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THERAPEUTIC MANAGEMENT OF VEXAS SYNDROME: A PRACTICAL REVIEW OF CLINICAL MANIFESTATIONS, COMPLICATIONS AND TREATMENT STRATEGIES

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ABSTRACT

Aim: To summarize current clinical knowledge on the manifestations, organ-specific complications, and medical management of VEXAS (Vacuoles, E1 enzyme, X-linked, Autoinflammatory, Somatic syndrome).

Methods: A review of peer-reviewed clinical cohorts, registry evaluations and consensus reports published between 2020 and 2026 was conducted using PubMed, MEDLINE, and Google Scholar. Studies addressing clinical phenotypes, organ manifestations, and treatment responses were analyzed.

Results: VEXAS syndrome is an adult-onset autoinflammatory disorder caused by somatic mutations in the *UBA1* gene within hematopoietic stem cells. It predominantly affects older males and is characterized by systemic inflammation alongside progressive hematologic abnormalities. High-dose oral glucocorticoids reliably achieve rapid flare control - however, steroid dependency is nearly universal and long-term toxicity is substantial. Conventional synthetic disease-modifying antirheumatic drugs (csDMARDs) show minimal efficacy. Janus kinase (JAK) inhibitors have emerged as the most consistent targeted medical therapy for controlling inflammatory symptoms and reducing steroid exposure. Interleukin-1 (IL-1) and interleukin-6 (IL-6) inhibitors provide partial benefit but carry risks of secondary loss of response, severe injection-site reactions, or serious infections. For patients with high-risk myelodysplastic syndrome (MDS) features or severe cytopenias, the hypomethylating agent azacitidine reduces clonal burden and improves blood counts. Allogeneic hematopoietic stem cell transplantation (allo-HSCT) is the only curative treatment for eligible candidates. Long-term therapeutic anticoagulation is indicated for unprovoked thrombosis, and routine antimicrobial prophylaxis is essential.

Conclusion: Managing VEXAS syndrome requires an integrated strategy: rapid suppression of acute inflammation with corticosteroids, maintenance with targeted therapy (ruxolitinib), clonal suppression (azacitidine or allo-HSCT) for marrow failure and aggressive infection prophylaxis.

KEYWORDS

VEXAS Syndrome, *UBA1* Mutation, Autoinflammation, Ruxolitinib, Azacitidine, Hematopoietic Stem Cell Transplantation, Relapsing Polychondritis

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1. Introduction

VEXAS syndrome is a severe adult-onset autoinflammatory disorder first identified in late 2020 by Beck and colleagues at the National Institutes of Health (NIH) [1]. The condition is defined by its acronym: Vacuoles in myeloid and erythroid precursor cells, E1 ubiquitin-activating enzyme deficiency, X-linked inheritance, Autoinflammatory systemic disease, and Somatic mutations acquired later in life [1, 2].

Because *UBA1* is located on the X chromosome, VEXAS primarily affects males, with clinical onset usually occurring in the sixth to eighth decades of life (median age 64–71 years) [1, 3]. Rare female cases have been documented, typically involving monosomy X (Turner syndrome) or skewed X-chromosome inactivation [4, 5]. Recent genomic screening studies suggest the disease is substantially more common than initially suspected, affecting approximately 1 in 4,000 men over 50 years of age [6].

Prior to molecular identification of *UBA1*, patients often endured years of diagnostic delay, receiving various diagnostic labels such as relapsing polychondritis, Sweet syndrome, polyarteritis nodosa, adult-onset Still's disease or unclassifiable myelodysplastic syndrome (MDS) [1, 5, 7]. The underlying pathogenic mechanism involves somatic mutations at codon 41 (Met41) or adjacent splice sites of *UBA1* in hematopoietic progenitor cells [1, 8]. Loss of canonical cytoplasmic UBA1b function forces translation from an alternative downstream start codon (Met67), generating a catalytically impaired isoform (UBA1c) [1]. This defect disrupts cellular ubiquitylation, triggers endoplasmic reticulum stress and unfolded protein response pathways, and drives hypersecretion of inflammatory cytokines, including IL-1 β , IL-6, TNF- α , and interferons [1, 8, 9].

Managing VEXAS is particularly challenging because traditional antirheumatic drugs are largely ineffective, high-dose corticosteroids cause severe long-term morbidity, and bone marrow failure progressively worsens [10, 11]. This review outlines the organ-specific manifestations of VEXAS syndrome and summarizes current evidence regarding medical and procedural treatments.

2. Methodology

A literature review was conducted to synthesize current evidence on the clinical manifestations and therapeutic options for VEXAS syndrome, focusing on studies published between 2020 and 2026. Searches were performed across PubMed, MEDLINE, and Google Scholar using the terms: *VEXAS syndrome*, *UBA1 mutation*, *autoinflammation*, *ruxolitinib*, *Janus kinase inhibitors*, *azacitidine*, *relapsing polychondritis* and *hematopoietic stem cell transplantation*. Cohort studies, prospective series, registry analyses, and therapeutic consensus guidelines were prioritized for inclusion.

3. Cutaneous manifestations

Skin lesions occur in 80% to 90% of VEXAS patients and frequently serve as the initial presenting sign [1, 12]. The dermatologic picture is broad:

Neutrophilic dermatoses (Sweet Syndrome-like): Tender, erythematous-to-violaceous plaques and papules affecting the trunk, extremities, and neck [1, 12].

Leukocytoclastic vasculitis: Palpable purpura, petechiae, and ecchymotic lesions on the lower extremities [1, 12].

Subcutaneous nodules: Painful nodules that clinically mimic cutaneous polyarteritis nodosa [12, 13].

Periorbital edema: Prominent violaceous periorbital swelling and erythema mimicking cellulitis or dermatomyositis [12, 14].

Management

Skin eruptions respond promptly to systemic oral prednisone (0,5-1 mg/kg/day), with resolution of pain and flattening of lesions within days [1, 15]. However, flares recur during steroid tapering in almost all patients [3]. For localized recurrences, potent topical corticosteroids (clobetasol propionate) or intralesional triamcinolone injections (5-10 mg/mL) can help control skin flares without increasing oral steroid doses [12].

Among targeted therapies, the Janus kinase (JAK) inhibitor ruxolitinib (10-20 mg twice daily) achieves complete or near-complete skin clearance in more than 75% of patients [16, 17]. The IL-1 receptor antagonist anakinra (100 mg/day subcutaneously) is also effective for neutrophilic lesions. However, up to 40% of patients experience severe, indurated injection-site reactions that limit its utility [18, 19].

4. Auricular, nasal, and laryngeal chondritis

Cartilage inflammation occurs in 35% to 50% of VEXAS patients, leading to common misdiagnoses of relapsing polychondritis (RP) [1, 7].

Auricular chondritis: Painful swelling and erythema of the external ear that characteristically spares the soft lobule [7]. Recurrent bouts cause cartilage resorption, resulting in permanent "cauliflower ear" deformities [7].

Nasal chondritis: Epistaxis, pain, and structural cartilage loss leading to saddle-nose deformities [7].

Laryngotracheal chondritis: The most hazardous complication, presenting with hoarseness, stridor, and dynamic tracheal collapse [7, 20].

Management

Acute cartilage inflammation responds to oral prednisone at 0,5-0,75 mg/kg/day [7, 15]. Conventional synthetic DMARDs (methotrexate, azathioprine) do not reliably prevent cartilage relapses [7, 21]. For maintenance, ruxolitinib or anakinra successfully suppresses recurring chondritis [7, 17, 18]. Patients with respiratory symptoms warrant evaluation with dynamic CT imaging and spirometry flow-volume loops [7, 20]. Critical subglottic stenosis or tracheomalacia may necessitate balloon dilatation, tracheal stenting or emergent tracheostomy [20].

5. Pulmonary infiltrates and interstitial lung disease

Pulmonary involvement is documented in 40% to 60% of patients and is an independent predictor of mortality [1, 3, 10].

Clinical presentation: Dry cough, progressive dyspnea, and hypoxemia [10].

Radiographic findings: Patchy ground-glass opacities, consolidating infiltrates (organizing pneumonia), interlobular septal thickening and sterile exudative pleural effusions [10].

Diagnostic workup: Bronchoalveolar lavage (BAL) is necessary during initial evaluation or acute deterioration to exclude opportunistic infections (*Pneumocystis jirovecii*, fungal or viral pathogens) [10, 22]. In sterile VEXAS pneumonitis, BAL reveals marked neutrophilic alveolitis [10].

Management

Active pulmonary infiltrates require induction therapy with high-dose oral prednisone (1 mg/kg/day) or intravenous pulse methylprednisolone (500-1000 mg/day for 3 days) [10, 15]. For long-term control, ruxolitinib stabilizes pulmonary infiltrates and improves lung function [16, 17]. In patients with severe coexisting bone marrow dysplasia or disease refractory to JAK inhibition, the hypomethylating agent azacitidine has demonstrated clearance of pulmonary infiltrates through reduction of the *UBA1*-mutant clone [10, 23]. Supplemental oxygen and structured pulmonary rehabilitation provide supportive benefit [10].

6. Thromboembolic and vascular complications

Thrombosis occurs in 35% to 50% of VEXAS patients, making thromboembolism a defining clinical challenge [1, 21].

Venous Thromboembolism (VTE): Deep vein thrombosis (DVT) and pulmonary embolism (PE) predominate, alongside atypical presentations such as portal, mesenteric, or cerebral venous sinus thrombosis [21].

Arterial disease & vasculitis: Ischemic strokes, myocardial infarction, acute peripheral ischemia and medium to large vessel vasculitis mimicking polyarteritis nodosa or giant cell arteritis [13, 21].

Management

Because the recurrence rate after stopping anticoagulation exceeds 60%, an unprovoked VTE in active VEXAS warrants indefinite therapeutic anticoagulation [15, 21]. Direct oral anticoagulants (DOACs; apixaban, rivaroxaban) or low-molecular-weight heparin (LMWH) are first-line treatment [21].

When progressive disease-related thrombocytopenia occurs, dosing adjustments are necessary: if platelet counts drop between 30000- 50000/ μ L, anticoagulant doses are reduced by 50%. Counts below 30000/ μ L generally require withholding anticoagulation due to severe bleeding risks [15, 21]. Concurrently, anti-inflammatory therapy (e.g. ruxolitinib) is necessary to suppress underlying endothelial inflammation and reduce the driving prothrombotic state [17, 21].

7. Musculoskeletal and ocular manifestations

Musculoskeletal and ocular inflammation significantly impair functional capacity and quality of life [1, 14, 24].

7.1 Musculoskeletal features

Joint involvement occurs in 60% to 70% of patients, presenting as symmetric or asymmetric, non-erosive polyarthritis or oligoarthritis primarily affecting the knees, wrists, and ankles [1, 24]. Proximal muscle weakness, severe myalgias and mild elevations in creatine kinase (CK) also occur [24].

Treatment: Joint symptoms respond rapidly to oral prednisone (0,3-0,5 mg/kg/day) [24]. For sustained control, JAK inhibitors (ruxolitinib) or IL-6 blockers (tocilizumab) provide reliable steroid-sparing benefit [17, 25]. NSAIDs should be used cautiously or avoided given baseline risks of chronic kidney disease and steroid-related gastrointestinal toxicity [15].

7.2 Ocular inflammation

Ocular complications occur in 40% to 50% of patients and carry a significant risk of permanent visual impairment [1, 14]. Manifestations include severe episcleritis, necrotizing anterior or posterior scleritis, anterior uveitis, and retro-orbital pseudotumors causing proptosis, diplopia, and compressive optic neuropathy [14].

Treatment: Vision-threatening ocular disease requires immediate pulse intravenous methylprednisolone (1 g/day for 3 days) followed by high-dose oral prednisone (1 mg/kg/day) alongside ophthalmology consultation [14, 15]. Ruxolitinib maintains remission and prevents ocular recurrences [14, 17].

8. Bone marrow pathology, cytopenias and clonal evolution

The bone marrow is the primary biological driver of VEXAS syndrome [1, 26].

Vacuolation: The diagnostic histologic hallmark is cytoplasmic vacuolation in myeloid and erythroid precursors (promyelocytes and proerythroblasts) on marrow aspirates [1, 26].

Cytopenias: Macrocytic anemia develops in almost all patients, alongside progressive thrombocytopenia and lymphopenia [1, 3, 26].

Clonal progression: Co-mutations in genes associated with clonal hematopoiesis (such as *DNMT3A*, *TET2*, and *ASXL1*) are common [1, 26]. Progression to high-risk MDS or transformation into acute myeloid leukemia (AML) occurs in 5% to 12% of long-term cohorts [3, 26].

Supportive Hematologic Care

Transfusions of packed red blood cells and platelets are provided as needed [10, 15]. Erythropoiesis-stimulating agents (ESAs) have very low response rates due to cell-intrinsic defects in *UBA1*-mutant erythroid precursors [10, 26]. Granulocyte colony-stimulating factor (G-CSF) is generally avoided because it can trigger severe autoinflammatory flares and worsen neutrophilic skin lesions [10, 15].

9. Comparative Summary of Main Therapeutic Options

Class / Modality	Specific Agents & Regimens	Clinical Utility & Response	Limitations & Adverse Effects
Systemic Glucocorticoids	Prednisone (0.5-1 mg/kg/day); IV methylprednisolone pulses	Rapid flare control (>90% initial response across symptoms).	Universal relapse when tapered <15-20 mg/day; causes severe bone loss, diabetes, and infections.
Janus Kinase (JAK) Inhibitors	Ruxolitinib (10-20 mg BID); Tofacitinib, Upadacitinib	First-line targeted steroid-sparing agent (>70% response rate); clears skin, chondritis, and lung disease.	Exacerbation of cytopenias (JAK2 inhibition); high incidence of herpes zoster (prophylaxis mandatory).
IL-1 and IL-6 Biologics	Anakinra (100 mg s.c. daily); Tocilizumab (8 mg/kg IV every 4 weeks)	Second-line options for systemic fever, arthritis, and chondritis.	Anakinra causes severe injection-site reactions; tocilizumab carries secondary loss of response and GI perforation risk.
Conventional csDMARDs	Methotrexate, Azathioprine, Leflunomide	Historically prescribed for misdiagnosed conditions (RP, vasculitis).	Poor efficacy (<15% response rate); frequently worsens cytopenias and liver tests. Not recommended.

Class / Modality	Specific Agents & Regimens	Clinical Utility & Response	Limitations & Adverse Effects
Hypomethylating Agents (HMAs)	Azacitidine (75mg/m ² /day for 7 days every 28 days)	Effective in patients with severe cytopenias, transfusion dependence, or MDS overlap; reduces clone size.	Transient myelosuppressive nadir during early cycles; requires 4–6 cycles to assess response.
Allogeneic Stem Cell Transplant (Allo-HSCT)	Reduced-intensity conditioning (RIC) regimens	Only curative therapy; achieves complete molecular remission and donor chimerism in >85% of survivors.	Non-relapse mortality of 15–25%; risks of graft-versus-host disease (GvHD) and serious infections.

10. Infection Prophylaxis and Supportive Care

Infectious complications represent the leading cause of death in VEXAS syndrome, responsible for 40% to 50% of fatalities [3, 11, 22]. Contributing factors include baseline disease cytopenias, innate immune dysregulation, older patient age and prolonged exposure to immunosuppression [22].

10.1 Antimicrobial Prophylaxis

1. ***Pneumocystis jirovecii* Pneumonia (PJP):** Trimethoprim-sulfamethoxazole (TMP-SMX, 800/160 mg three times weekly or 400/80 mg daily) is indicated for all patients receiving prednisone >20 mg/day for >4 weeks, those taking JAK inhibitors, or those on combination therapies [15, 22]. Atovaquone (1500 mg/day) or inhaled pentamidine are used in patients with sulfa intolerance or severe cytopenias [15].

2. **Varicella Zoster Virus (VZV):** Antiviral prophylaxis with valacyclovir (500 mg/day) or acyclovir (400 mg BID) is mandatory for patients receiving JAK inhibitors [15, 16]. Recombinant zoster vaccination (Shingrix) should be administered [15].

10.2 Bone Protection

Baseline bone mineral density testing (DXA) is recommended at diagnosis [15]. Daily supplementation with calcium (1000-1200 mg/day) and vitamin D3 (1000-2000 IU/day) alongside antiresorptive agents (oral bisphosphonates or denosumab) should be initiated to mitigate rapid glucocorticoid-induced osteoporosis [15].

11. Conclusions

VEXAS syndrome is a severe autoinflammatory disorder driven by acquired clonal hematopoiesis. Effective clinical care requires shifting away from conventional DMARDs toward an integrated, multi-stage strategy:

1. **Rapid Induction:** High-dose systemic glucocorticoids rapidly control acute, organ-threatening inflammatory flares.

2. **Maintenance & Steroid Sparing:** Targeted JAK inhibition with ruxolitinib serves as the primary medical maintenance strategy, controlling multi-organ autoinflammation and permitting significant glucocorticoid reduction.

3. **Clonal Suppression:** In patients with progressive bone marrow failure, severe cytopenias or MDS overlap, hypomethylating therapy with azacitidine suppresses the mutated clone and stabilizes blood counts.

4. **Curative Approach:** Allogeneic hematopoietic stem cell transplantation remains the only curative modality and should be evaluated early in eligible patients with severe or refractory disease.

5. **Complication Prevention:** Long-term anticoagulation for unprovoked thrombosis and antimicrobial prophylaxis against *Pneumocystis* and herpes zoster are essential to reduce mortality.

Conflicts of Interest: No conflicts of interest to declare.

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