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Vasculitis associated with VEXAS syndrome

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Abstract

Objectives: To define the prevalence, distribution and characteristics of patients with VEXAS (vacuoles, E1-enzyme, X-linked, autoinflammation, somatic) syndrome who have confirmed vasculitis.

Methods: Patients with VEXAS syndrome, verified by positive UBA1 mutation, were included. Chart review was performed to identify patient characteristics and outcomes. Vasculitis diagnosis was based on either histopathology showing vascular inflammation or non-invasive angiography findings. Summary statistics were calculated.

Results: Eighty-nine patients met inclusion criteria. All were male with a median age of onset of 66.9 years (interquartile range 60.1, 72.7). Median (interquartile range) follow-up was 3.8 (2.2–5.5) years, during which 21 patients (23.6%) had evidence of vasculitis. Vasculitis subtypes included small vessel vasculitis (19.1%), cutaneous medium vessel vasculitis (2.2%) and large vessel vasculitis (2.2%). No patient had more than one vessel size involved. Histopathology in small vessel vasculitis patients was consistent with cutaneous leukocytoclastic vasculitis in the majority, though one patient had leukocytoclastic peritubular capillaritis on renal biopsy. Cranial symptoms (headache, vision changes or jaw pain) were noted in 18.0%. Two additional patients not experiencing cranial symptoms exhibited large vessel involvement with confirmed carotid thickening on non-invasive angiography; one of these had a positive temporal artery biopsy.

Conclusion: VEXAS syndrome manifests as a variable vessel vasculitis in a quarter of patients, with cutaneous small and medium vessel involvement being particularly common. Some patients may have positive ANCA serologies or even renal vasculitis leading to misdiagnosis. Cranial symptoms are common and may mimic GCA, though documented large vessel inflammation is rare.

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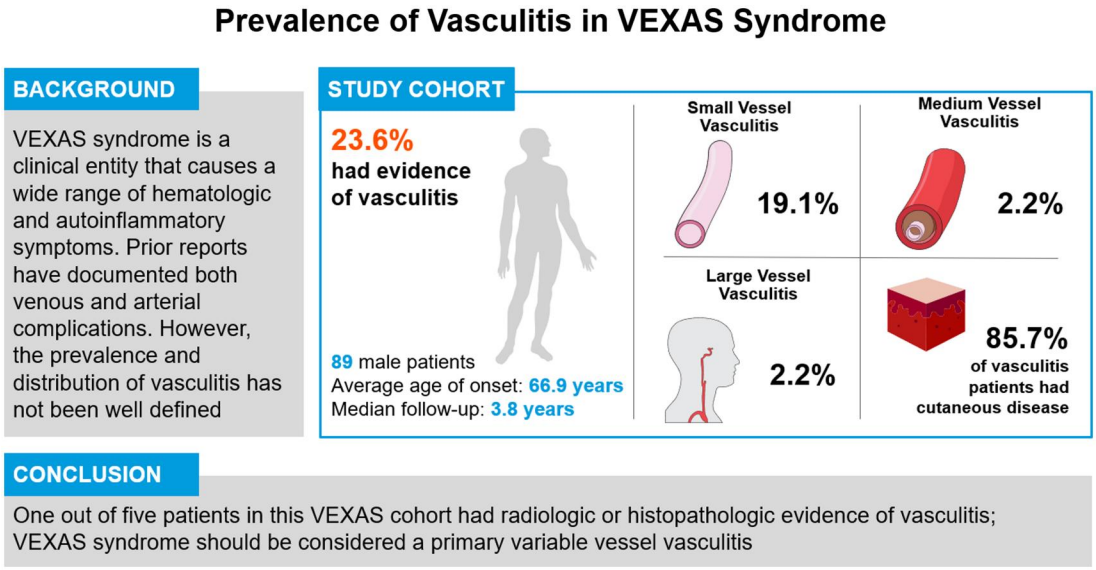
Graphical abstract



Keywords: VEXAS syndrome, UBA1 mutation, vascular inflammation, vasculitis, autoinflammatory disease, giant cell arteritis, somatic mutation.

Rheumatology key messages

- Small, medium and large vessel vasculitis can occur in VEXAS syndrome.
- Vasculitis is appreciated in one out of five.
- Cranial symptoms and large vessel imaging may mimic GCA in VEXAS syndrome.

Introduction

VEXAS (vacuoles, E1 enzyme, X-linked, autoinflammatory, somatic) syndrome is a recently defined clinical entity that causes haematologic and autoinflammatory symptoms. This autoinflammatory illness is separate from traditional autoinflammatory syndromes given that the mutation is somatic and develops in later life. Estimated prevalence is around 1 in 4269 in men and 1 in 26 238 in women over the age of 50 years [1]. Since its initial description in 2020, it was recognized to mimic many other rheumatologic conditions. For example, in the incident VEXAS cohort of 25 patients described by Beck and colleagues, 60% had been diagnosed with relapsing polychondritis, 12% with polyarteritis nodosa and 4% with GCA [2].

Both arterial and venous disease have been well documented in these patients, with a particularly high rate of thromboembolic disease [3]. Our institution and others have noted cutaneous small vessel vasculitis as the most frequent vasculitic manifestation, commonly in the form of leukocytoclastic vasculitis (20–25%) [4]. There have been rare reports of small vessel vasculitis affecting internal organs, including pulmonary capillaritis and necrotizing crescentic glomerulonephritis, the latter associated with MPO-antibody positivity [5, 6].

Medium vessel vasculitis, when reported, has been primarily cutaneous [2, 5, 7–10]. Reports of intra-abdominal medium vessel vasculitis or aneurysms have remained exceptionally rare, with only a solitary case reporting multiple hepatic aneurysms on angiography [8]. While small and medium vessel vasculitis are the most frequently reported manifestations of vasculitis, large vessel involvement has also been observed including positive temporal artery biopsy and abnormal thickening of the subclavian and aorta on vascular imaging [11, 12].

Given the heterogeneity in the presentation of vasculitis features among VEXAS, the purpose of this study was to report the frequency, distribution, clinical and radiographic features of vasculitis associated with VEXAS syndrome.

Methods

Patients with a clinical syndrome consistent with VEXAS and positive UBA1 mutation evaluated at any Mayo Clinic site between 1 January 2020 and 1 July 2024 were included for analysis. Known UBA1 hotspot mutations included p.Met41Thr (c.122T>C), p.Met41Val (c.121A>G), p.Met41Leu (c.121A>C) or intron 2 splice-site mutation (c.118-1G>C, c.118-2A>C). Retrospective chart review was conducted to record demographic information, clinical presentation, laboratory results, relevant imaging findings, histopathology and treatment. Prevalence of vasculitis and important clinical manifestations were summarized. Date of diagnosis was defined as the date a UBA1 mutation positive tissue or blood sample was obtained from the patient. Patients with bone marrow biopsies obtained prior to 2020 were included if later testing was UBA1 positive.

Vasculitis was defined as pathologic or radiographic evidence of vessel inflammation. Patients were compared with the following classification criteria based on vessel size: small vessel vasculitis with ANCA positivity was reviewed based on the 2022 ACR/EULAR classification criteria for granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis and microscopic polyangiitis [13–15]; medium vessel vasculitis was compared with the 1990 ACR classification criteria for polyarteritis nodosa [16]; large vessel vasculitis were reviewed with the 2022 ACR/EULAR classification criteria for GCA [17].

Patients were grouped based on whether they had a diagnosis of vasculitis; differences were tested using Fisher's exact tests for categorical variables or Wilcoxon rank sum tests for continuous variables. SAS version 9.4 was utilized for the analysis.

This study complies with the Declaration of Helsinki and was approved by the Mayo Clinic Institutional Review Board and was deemed exempt from requiring individual patient consent given the retrospective nature of the study.

Results

Eighty-nine patients met inclusion criteria; all were male with an average age of onset at 66.9 years (interquartile range 60.1, 72.7). The most common UBA1 mutations were p.Met41Thr (50.6%), p.Met41Val (24.7%) and p.Met41Leu (16.9%). Intron 2 splice mutations were present in a minority (5.6%). Macrocytic anaemia was present in 85.4% and thrombocytopenia in 37.1%. The majority (88.8%) were on prednisone ranging from 5 to 60 mg daily at the time of last follow-up. Twenty-three patients (25.9%) required prednisone doses >20 mg daily for inflammatory control. Median (interquartile range) follow up was 3.8 (2.2–5.5) years during which 21 patients (23.6%) had pathology or imaging positive for vascular inflammation. Four of these patients met classification criteria for a specific vasculitis (one each for GCA, polyarteritis nodosa, granulomatosis with polyangiitis and microscopic polyangiitis).

Small vessel vasculitis

Seventeen (19.1) patients had histopathologic evidence of small vessel vasculitis. The majority (16 patients) had leukocytoclastic vasculitis on cutaneous biopsy (Fig. 1). Of these patients with leukocytoclastic vasculitis on cutaneous biopsy, two were found to have positive ANCA testing. One had associated arthralgias, lung opacities, and pANCA positivity without MPO-antibody fulfilling 2022 ACR/EULAR classification criteria for microscopic polyangiitis. The second had leukocytoclastic vasculitis along with reports of a previous ANCA positive lab test, though the original records of specific ANCA staining could not be found.

A third patient, who did not have cutaneous vasculitis documented, had cANCA and PR3-antibody positivity with leukocytoclastic peritubular capillaritis not involving the glomeruli on kidney biopsy, which has been previously reported [18]. This patient met 2022 ACR/EULAR classification criteria for granulomatosis with polyangiitis.

Medium vessel vasculitis

Two patients (2.2%) had cutaneous pathology confirming medium vessel vasculitis (Fig. 1). One additional patient carried a diagnosis of polyarteritis nodosa based on renal infarcts, sensorineural hearing loss, sensory neuropathy and

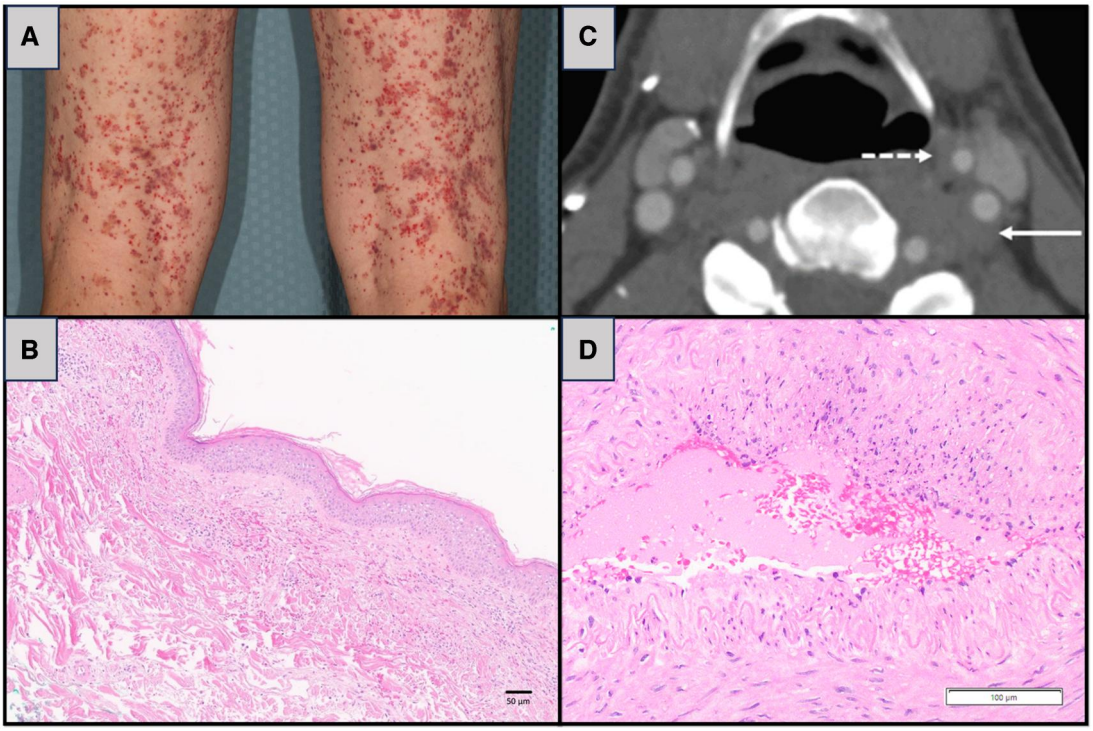


Figure 1. Small and large vessel vasculitis in VEXAS syndrome. (**A**) Gross image of petechial rash in a patient with VEXAS syndrome. (**B**) Pathology from above patient with sparse neutrophilic inflammation surrounding superficial dermal blood vessel walls consistent with small vessel vasculitis, scale bar is 50 µm. (**C**) Left internal (solid arrow) and external (dashed arrow) carotid circumferential periarterial thickening on CT angiography in a patient with VEXAS syndrome. (**D**) Endothelialitis with mixed inflammation is present in the intima of a muscular temporal artery, scale bar 100 µm, haematoxylin and eosin stain. VEXAS: vacuoles, E1-enzyme, X-linked, autoinflammation, somatic.

vision loss secondary to ischaemic optic neuropathy. This patient did meet 1990 ACR classification criteria with positive biopsy, testicular swelling, elevated creatinine and neuropathy. CT angiography of the abdomen and pelvis, however, did not confirm vessel wall thickening or aneurysm formation.

Large vessel vasculitis

Cranial symptoms were reported in 14 patients (12 headache, 1 jaw pain, 2 vision changes and 1 abnormal temporal artery exam with bulging of the vessel and overlying tenderness). Twenty-two patients (24.7%) underwent evaluation for GCA due to concern of either systemic and/or cranial symptoms. Two temporal artery US were performed: both were negative for wall thickening or oedema.

Eighteen patients (20.2%) underwent temporal artery biopsy of which only one was positive with focal neutrophilic infiltrate in the intima (Fig. 1). The patient with the positive temporal artery biopsy also had CT angiographic evidence of carotid peri-arterial thickening (Fig. 1). One additional patient was found to have carotid inflammation on MR angiogram. Both patients with carotid findings had UBA1 p.Met41Val mutations and notably did not experience cranial symptoms. One of these patients met 2022 ACR/EULAR criteria for GCA based on positive temporal artery biopsy and elevated inflammatory markers.

The patient previously described with bulging of the temporal artery on physical exam with associated tenderness did develop vision loss, though temporal artery biopsy was negative.

Comparative analysis

Patients with any vessel vasculitis were compared with those without (Table 1). Ocular symptoms were the only variable statistically associated with vasculitis (P =0.013). Ocular

Table 1. Characteristics of VEXAS patients with and without vasculitis

	Presence of small, medium or large vessel vasculitis			
	No (N = 68)	Yes (N = 21)	Total (N = 89)	P-value
Age at haematologic or suspicious inflammatory symptom onset				0.108[a]
Mean (s.d.)	67.1 (9.11)	63.5 (8.20)	66.3 (8.99)	
Median (IQR)	67.5 (60.1, 73.4)	63.6 (59.0, 68.4)	66.9 (59.8, 71.5)	
Race, *n* (%)				
White	65 (95.6)	21 (100.0)	86 (96.6)	>0.999[b]
Asian	1 (1.5)	0 (0.0)	1 (1.1)	>0.999[b]
Other	2 (2.9)	1 (4.8)	3 (3.4)	0.559[b]
Mutation type, *n* (%)				0.544[b]
p.Met41Thr	35 (51.5)	10 (47.6)	45 (50.6)	
p.Met41Val	15 (22.1)	7 (33.3)	22 (24.7)	
p.Met41Leu	13 (19.1)	2 (9.5)	15 (16.9)	
Intron 2 splice region	3 (4.4)	2 (9.5)	5 (5.6)	
Unknown	2 (2.9)	0 (0.0)	2 (2.2)	
Constitutional symptoms, *n* (%)				
Fever	37 (54.4)	12 (57.1)	49 (55.1)	>0.999[b]
Weight loss	34 (50.0)	7 (33.3)	41 (46.1)	0.216[b]
Night sweats	15 (22.1)	3 (14.3)	18 (20.2)	0.546[b]
Lymphadenopathy	5 (7.4)	4 (19.0)	9 (10.1)	0.206[b]
Haaematologic manifestations, *n* (%)				
Macrocytic anaemia	59 (86.8)	17 (81.0)	76 (85.4)	0.496[b]
Thrombocytopenia	25 (36.8)	8 (38.1)	33 (37.1)	>0.999[b]
MDS	6 (8.8)	4 (19.0)	10 (11.2)	0.238[b]
Venous thromboembolic disease, *n* (%)				
DVT	24 (35.3)	8 (38.1)	32 (36.0)	0.801[b]
PE	6 (8.8)	2 (9.5)	8 (9.0)	>0.999[b]
SVT	11 (16.2)	2 (9.5)	13 (14.6)	0.725[b]
Ocular symptoms, *n* (%)	30 (44.1)	16 (76.2)	46 (51.7)	**0.013**[b]
Nasal or auricular chondritis, *n* (%)	25 (36.8)	9 (42.9)	34 (38.2)	0.618[b]
CRP				0.079[a]
Mean (s.d.)	60.9 (64.47)	76.4 (53.20)	64.6 (61.99)	
Median (IQR)	33.0 (14.0, 94.0)	69.0 (26.5, 113.5)	42.0 (17.0, 95.0)	
Prednisone dose at last follow-up, *n* (%)				0.940[b]
None	8 (11.8)	2 (9.5)	10 (11.2)	
0–5 mg daily	4 (5.9)	0 (0.0)	4 (4.5)	
6–10 mg daily	16 (23.5)	6 (28.6)	22 (24.7)	
11–15 mg daily	9 (13.2)	2 (9.5)	11 (12.4)	
16–20 mg daily	15 (22.1)	4 (19.0)	19 (21.3)	
21–30 mg daily	8 (11.8)	4 (19.0)	12 (13.5)	
31–60 mg daily	8 (11.8)	3 (14.3)	11 (12.4)	
Patient expired, *n* (%)	13 (19.1)	5 (23.8)	18 (20.2)	0.757[b]

[a] Wilcoxon rank sum P-value.
[b] Fisher's exact P-value.
DVT: deep venous thrombosis; IQR: interquartile range; SVT: superficial vein thrombosis; VEXAS: vacuoles, E1-enzyme, X-linked, autoinflammation, somatic.

symptoms included periorbital oedema [16], uveitis [9], scleritis [7], conjunctivitis [5] and vision loss [1].

Discussion

Vasculitis has been increasingly reported in patients with VEXAS syndrome. We sought to clarify the frequency, distribution and characteristics of vessel inflammation within our cohort. Our data show that a quarter of patients with VEXAS could develop vasculitis based on pathology or imaging and this can be variable in vessel size. Small vessel inflammation, particularly cutaneous, is the most common manifestation. However, medium vessel and large vessel vasculitis was also observed among our cohort. When medium vessel vasculitis occurs, it is typically cutaneous and without involvement of the mesentery, a notable contrast to polyarteritis nodosa [19].

The recognition of both cranial symptoms and diagnosed large vessel inflammation in this cohort suggest VEXAS may be a common mimicker of GCA. Although 22 patients underwent evaluation for GCA, it is noteworthy that only one patient fulfilled 2022 ACR criteria. Whether the cranial symptoms are caused by direct vascular inflammation in VEXAS syndrome, as is known to be the case with GCA, is yet to be determined. Two patients had peri-arterial carotid thickening, one of which also had neutrophilic inflammation on temporal artery biopsy. The overall frequency of head and neck artery involvement in VEXAS remains uncertain and should be confirmed in larger cohorts and further evaluated in prospective studies.

It was considered that the thrombosis risk in VEXAS may be inherently linked to vascular inflammation, and that the risk of vasculitis would be highest amongst those with history of arterial or venous thrombosis. This would be a model like Behçet's syndrome, where vasculitis itself leads to thrombosis and immunosuppression alone can mitigate risk [20]. However, as noted in Table 1, our retrospective analysis did not find a correlation between those with documented thrombosis and vasculitis indicating they may have different pathophysiologic mechanisms within VEXAS syndrome.

The question has also been raised of whether patients presenting with vasculitis should be screened for VEXAS syndrome when accompanying features are present. An Italian centre reported its experience of 147 male patients seen in their vasculitis clinic who were screened for the presence of symptoms suggestive of VEXAS syndrome symptoms, including fever and one of the following: dermatologic manifestations, pulmonary infiltrates, auricular or nasal chondritis, or venous thromboembolism [6]. The patients additionally had to have elevated CRP with at least some haematologic abnormality. Seven patients met their inclusion criteria, of which five underwent genetic screening for UBA1 mutations and three were positive for *UBA1* mutation consistent with VEXAS. The manifestations in these three patients included ANCA-associated vasculitis complicated by necrotizing and crescentic glomerulonephritis, cutaneous vasculitis, and an 'undifferentiated inflammatory syndrome with vasculitis features' (notably, no histologic or radiographic evidence of vasculitis in this patient) [6]. Two other groups have evaluated for UBA1 mutations and clonal hematopoiesis within vasculitis cohorts, noting similarly low prevalence [21, 22].

Limitations in the present study included the low statistical power to assess between group differences, its retrospective nature and the lack of standardized investigations in patients with VEXAS syndrome. It is possible that the prevalence of large vessel vasculitis and medium vessel vasculitis may be higher in the population but are not routinely screened for. Given the low positive rate of temporal artery biopsy in our cohort, and the focal nature of the neutrophilic infiltrate on the one positive biopsy, it may be reasonable to consider temporal artery US prior to or in lieu of biopsy. In addition, non-invasive angiography or PET-CT may be preferred measures of screening this population for large vessel involvement when clinically suspected. In the case of medium vessel vasculitis, a deep enough cutaneous biopsy would need to be performed. Conversely, the rate of small vessel vasculitis may have been overestimated since centralized review of the pathology in our centre was not required. An article by Zakine *et al.* suggested that leukocytoclasia may be more common on skin biopsy than true vasculitis [4] though other reviews support similar rates of cutaneous vasculitis within VEXAS syndrome to our own centre's report [23].

Our cohort findings suggest that VEXAS syndrome should be considered a primary systemic vasculitis, particularly a variable vessel vasculitis. Small vessel vasculitis has been commonly reported, medium vessel vasculitis was confirmed in the cutaneous tissue of this population, and less commonly visceral medium vessel vasculitis has been reported previously [8]. Large vessel vasculitis was clearly demonstrated on non-invasive angiography with confirmation of an inflammatory infiltrate on the temporal artery biopsy. These findings highlight that vessels of small, medium and large categories are involved as defined in the 2012 International Chapel Hill Consensus Conference on the Nomenclature of Vasculitides [24]. The prevalence in our series is notably higher than in other forms of variable vessel vasculitis such as Behçet's disease (12% with arterial inflammation reported [25]) and Cogan's syndrome (estimated 10% will develop aortitis [26]).

Assessing individuals with vasculitis for VEXAS syndrome, especially those exhibiting classical features, presents a complex process. Our observations reveal that patients diagnosed with VEXAS, when concurrently presenting with documented vasculitis, exhibit atypical clinical manifestations and glucocorticoid dependency. Atypical vessel involvement, as in the case of our patient with peritubular capillaritis on renal biopsy, or the patient with medium vessel involvement of the bronchial arteries in the initial *NEJM* series, can be seen [2]. Given the findings amongst this patient cohort and the prior literature, those aged 50 years and above, particularly males, with atypical presentations of vasculitis and/or glucocorticoid dependency should raise concern for VEXAS syndrome with consideration of UBA1 testing. The initial step toward identifying effective treatment modalities lies in meticulously identifying and cataloguing this specific population.

Data availability

De-identified data can be requested from the study authors.

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