International Journal of Advances in Medicine
Buch KP et al. Int J Adv Med. 2025 Mar;12(2):239-241
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: https://dx.doi.org/10.18203/2349-3933.ijam20250382

Case Report

A rare case of adult-onset Still's disease with peripheral neuropathy

Kathan Pankaj Buch[1]*, Anshu Kishor Solanki[1], Shrey Rajesh Gondaliya[2]

[1]Department Of Internal Medicine, Yash Multispeciality Hospital, Bhavanagar, Gujarat, India
[2]Department Of Internal Medicine, Jagruti Multispeciality Hospital, Jamnagar, Gujarat, India

Received: 20 January 2025
Accepted: 12 February 2025

***Correspondence:**
Dr. Kathan Pankaj Buch,
E-mail: kathanusmle@gmail.com

ABSTRACT

Adult onset stills disease is a rare multisystem inflammatory autoimmune disorder which is characterized by a triad of fever, joint pain, and a transient salmon-pink maculopapular rash. Here we present a rare case of 27-year-old female who presented with symptoms of fever, polyarthritis for 2 months, and ulceration of right foot and numbness of left hand for 20 days. The diagnosis of adult-onset Still's disease (AOSD) was confirmed as patient fulfilled the Yamaguchi criteria. The presentation, diagnostic work up and management is discussed and the current literature pertaining to this interesting case is reviewed.

Keywords: Neuropathy, Still's disease, Fever

INTRODUCTION

Adult-onset Still's disease (AOSD) is a rare, multisystem, autoinflammatory disorder which affects adolescents and young adults.[1] Prevalence of AOSD is estimated to be less than one case per 100,000 people. AOSD is rare, and hence there are currently no consensus on its incidence and prevalence in different populations.2 Until very recently the pathophysiology of this disease was obscure however various factors are now being implicated. Infectious triggers include EBV; parvovirus B19; CMV; HHV; HIV; Coxsackievirus etcetera.[3]

This case is of importance because it shows a neurological manifestation of AOSD which is a rare occurrence. Various neurological disorders associated with AOSD include myositis, fatigue, cerebral infarction, cranial nerve palsy and aseptic meningitis.[4] Recently role of Human Leukocyte antigen has been established in the cause of AOSD, HLA-DR4 and HLA DRw6 have been implicated.[5]

CASE REPORT

Patient presented with complaints of fever for 2 months, pain in both knees, wrists, and elbows for 2 months, non-traumatic right foot ulcer with blackish discoloration of tip of small toe for 20 days, weakness, and numbness of left-hand for 20 days.

Patient was apparently well 2 months back when she started experiencing joint pain, which was gradual in onset, symmetrically involving both knees, elbows, and wrists, associated with morning stiffness and decreasing in severity after 1-2 hours of doing her daily chores in the morning. Locally available oral medication intake was associated with partial relief of the pain of which she used to take 2-3 tablets in a day. This was followed by spontaneous development of a painful ulcer on the outer aspect of right ankle which was small to begin with and progressed to a size of approximately 5×3 cm at presentation and not associated with any discharge from the ulcer site. This was also associated with blackish discoloration of the right 5th digit which was spontaneous in onset and confined to the tip. She also has previous history of similar ulceration at this site 1-1.5 years back which healed with scarring at the site. Patient also noticed weakness in left hand which was gradual in onset with patient having noticed slipping of utensils from the left hand initially which progressed to complete inability to make a fist. This was also associated with numbness over

the left palm and volar aspect of fingers of left hand. She also complained of fever for 2 months, prior to presentation which was insidious in onset, associated with evening rise, decreased in severity on medication, associated with mild chills and not associated with the rigors.

No history of significant trauma, lower back pain, swelling over limbs or fingers, bluish discoloration of fingers, hair loss, oral or genital ulceration, dryness of mouth or the eyes.

No history of difficulty in walking, climbing stairs, slipping of shoes, difficulty in doing chores with right hand, pain in the neck, numbness in other limbs, diurnal variation of weakness.

No history of headache, blurring of vision, cough, chest pain, abdominal pain, altered bowel habits, burning micturition, passage of bloody urine.

Examination

Vitals on admission-BP-right arm-150/90 mm Hg, pulse-104 beats per minute, temperature-100 degrees F, SpO_2-99% room air, built: Average built, height: 150 cm, weight: 44 kg, BMI: 19.55 kg/m^2, head to toe examination: pallor, bilateral submandibular, upper jugular, inguinal lymphadenopathy, ulcer on the lateral aspect of the right foot of size 5×3 cm, gangrene at the tip of the little toe of the right foot. Fundus exam-disc edema-hypertensive papillopathy

Systemic examination

CNS: intact higher mental function, GCS=E4, V5, M6=15/15, motor left hand-grade 3/5 power in flexors, adductors, abductors, decreased sensation of touch, pain, temperature in the left palm.

Cardiovascular system: S1S2, no murmurs

Respiratory system: B/L Normal air entry, no added sounds

Per abdomen: Soft, non-tender, no organomegaly

Local musculoskeletal examination: Tenderness over bilateral knees, elbows, and wrists with the minimal swelling

Laboratory, radiological and histopathological investigation shave been summarized in the Table 1 and 2 below.

In Table 1 elevated serum urea and creatinine are suggestive of renal dysfunction whereas altered iron profile with elevated ferritin levels suggesting of an inflammatory pathology.

Table 1: Laboratory investigations.

Parameters	Investigations
Hematology	
Hemoglobin (mg/dl)	6.7
TLC (cells per cumm)	20,320 with 86% neutrophils
Platelets (lakhs/cumm)	6.42 lacs
Reticulocyte count	1%
Peripheral blood smear	Microcytic hypochromic anemia with neutrophilic leukocytosis
Biochemistry	
Blood urea (mg/dl)	58
S. creatinine (mg/dl)	2.86
Anti CCP	Negative
ANA	Negative
ANCA profile	Negative
ESR (mm/ hr.)	63
CRP (mg/l)	107
Procalcitonin (ng/ml)	0.1
Serum ferritin (ng/ml)	1000
Serum iron (mcg/dl)	38.56
TIBC (mcg/dl)	451
UIBC (mcg/dl)	412
% transferrin saturation	8.55%
LDH (IU/l)	110

Table 2: Imaging and other investigations.

Variables	Investigations
Imaging	
Chest X-ray	Within normal limits
ECG	Left axis deviation
USG whole abdomen and renal artery doppler	Raised resistive index in left kidney (0.7)
Arterial doppler right lower limb	Normal flow
Nerve conduction velocity	Left median and ulnar nerves showed sensorimotor axonal neuropathy.
2D-echocardiography	Concentric left ventricular hypertrophy
Histopathology	
Wedge biopsy from foot ulcer	Inflammatory infiltrates comprising mainly neutrophils, sub epithelium consists of fibro collagenous tissue with foci of acute and chronic inflammatory infiltrates
Biopsy of oropharyngeal lesion	Acute on chronic inflammatory pathology

In Table 2 NCV studies showing sensorimotor neuropathy and histopathological examination of the ulcers show neutrophilic infiltrates.

DISCUSSION

To diagnose a patient with AOSD, Yamaguchi's criteria must be satisfied. We also need to rule out active infection, malignancy, and rheumatological disorders. Five criteria need to be fulfilled with at least two major criteria to diagnose a patient with AOSD.

Major criteria include, typical rash, arthralgia for more than 2 weeks, fever of more than 39 degree Celsius for more than 1 week and White blood cells more than 10,000 with more than 80% polymorphic granulocytes and minor criteria include, sore throat, altered liver function, splenomegaly or hepatomegaly, lymphadenopathy, negative rheumatoid factor, and anti-nuclear antibody.[6] Our patient fulfilled three major criteria and four minor criteria. After ruling out other causes, the patient was diagnosed with mild to moderate severity of this disease and given a course of antibiotics to treat any underlying infection after which the patient was shifted to oral prednisone which was initiated at a dose of 0.5 mg/kg per day with non-steroidal anti-inflammatory drugs which resulted in resolution of symptoms and the patient was slowly tapered from steroids and showed resolution of signs and symptoms. On the two-month follow-up the patient showed resolution in anaemia, neuropathy, and renal dysfunction as well.

Management of moderate to severe disease warrants use of Disease activity modifying drugs like methotrexate and biologics like interleukin one inhibitor like anakinra or interleukin six inhibitor tocilizumab. Newer drugs like canakinumab, rilonacept and tumour necrosis factor inhibitors like infliximab have been used as well.[7]

The prevalence of neurological involvement in AOSD in patients was 7.5% and patients who had neurological symptoms were at a higher risk of developing macrophage activating syndromes.

CONCLUSION

Maintaining high level of suspicion is essential for timely diagnosis and management of AOSD. Sensorimotor axonal neuropathy involving median and ulnar nerves is rare occurrence in patients with AOSD; however, a physician should always be aware of this co-occurrence and evaluate further for prompt treatment of patient.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Giacomelli R, Ruscitti P, Shoenfeld Y. A comprehensive review on adult onset Still's disease. J Autoimmun. 2018;93:24-36.
2. Efthimiou P, Yadlapati S. Adult-Onset Still's Disease. Auto-Inflammatory Syndromes. 2018;33(3)261-76.
3. Van de Putte LB, Wouters JM. Adult-onset Still's disease. Baillieres Clin Rheumatol. 1991;5(2):263-75.
4. Zhao M, Wu D, Shen M. Adult-onset Still's disease with neurological involvement: a single-centre report. Rheumatology (Oxford). 2021;60(9):4152-57.
5. Wouters JM, Reekers P, van de Putte LB. Adult-onset Still's disease. Disease course and HLA associations. Arthritis Rheum. 1986;29(3):415-8.
6. Efthimiou P, Paik PK, Bielory L. Diagnosis and management of adult onset Still's disease. Ann Rheum Dis. 2006;65(5):564-72.
7. Macovei LA, Burlui A, Bratoiu I, Rezus C, Cardoneanu A, Richter P, et al. Adult-Onset Still's Disease-A Complex Disease, a Challenging Treatment. Int J Mol Sci. 2022;23(21):12810.

Cite this article as: Buch KP, Solanki AK, Gondaliya SR. A rare case of adult-onset Still's disease with peripheral neuropathy. Int J Adv Med 2025;12:239-41.