

Objectives: To highlight the rheumatologic complications of beta-thalassemia, which include musculoskeletal manifestations such as arthritis and arthropathies, joint effusions, osteoporosis, bone fractures, and arthralgias as well as connective tissue diseases such as pseudoxanthoma elasticum (PXE).

Methods: We searched the literature through Google Scholar and PubMed for all publications reporting on rheumatologic manifestations in beta-thalassemia. The following keywords were used: thalassemia, rheumatoid arthritis, osteoarthritis, iron-overload, septic arthritis, deferoxamine-related arthropathy, crystal arthritis, effusions, osteoporosis, fractures, bone marrow hyperplasia, scoliosis, aseptic necrosis, arthralgia, myalgia, and pseudoxanthoma elasticum. All reports published between 1970 and 2016 were included.

Results: Arthralgia and low back pain are among the most common musculoskeletal manifestations of thalassemia. Many studies are reporting an increased rate of RA in hemoglobinopathies such as thalassemia, the association of which may be attributed to genetic factors. Arthritis may develop secondary to iron deposition in the synovial tissue or due to iron chelators such as deferoxamine, which may provoke a self-limited arthritis due to synovial destruction sustained by free radicals production during iron interchange. Several studies reported increased incidence (16%–30%) of arthropathy in beta-thalassemia patients on deferoxamine therapy. Multiple etiologies contribute to osteoarthritis in thalassemia patients such as bone marrow expansion, iron deposition within the joint, and hypoparathyroidism. Only few cases of Salmonella enteritidis septic arthritis were described in thalassemia patients. The associations between osteoporosis and hypogonadism, diabetes, and vitamin D and calcium deficiency were found significant. Skin lesions of PXE were reported in up to 16% in beta-thalassemias.

Conclusions: The pathophysiology of thalassemia and subsequent blood transfusions affect almost every organ system. Rheumatologic manifestations ranging from musculoskeletal complications to connective tissue diseases are common among thalassemia patients. Rheumatologists are highly encouraged to be involved in the multidisciplinary approach for the management of beta-thalassemia.

Disclosure of Interest: None declared

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AB0990 PERI-ARTERITIS AS A NOVEL PARANEOPLASTIC PRESENTATION OF MYELODYSPLASTIC SYNDROME: A CASE SERIES AND REVIEW OF THE LITERATURE

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Background: Myelodysplastic syndromes (MDS) are characterized by ineffective and dysplastic hematopoiesis resulting in peripheral blood cytopenias with varying risk for progression to acute myelogenous leukemia (AML).¹

Autoimmune manifestations have been well described in association with MDS.² Peri-arteritis has rarely been reported.

Objectives: Here, we present a novel association of MDS with peri-arteritis.

Methods: Three patients within our institution and one in the literature were identified. Patients were diagnosed with MDS according to 2008 World Health Organization classification with concomitant occurrence of peri-arteritis on imaging.³

Results: See Table 1.

Conclusions: Autoimmune manifestations associated with MDS occur in 7–25% of patients. The most commonly reported conditions include vasculitis, inflammatory arthritis and connective tissue disease. There is a paucity of literature describing peri-arteritis associated with MDS.² Review of the literature revealed only one other case of peri-arteritis associated with MDS outside of those identified at our facility.⁴

With this case series, we report peri-arteritis as a novel presentation of MDS. Peri-vascular inflammation may mimic idiopathic retroperitoneal fibrosis, however, persistent cytopenias, lack of response to immunosuppressive therapy and persistence of elevated inflammatory markers should prompt the physician to evaluate for an underlying diagnosis of MDS.

Abstract AB0990 – Table 1

Patient	1	2	3	4 ⁴
Age	67	37	67	70
Sex	Female	Female	Female	Male
MDS type	CMML-1	MDS with multi-lineage dysplasia; progression to AML	MDS Unclassifiable	MDS with multi-lineage dysplasia
Presenting Symptoms	Hydronephrosis, abdominal pain, weight loss	Serositis, dyspnea, fevers, rash	Rapidly evolving peri-arteritis, abdominal pain	Organizing pneumonia, fever, arthralgias
Bone Marrow Biopsy	Hypercellular, features of CMML-1	Normocellular, multilineage dysplasia, 4%blasts, 20q deletion	Hypercellular, granulocytic hyperplasia, 2% blasts	Normocellular, multilineage dysplasia
ANCA and ANA	Negative	Negative	Negative	Negative
ESR (0–15mm/hr)	21	76	57	
CRP (<0.9mg/dL)	5.4	31.2	4.9	9
Radiographic extent of peri-arteritis	Infra-renal abdominal aorta and common iliac arteries	Thoracic aorta and branch vessels	Carotid arteries, thoracic and abdominal aorta, femoral vessels	Internal carotid artery
Treatment	Corticosteroids	Corticosteroids, anakinra, methotrexate, IVIG	Corticosteroids	Corticosteroids
Clinical Course	Ureteral stenting and nephrostomy tube placement improved symptoms. Inflammatory markers remained elevated despite treatment.	Multiple treatment modalities failed to improve symptoms or inflammatory markers. MDS transformed to AML requiring stem cell transplant. Her disease was then quiescent.	Despite treatment, disease was complicated by aortic dissection which was managed non-operatively. Azacitidine was initiated but she died of infectious complications.	Epipharyngeal mass improved with corticosteroids. Disease course was complicated by infection.

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AB0991 PREVALENCE OF FIBROMYALGIA AND DEPRESSION IN PATIENTS WITH AUTOIMMUNE /INFLAMMATORY SYNDROME INDUCED BY ADJUVANTS COMPARED TO PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Autoimmune/inflammatory syndrome induced by adjuvants (ASIA) has been associated with previous exposure to various agents such as mineral oil, silicone and vaccines, which act as adjuvants eliciting chronic stimulation of the immune system and manifestation of autoimmune diseases. There is little information about fibromyalgia (FM), depression and the deterioration of the quality of life in ASIA patients.

Objectives: To determine the prevalence of FM, depression and both in ASIA patients associated to mineral oil versus systemic sclerosis (SSc) patients.

Methods: A comparative cross-sectional study was performed in patients with ASIA according to Shoenfeld's criteria associated to injection of mineral oil with cosmetic purposes. The prevalence of fibromyalgia according to criteria of ACR, depression by Beck questionnaire and quality of life with SF-36 in patients with ASIA versus patients with systemic sclerosis matched by age were evaluated.

Results: There were 100 women, 50 ASIA patients (mean age 49±9.7 years) compared to 50 SSc patients (mean age 49±10 years). Of patients with ASIA, 40 patients had non-specific rheumatic disease and 10 met ACR criteria for SSc in 4 patients, SLE 3 and overlap syndrome 1 (SLE plus SSc) and rheumatoid arthritis 2. In ASIA vs. SSc we found depression in 72% vs 46% respectively, p<0.005, fibromyalgia in 66 vs 48%, p<0.008 and fibromyalgia plus depression in 56% vs. 28, p<0.005. Patients with ASIA had a worse quality of life than SSc by SF36 and the most affected parameter was the emotional one.

Conclusions: ASIA patients associated to injection of mineral oil had higher prevalence of depression, fibromyalgia and greater deterioration of quality of life compared to SSc patients. There is a possible link between ASIA, depression and FM.

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