American Journal of Case Reports

ISSN 1941-5923
© Am J Case Rep, 2017; 18: 580-588
DOI: 10.12659/AJCR.903178

Received: 2017.01.04
Accepted: 2017.02.22
Published: 2017.05.26

Quotidian High Spiking Fevers in Adult Still's Disease

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Conflict of interest: None declared

Patient: Female, 29
Final Diagnosis: Adult Still's Disease
Symptoms: Fever • arthralgia • sore throat • shortness of breath
Medication: —
Clinical Procedure: —
Specialty: Rheumatology

Objective: Rare disease

Background: Adult Still's disease (ASD) is a rare systemic inflammatory condition, which commonly presents with the triad of quotidian fevers, rash, and non-specific rheumatologic symptoms such as myalgia and arthralgia. The etiology and pathogenesis are poorly understood and both the clinical presentation and laboratory data are typically nonspecific. As such, the presentation is often confused with infection, other autoimmune processes, and malignancy.

Case Report: We present a case of a 29-year-old Hispanic female who presented with fever, sore throat, myalgia, and shortness of breath. Initially diagnosed with suspected pneumonia, extensive workup led to the final diagnosis of ASD due to the persistence of her symptoms, which met Yamaguchi Criteria, as well as exclusion of other possible etiologies.

Conclusions: ASD is a rare systemic inflammatory condition and its nonspecific presentation often leads to diagnostic delay and disease complications. We discuss the incidence, etiology, pathology, diagnosis, and standards in management of ASD. This case emphasizes the need for high clinical suspicion of ASD, and early exclusion of other etiologies, especially with failure of first-line treatment, to limit patient suffering and complications.

MeSH Keywords: Fever of Unknown Origin • Rheumatology • Still's Disease, Adult-Onset

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/903178

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Background

Adult Still's disease (ASD) is a rare systemic inflammatory condition, which most commonly presents with daily fevers, rash, and non-specific rheumatologic symptoms such as myalgia and arthralgia [1]. The etiology and pathogenesis are poorly understood and both the clinical presentation and laboratory data are typically nonspecific. The disease is typically confused with infection, other autoimmune processes, and malignancy. The rarity of the disease and nonspecific presentation often leads to diagnostic delays and disease complications [2]. We present the case of a 29-year-old Hispanic female who was eventually diagnosed with ASD after a 24-day hospitalization with fever of unknown origin (FUO). We discuss her workup and clinical course and further demonstrate the challenges of ASD diagnosis. This case illustrates the need for early consideration of the diagnosis, especially for patients from underdeveloped and developing countries, and the need for research to help better identify at-risk groups and to expedient diagnosis.

Case Report

A 29-year-old Hispanic female from Guatemala presented with a three-day history of fever, sore throat, body aches with associated shortness of breath. One day prior to presentation, she visited her primary care provider who started her on ciprofloxacin with no improvement. She denied having prior similar episodes, any significant past medical issues, or recent history of travel, sick contacts, or alcohol, tobacco, or recreational drug use. Past surgical history was significant only for a C-section. She denied any allergies and family history was positive only for end-stage renal disease (ESRD) in her father. Her last menstrual period was two weeks prior to presentation.

On examination, she was febrile (39.3°C), tachycardic (128 beats per minute), with a blood pressure of 93/68 mm Hg, respiratory rate of 20 breaths per minute and 95% oxygen saturation on room air. The patient appeared awake, alert, and in mild distress. Her skin was warm and dry without any rash, bruising, or cyanosis. She had full strength with no focal neurologic signs. The pharynx was mildly erythematous without exudates or hemorrhage. Her neck was supple without any lymphadenopathy and a flat jugular venous pressure (JVP). Her lungs were clear to auscultation bilaterally with diminished breath sounds on the left more than the right. Cardiac examination revealed sinus tachycardia and a systolic ejection murmur graded as 3/6. Abdomen was soft, non-distended, tender with epigastric pain but without evidence of guarding, rebound tenderness, masses, or costovertebral angle tenderness (CVA) tenderness. Bowel sounds were normoactive in all four quadrants. Pertinent laboratory tests on initial workup revealed a leukocytosis, microcytic anemia, transaminitis, and critically elevated cardiac troponins. The full initial workup and results are available in Table 1.

Diagnostic studies included an abdominal ultrasound (US), which showed a mild hepatomegaly, chest x-ray (CXR) demonstrating a left lower lobe infiltrate suggestive of pneumonia (Figure 1), and an electrocardiogram (ECG) revealing sinus tachycardia with nonspecific ST segment changes. The patient was diagnosed with sepsis secondary to community-acquired pneumonia, transaminitis due to sepsis, and elevated troponins likely secondary to myocarditis. She was admitted to the telemetry unit and started on ceftriaxone and azithromycin as well as supplemental oxygen. For the myocarditis, the patient was started on colchicine and PRN acetaminophen.

CT scan of her chest, abdomen, and pelvis revealed bilateral pleural effusions with compressive atelectasis in the lower lobes (Figure 2).

After no improvement in her symptoms, the antibiotic regimen was upgraded to piperacillin/tazobactam and vancomycin. However, the patient remained febrile (see temperature trends in Figure 3) and despite multidisciplinary efforts, the patient developed acute hypoxic respiratory failure, requiring BiPAP support. She was transferred to the medical intensive care unit (MICU).

After six days of intensive care management, the patient's condition started improving and she was switched to oxygen by nasal cannula. She was transferred to the medical floor where the fever spikes were accompanied by a re-elevation in WBC count, prompting a *Clostridium difficile* toxin stool screen which was negative and change of antibiotic therapy from piperacillin/tazobactam to meropenem. She developed acute kidney injury (AKI) with a creatinine rise to 1.64 which was determined to be toxic acute tubular necrosis due to nonsteroidal anti-inflammatory drug (NSAID) treatment for her other conditions. She was treated with IV fluid hydration and discontinuation of ibuprofen.

By day 13, the patient was still febrile and urine culture was positive for *Candida* infection, so she was started on fluconazole. In addition, she became severely anemic, requiring two blood transfusions with packed red blood cells. Hematology was consulted and performed a bone marrow biopsy and aspiration. Biopsy revealed 90% cellularity demonstrating a complete maturation of hematopoietic elements, normal spiculation, abundant phagocytic stromal breakdown, and myeloid to erythroid ratio at the high-end of normal at 4: 1. Additionally, she had an increased number of megakaryocytes which were not in clusters or atypical in nature. Lymphoma was ruled-out as there was no increase in CD34 + blast cells, no staining for CD15+ or CD30+ and no Reed-Sternberg cells.

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Table 1. Summary of initial diagnostic tests and laboratory result.

Complete Blood Count	Result	Reference Values
White blood cell count	**19.0×10³/CMM**	4.0–11.5×10³/CMM
Hemoglobin	**11.5 g/dL**	12.0–16.0 g/dL
Hematocrit	**34.3%**	37.0–47.0%
Platelets	**338×10³/CMM**	140–440×10³/CMM
White blood cell count manual differential		
Segmented Neutrophils	88%	42–75%
Bands	1%	0–8%
Blood urea nitrogen	8 mg/dL	6–22 mg/dL
Creatinine	0.63 mg/dL	0.5–1.20 mg/dL
Liver function tests		
Total bilirubin	0.6 mg/dL	0.3–1.2 mg/dL
Alkaline phosphatase	**165 IU/L**	50–136 IU/L
Aspartate transaminase	**179 IU/L**	8–42 IU/L
Alanine transaminase	**155 IU/L**	30–65 IU/L
Cardiac enzymes		
Lactate	1.5 mmol/L	0.4–2.0 mmol/L
Troponin I	**2.5 ng/mL**	<0.3 ng/mL
β-human chorionic gonadotropin	<5.0 mIU/mL	5.0–10.0 mIU/mL

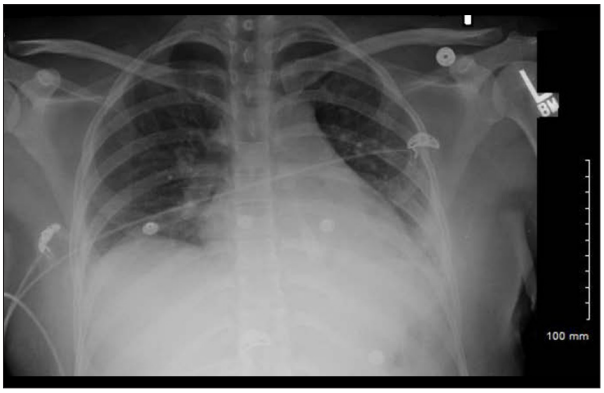

Figure 1. Frontal chest radiograph demonstrating a left lower lobe infiltrate suggestive of pneumonia.

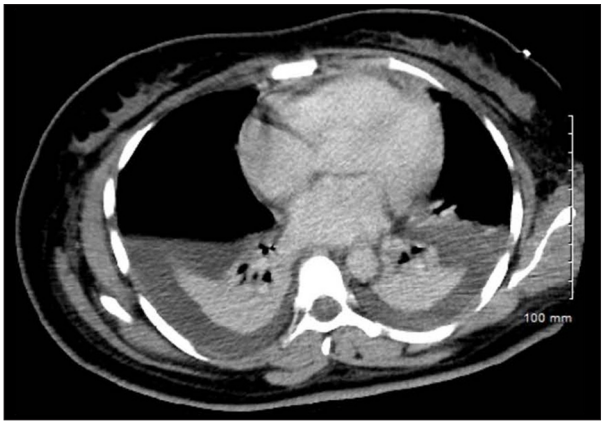

Figure 2. CT chest, abdomen and pelvis revealing bilateral pleural effusions with compressive atelectasis.

On day 14, after her creatinine normalized, a CT scan with contrast revealed new edematous appearance of both kidneys with diminished medullary enhancement consistent with underlying interstitial nephritis likely due to her earlier AKI (Figure 4). Imaging continued to show pleural effusions, however, subsequent thoracocentesis remained normal and negative for malignant/infectious etiologies. On day 22 chest x-ray revealed a new right lower lobe (RLL) infiltrate. Subsequently, a gallium scan was consistent with an inflammatory process of the lungs correlating to her infiltrates (Figure 5).

At this time, Adult Still's disease (ASD) was considered as one of the differentials, but given her origin from an underdeveloped country, presence of the lung infiltrates, and elevated WBC count, it was considered less likely. Rheumatology was

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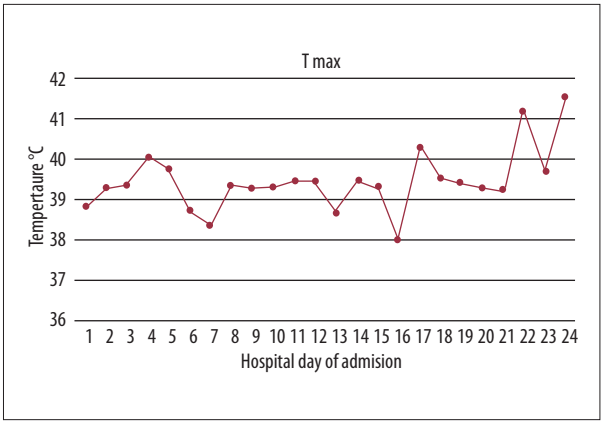

Figure 3. Temperature trends throughout the hospital stay reveals high-spiking fevers.

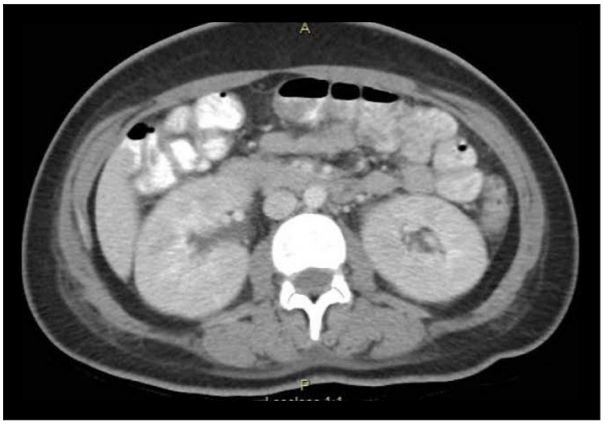

Figure 4. CT abdomen with contrast demonstrating edematous appearance of both kidneys with diminished medullary enhancement consistent with underlying renal disease/ glomerulonephritis/interstitial nephritis.

consulted and ordered a comprehensive panel of autoimmune markers, which were within normal limits with minor exceptions. Notably, several inflammatory markers were elevated including ESR (erythrocyte sedimentation rate), CRP (C-reactive protein) and a significantly elevated serum ferritin of 11,291. Table 2 shows a list of consults, dates, and tests performed throughout her hospital stay.

Throughout her hospital course, several symptoms were nearly universal and defined her hospital course. These included daily spiking fevers, respiratory symptoms, a rising and falling WBC count, and elevated transaminases and anemia. After a 24-day hospital stay at the community hospital, decision was made to transfer the patient to tertiary care institution for further diagnostic and therapeutic intervention.

Upon her arrival at the tertiary care center, the patient was admitted to the ICU, where she was noted by Rheumatology to have a faint rash, which in the context of her other symptoms

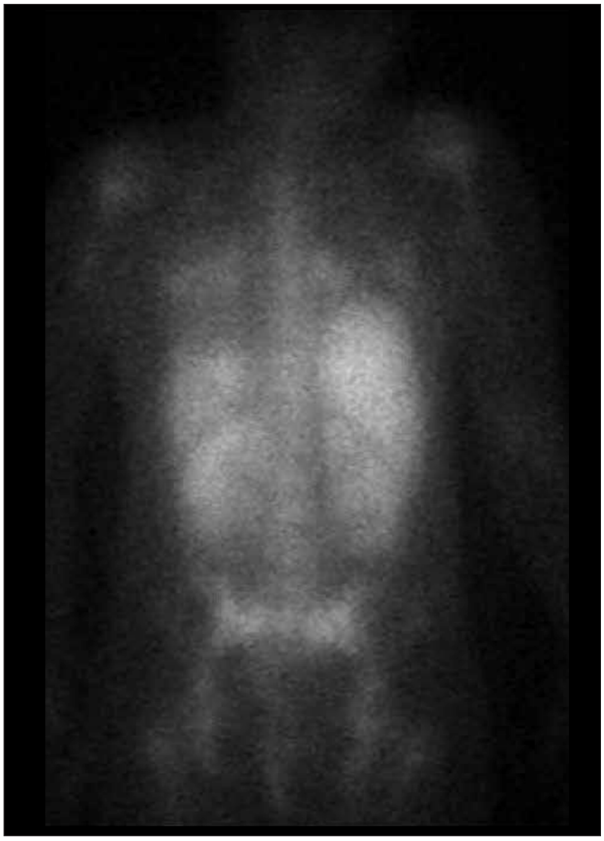

Figure 5. Gallium scan demonstrates findings consistent with an inflammatory process of the lungs.

further raised suspicion of ASD. The decision was made to start her on IV methylprednisolone. Her transaminitis was thought to be secondary to acute liver injury and repeat workup for acute viral and autoimmune hepatitis was found to be negative. Within 24 hours of starting IV methylprednisolone, the patient became afebrile and after three days of IV steroids she was started on prednisone 50 mg orally every day (PO qd) with subsequent discharge to home on a slow prednisone taper. The patient was followed as an outpatient and her liver tests remained elevated with an AST and ALT at 446 and 98 respectively, requiring outpatient hepatology follow up. At her three-month follow-up, her ferritin had returned to normal and was noted to be 29. She has had no new complaints or flares.

Discussion

This case illustrates the difficulties of diagnosing Adult Still's disease (ASD), primarily due to a lack of sensitive and specific serologic markers as well as nonspecific clinical presentation. Several cases have been reported in the literature documenting significant delays in diagnosis. Patients have initially been misdiagnosed with rhabdomyolysis and pneumonia in one case [3], and a case series of 20 patients found initial

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Table 2. Consulting departments, diagnostic tests and results throughout clinical course.

Diagnostic test and Department	Result	Reference value
• Cardiology		
Serial Troponin I	**Hospital day 1: 2.5 ng/mL** **Hospital day 3: 1.1 ng/mL** Hospital day 22: <0.3 ng/mL	<0.3 ng/mL
2D Echocardiogram	LV EF 60–65% **Moderate tricuspid valve regurgitation**	LV EF 60–65%
• Hematology		
Total proteins, serum	**5.8 g/dL**	6.0-8.5 g/dL
Total globulin	3.2 g/dL	2.2-3.9 g/dL
α1 globulin	0.3 g/dL	0.0–0.4 g/dL
α2 globulin	0.9 g/dL	0.4–1.0 g/dL
β globulin	0.9 g/dL	0.7–1.3 g/dL
γ globulin	1.0 g/dL	0.4–1.8 g/dL
Erythrocyte sedimentation rate	**>140 mm/h**	0-20 mm/h
Haptoglobin, serum	**599 mg/dL**	34-200 mg/dL
M spike	Not observed	Not observed
Immunoglobulins, serum		
IgA	216 mg/dL	70–400 mg/dL
IgG	2107 mg/dL	700–1600 mg/dL
IgM	93 mg/dL	40–230 mg/dL
Iron studies		
Iron Saturation, serum	**8%**	15–50%
Iron	**27 µg/DL**	35–150 µg/dL
TIBC	**119 mcg/dL**	250–450 µg/dL
Soluble transferrin	17.6 nmol/L	12.2–27.3 nmol/L
Serum ferritin	**11,291 ng/mL**	15–150 ng/mL
Serum creatine kinase MB isoenzyme	2.9 ng/mL	0.0–5.3 ng/mL
Folate (Folic acid, serum)	5.7 nmol/L	>3.0ng/mL
Immunofixation results	Polyclonal IgG gammopathy with κ & λ increase	
Lactate	**4.4 mmol/L**	0.4–2.0 mmol/L
Vitamin B12 (Cobalamin)	**1428 pg/L**	211–946 pg/mL
Platelet factor 4	**230 IU/mL**	<75 IU/mL
• Infectious Disease		
Whole blood culture	Negative	Negative for aerobic and anaerobic organisms
Parasite screen	Negative	Negative
Anti-Streptolysin O Ab	**345.1 IU/mL**	0.0–200.0 IU/mL

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Table 2 continued. Consulting departments, diagnostic tests and results throughout clinical course.

Diagnostic test and Department	Result	Reference value
Babesia microti PCR	Negative	Negative
C. difficile toxin stool	Negative	Negative
CMV IgG Ab	**9.10 U/mL (positive)**	<0.60 U/mL (negative)
CMV IgM Ab	<30.0 AU/mL (negative)	<30.0 AU/mL (negative)
Cryptococcal Ag	Negative	Negative
Dengue fever IgG/IgM	Negative	Negative
Ebstein Barr DNA quant PCR	0 copies/mL	0 copies/mL
EBV Ab Viral Capsid Antigen IgM	<36.0 U/mL (negative)	<36.0 U/mL (negative)
Ehrlichia	Negative	Negative
Hepatitis panel		
Hepatitis A IgM antibody	Negative	Negative
Hepatitis B core IgM antibody	Negative	Negative
Hepatitis B surface antigen screen	Negative	Negative
Hepatitis C antibody	<0.1 signal-to-cut off ratio	0.0-0.9 signal-to-cut off ratio
HIV screen 4th generation	Non-reactive	Non-reactive
Influenza panel		
Influenza A and B antigen	Negative	Negative
Influenza A/B PCR	Negative	Negative
L. pneumophilia serology antigen	Negative	Negative
Leptospira Ab	Negative	Negative
Lyme IgG/IgM Ab	Negative	<0.91ISR (negative)
Malaria Ag	Negative	Negative
Streptococcus A Antigen Rapid	Negative	Negative
Strongyloides	Negative	Negative
Throat culture for Streptococci	Negative	Negative
Tuberculosis panel		
Acid-fast bacilli (AFB) non-sputum stain reflex to culture	Negative	Negative
QuantiFERON-TB TB Ag minus nil value	<0.01 IU/mL (negative)	<0.35 IU/mL (negative)
QuantiFERON-TB TB Gold	Indeterminate	Negative
Zika Virus	Negative	Negative
• **Pulmonology**		
Arterial blood gases (ABG)		
pH	7.49	7.35-7.45
Arterial blood carbon dioxide (pCO_2)	43.0 mmHg	35–45 mmHg

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Table 2 continued. Consulting departments, diagnostic tests and results throughout clinical course.

Diagnostic test and Department	Result	Reference value
Arterial blood oxygen (pO_2)	112 mmHg	90–100 mmHg
Bicarbonate, serum venous (HCO_3^-)	**32.8 mEq/L**	22–26 mEq/L
Base excess (BE)	**8.5 mEq/L**	–2 to +2 mEq/L
Oxygen saturation	99%	90–100%
• **Radiology**		
Thoracocentesis	Negative on culture with no malignant cells. Cells comprised mainly of neutrophils, lymphocytes, mesothelial cells and macrophages.	
• **Rheumatology**		
Anti-nuclear antibody (ANA) direct	Negative	Negative
Anti-DNA SS IgG Ab	**146 EU (positive)**	<20 EU (negative)
Anti-DNA DS Ab	1 IU/mL (negative)	<9 IU/mL (negative)
Anti-GBM Ab	3 Units (negative)	0–20 Units
Atypical P-ANCA	<1: 20 (negative)	<1: 20 (negative)
C-ANCA	<1: 20 (negative)	<1: 20 (negative)
P-ANCA	<1: 20 (negative)	<1: 20 (negative)
C-reactive protein	**21.14 mg/dL**	<0.4 mg/dL
Complement, serum		
C3	**168 mg/dL**	90–180 mg/dL
C4	23 mg/dL	10–40 mg/dL
Creatine phosphokinase (CPK)	**19 U/L**	38–173 U/L
Cryoglobulin QI serum reflex	Negative	Negative
LDH	**411 U/L**	100–190 U/L

diagnoses to incorrectly include streptococcal tonsillitis/pharyngitis, infective endocarditis, sepsis, and bacterial meningitis [4]. One case series of 57 patients reported a mean time to diagnosis of four months [5]. This problem is further compounded especially in dark skinned patients as well as those originating from developing countries. As a result, the spectrum of differential diagnosis is wide, including primarily infectious, neoplastic and autoimmune etiologies.

Definitive diagnosis is based on the typical presentation of fevers, rash, myalgia and arthralgia, and the exclusion of other possible causes. Viral syndromes are usually excluded if the symptoms persist longer than three months, and tests for infection, including cultures and serologic test, can be helpful early in the disease course [6]. In the present case, infectious etiology was considered repeatedly, due to the patient being from a developing country and the presence of infectious markers on repeat radiologic investigations, however, serologic tests were consistently negative. ASD is frequently mistaken initially for infection as demonstrated in one series of patients presenting with fever of unknown origin (FUO), in which 90% of those who were eventually diagnosed with ASD had been treated with antibiotics [4].

Neoplastic processes – particularly hematologic malignancies – can also mimic ASD, and hematologic profiles usually help differentiate between these possible causes, however, bone marrow or lymph node biopsies may sometimes be necessary. Autoimmune conditions that may be confused with ASD include spondyloarthropathies, dermatomyositis, Kikuchi's syndrome, Sweet's syndrome, granulomatous disorders and vasculitides [6].

The triad of symptoms classically associated with ASD include high-spiking fevers, a characteristic rash, and arthralgia. The fever is usually greater than 39°C and follows a quotidian or double quotidian pattern. The rash typically appears as an

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evanescent, salmon-colored, maculopapular process, predominantly found on the trunk and proximal extremities. It may be pruritic and can be confused with a drug allergy [6]. Historically, the rash was considered to be one of the most useful features for diagnosis, however, due to its transient nature, it must be specifically looked for or it may be missed [7]. The incidence of rash has been shown to be between 51% and 87% [6]. The rash typical of Still's disease was never noted in our patient prior to transfer, and was either absent or may not have been appreciated. Most patients with ASD will also have associated arthralgia or arthritis, most commonly affecting the knees, wrists and ankles [6]. As was found in the present case, sore throat is also a common presenting symptom, with multiple case series demonstrating the finding in the majority of patients diagnosed with ASD [1,5].

Laboratory analysis typically reflects the non-specific systemic inflammatory condition, with leukocytosis, elevated ESR, CRP and ferritin [1]. Serum ferritin has been suggested as a promising marker for Still's disease, with the glycosylated fraction of ferritin potentially being more specific [6,8,9]. A serum ferritin greater than five times the upper limit of normal and a glycosylated ferritin level of ≤20% has a specificity of 92.9%; however, it has a sensitivity of only 43.2% [9]. Our patient demonstrated a significantly elevated ferritin level of 11,291. Unfortunately, ferritin is also elevated in infectious or neoplastic processes, and therefore only useful when extremely elevated or in the context of high clinical suspicion. Furthermore, glycosylated ferritin is not routinely checked during the diagnostic workup and was not ordered in the present case.

From the clinical and laboratory findings, multiple criteria have been proposed to aid in the diagnosis of ASD, including Yamaguchi, Fautrel, Cush and Calabro criteria [10–13]. The Yamaguchi criteria have become the most widely accepted, and is defined as the presence of five features, with at least two being major diagnostic criteria. Major diagnostic criteria include: Fever of at least 39°C (102.2°F) lasting at least one week, arthralgia or arthritis lasting at least two weeks, a non-pruritic macular or maculopapular skin rash that is salmon-colored in appearance, normally found over the trunk and extremities, especially with high fever, and a leukocytosis of ≥10,000/μL with at least an 80% granulocyte predominance. Minor criteria include: sore throat, lymphadenopathy, hepatomegaly or splenomegaly, abnormal liver function tests (particularly elevations in AST, ALT and LDH, and negative antinuclear antibody (ANA) and rheumatoid factor (RF) antibodies [10].

Once diagnosis is made, treatment generally consists of some combination of NSAIDs, glucocorticoids and anti-rheumatic agents [6]. There is a lack of randomized trials comparing treatment options; therefore, most data supporting treatment comes from retrospective series. Some patients with mild disease respond to NSAIDs. However NSAID monotherapy is frequently inadequate, with two studies demonstrating only 16% and 18% efficacy [5,14]. Most patients will require corticosteroid therapy, and corticosteroids may effectively control the disease in 63% to 76% of patients [14,15]. Initial dosage of prednisone ranges from 0.5–1.0 mg/kg/day, and response typically occurs within a couple hours to days [1]. Nonbiologic or biologic disease-modifying anti-rheumatic agents (DMARDs) may be necessary for long-term management or in cases refractory to treatment. Limited data exists to support the use of one DMARD over another; however, methotrexate is the most commonly used DMARD in ASD. In one study 88% of patients achieved partial remission of symptoms, while 69% achieved complete remission [16]. Another study demonstrated 73% efficacy with methotrexate monotherapy [14]. Methotrexate should be added when corticosteroids fail or in cases of steroid-dependence [1]. Other non-biologic DMARDs are also used, including cyclosporine, hydrochloroquine, azathioprine, and sulfasalazine, as either monotherapy or in combination [14].

Biologic agents represent an alternative treatment, and are typically suggested for patients who have had an inadequate response to corticosteroids and methotrexate. Multiple small case series have demonstrated remarkable efficacy with the use of infliximab – a TNF-α inhibitor – resulting in rapid remission of disease in all but one patient [17–19]. More modest results were demonstrated with infliximab in another case series, which showed a complete response in 27% of patients and a partial response in 60%, with minimal adverse effects [20]. Tocilizumab, an anti-IL-6 receptor antibody, has also been an effective treatment in patients who are cyclosporine resistant, and long-term therapy has been shown to induce remission of ASD [21]. Other patients resistant to treatment were effectively treated with anakinra, an IL-1 inhibitor. A small case series produced positive immediate outcomes of complete remission for all of the four cases treated with anakinra, with few to no side effects [22]. In patients with refractory ASD in which treatment with anakinra failed, canakinumab, a novel monoclonal antibody against IL-1β, demonstrated sustained efficacy in a very small series [23]. Larger prospective studies are needed to further assess efficacy and safety in the treatment of ASD.

Prognosis is usually good, even for patients suffering from chronic type disease, however, serious complications exist [3]. Macrophage activating syndrome (MAS), which is also referred to as reactive hemophagocytic lymphohistiocytosis (RHL) and other derivations, is an aggressive, potentially life-threatening syndrome of excessive immune activation and appears most frequently. Multiple retrospective studies have reported rates of occurrence ranging from 12% to 19% [5,24–26]. Other less frequent complications include myocarditis, cardiac tamponade, and constrictive pericarditis, endocarditis, shock, multiple organ failure, acute respiratory distress syndrome, intra-alveolar hemorrhage,

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disseminated intravascular coagulation, thrombotic microangiopathy and fulminant hepatitis [1]. Treatment of ASD may also be a source of complications, due to potential toxicities of various drug therapies. One study suggested that adverse events related to therapy occurred in 21% of patients on NSAIDs (predominantly gastrointestinal events), 75% of patients on corticosteroids (Cushing syndrome, aseptic osteonecrosis, steroid-induced diabetes, hypertension, infection), and 33% of patients on methotrexate (elevated liver enzymes, low blood counts and cough) [5]. In the present case, the patient's condition quickly improved with steroids and she is currently doing well.

In our patient, diagnosis was challenging, primarily because her constellation of symptoms did not immediately fit the typical pattern of Still's disease. Despite well-defined criteria for ASD it is not uncommon for atypical presentations to initially misguide diagnosis [3]. Our patient's presenting symptoms were fever, sore throat, tachycardia, and shortness of breath along with evidence of pneumonia and it significantly delayed consideration of ASD. Further complicating more expedient diagnosis was her immigration history from a developing country, which further raised suspicion of less common and rare infectious causes. This case emphasizes the need for high clinical suspicion of ASD, and exclusion of other etiologies early, so that immediate treatment can limit patient suffering and complications. Despite the relatively established base of literature on ASD, early diagnosis is still a challenge, and additional research is needed to identify patients early and identify any genetic predisposition that may also aid in diagnosis.

Conclusions

This case study demonstrates an atypical presentation of Still's disease in an adult patient (ASD) and the elusive nature of its diagnosis. Still's disease is a diagnosis of exclusion, and appropriate expedient workup is needed to rule out other etiologies for patients with this presentation. A key pattern of symptoms presented here should help clinicians in the future to consider the diagnosis earlier in the disease course. This includes high-spiking fevers resembling the profile of an infectious pattern but refractory to medical management, an extremely elevated ferritin, hepatomegaly, and a leukocytosis with granulocytic predominance.

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