examination she was febrile, ill-looking, and pale, her pulse rate was 80 beats/min, blood pressure in her right arm was 133/90 mmHg, her left arm was 130/89 mmHg, there

was no lymphadenopathy and no peripheral edema. The right-side radial artery pulse was weak whereas the left was normal. Posterior tibial and dorsalis pedis artery pulses were unable to feel bilaterally. Wet gangrene was noted in bilateral toes and dry gangrene was noted in the 2nd finger of the right hand. The ulcer was noted on the anterior aspect of the medial malleolus of the right lower limb. Other systemic examinations were normal.

Figure 1. Bilateral foot wet gangrene**Figure 2. Wet gangrene in hands****Figure 3. Right medial malleolus ulcer**

Corresponding Author: A Narani, Email: ara.jhc@gmail.com, ORCID: 0000-0002-8294-7788, Submitted August 2024 Accepted December 2024



This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License, which permits unrestricted use, distribution and reproduction in any medium provided the original author and source are credited

Table 1: Laboratory and special investigations

WBC	21.40	ASOT	400	Total protein	69	Serum creatinine	54
NEUTROPHIL	16.87	RF	<20	Albumin	21	ESR	116
LYMPHOCYTE	3.4	ANA	NEGATIVE	Globulin	48	UPCR	1.02
Hemoglobin	8.58	DsDNA	NEGATIVE	Total bilirubin	10.1	Serum ferritin	234
Platelet	570	TSH	0.88	Direct bilirubin	7	HepB surfaceAg	NEGATIVE
AST	68	Ft4	1.74	Na ⁺	130	Hep C Ab	NEGATIVE
ALT	88	PT	13.4	K ⁺	4	PANCA	NEGATIVE
ALP	209	APTT	22	CRP	154	CANCA	NEGATIVE
Man-toux	NEGATIVE	Cryoglobulin	POSITIVE	VDRL	NEGATIVE	HIV	NEGATIVE

Cryoglobulin may be false positive as the test done on while on enoxaparin, UFR protein +, pus cells 8-10, red cells- nil, Bilateral distal arterial duplex- monophasic flow in distal arteries could be due to arterial spasm/vasculitis. Arterial duplex of right upper limb normal, 2D Echo- normal, Blood culture- negative, sputum AFB- not seen, Urine culture- coliform isolated, sensitive to amikacin, co-amoxiclav, cotrimoxazole, nitrofurantoin, piperacillin, Wound swab- 2 types of coliforms, coagulase-negative staphylococcus isolated.

Skin biopsy- appearance is in keeping with early features of SVV, Blood picture- reactive neutrophil leukocytosis and thrombocytosis, anemia with marked rouleaux compatible with anemia of chronic disease, red cell microcytosis,

She had undergone amputation of 4 toes in her right foot. She developed wet gangrene on the left 2nd, 5th and right 2nd fingers. She was managed with intravenous antibiotics. She was started with an intravenous methylprednisolone pulse of 1g daily for 3 days and changed to oral prednisolone 45mg/day also she started with an intravenous cyclophosphamide pulse of 75mg fortnightly for 6 weeks and planned to continue 3 weekly intervals. With these treatments progression of the disease was controlled. She was discharged with a tail-off regimen of steroids.

Discussion

SVV can be categorized as ANCA associated or non-ANCA associated vasculitis. SVV caused by the deposition of immune complexes leads to hemorrhaging from the involved vessels.

A purple or reddish rash on the legs, buttocks, torso or upper body is the telltale symptom of SVV. Other symptoms of SVV include fever, weight loss, myalgia and arthralgia, kidney and gastrointestinal involvement. A physical examination can help confirm if the disease is limited to skin or systemic.

Skin biopsies can provide valuable diagnostic information. Relevant antibody and blood testing can be done during the diagnosis. SVV can co-occur with other autoimmune diseases such as rheumatoid arthritis, Crohn's disease, SLE, etc.

The first step of management is to address the causative agent. Some will also require corticosteroids in combination with other immunosuppressive medications such as azathioprine and methotrexate. Other potential medications may include colchicine, antihistamines, hydroxychloroquine and dapsone alone or in combination. Symptoms of SVV will often resolve within a few weeks to months if there is no organ involvement.

Local ulceration from cutaneous small vessel vasculitis can lead to wound infection or cellulitis, chronic leg ulceration and scar formation.

Referneces

1. <https://www.anaiddedermatologia.org.br/pt-case-for-diagnosis-cutaneous-small-articulo-S0365059621000040>
2. <https://www.rheumatologyadvisor.com/ddi/small-vessel-vasculitis/>
3. <https://pmc.ncbi.nlm.nih.gov/articles/PMC10236953/>
4. https://journals.lww.com/mjdy/fulltext/2014/07020/a_case_of_small_vessel_vasculitis.21.aspx
5. <https://www.merckmanuals.com/professional/musculoskeletal-and-connective-tissue-disorders/vasculitis/overview-of-vasculitis>
6. <https://www.news-medical.net/health/Vasculitis-Affecting-Small-Blood-Vessels-ANCA-Associated-and-Non-ANCA.aspx>
7. <https://dermnetnz.org/topics/cutaneous-small-vessel-vasculitis#:~:text=The%20most%20severe%20complications%20of,Chronic%20leg%20ulceration.>