CASE REPORT

A rare case of occult abdominal tuberculosis with Poncet's disease mimicking Adult onset Still's disease

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ABSTRACT

A 50-year-old female presented with fever, symmetrical arthralgias, rash, painful oral ulcerations and alopecia since 8 weeks. Examination showed mild hepatospleenomegaly. Investigations revealed leucocytosis, neutrophilia, elevated sedimentation rate and raised ferritin levels (3850 ng/ml). Computerized tomography (CT) abdomen showed hepatospleenomegaly, mild ascitis and mild bilateral pleural-effusion. After ruling out occult infections, tuberculosis, malignancies and autoimmune diseases by appropriate investigations, and due to raised ferritin levels, adult onset stills disease (AOSD) was diagnosed. Patient responded to oral steroids initially, but after 7 days developed severe abdominal pain. Repeat CT showed multiple enlarged, necrotic and matted retroperitoneal lymph nodes with caseating granuloma on histopathology suggesting tuberculosis. Patient was given four-drug anti-tubercular treatment and she improved. Thus our patient of occult abdominal tuberculosis with reactive arthritis (Poncet's disease) presented with hyperferritinemia mimicking AOSD. We postulate that extreme hyperferritinemia can be seen in tuberculosis and tuberculosis must be conclusively ruled out before diagnosing AOSD in tropics.

Key Words: Abdominal tuberculosis, adult onset stills disease, serum ferritin, serum ferritin levels

INTRODUCTION

World's largest number of Tuberculosis (TB) patients reside in India which makes around 26% of world TB population.[1] Of all extrapulmonary TB, abdomen is involved in 11% of cases. Gastrointestinal tract, peritoneum, lymph-node, liver, spleen and genitourinary tract can be involved in abdominal TB. Abdominal TB is a great mimicker and often difficult to diagnose. Because of non-specific signs and symptoms and frequent false negative results with standard tests, it is often confused with gastrointestinal inflammatory, malignant, infectious as well as autoimmune diseases and diagnosis is frequently delayed.

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CASE REPORT

A 50-year-old female patient presented with high grade fever and arthralgias since 8 weeks. Bilateral metacarpophalyngeal, wrist and large joints were swollen, painful and had morning stiffness lasting 30 minutes. Two weeks prior she developed a non-pruritic, non-photosensitive macular rash over the trunk which resolved spontaneously. Over the preceding 4 weeks, she was having severe anorexia, malaise, multiple painful oral ulcers and had lost 7-kg weight. Patient also complained of alopecia [Figure 1] developing with the onset of fever. There was no history of cough, diarrhea, vomiting, abdominal pain or any other significant infection.

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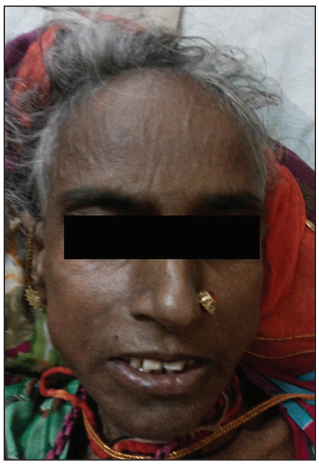

Figure 1: Figure showing non scarring alopecia

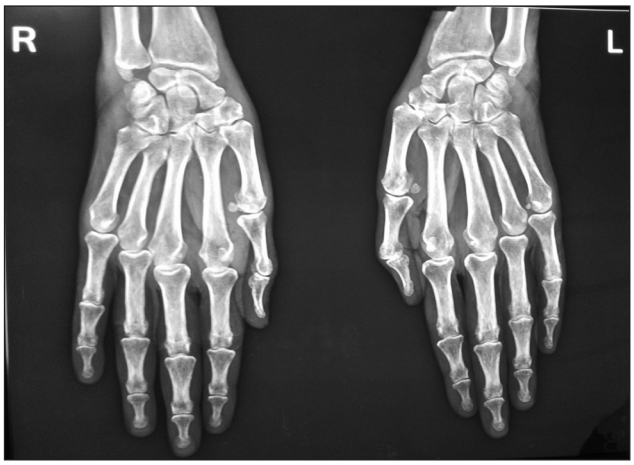

Figure 2: X-rays of both wrist joints and hands (AP view) showing mild periarticular soft tissue swelling with no evidence of destruction or joint space narrowing

Patient took treatment initially at a local health centre with acetaminophen 1.5 g/day for 5 days and amoxicillin 1.5 g/day for 5 days but with no relief, after which she was referred to us. Examination revealed oral temperature of 39°C, mild pallor, non-scarring alopecia and mild hepatospleenomegaly. Wrist, knee and ankle joints were bilaterally swollen and tender without any joint restriction. Rest of general and systemic examination was normal.

Investigations [Table 1] revealed, normocytic normochromic anemia and leucocytosis with marked neutrophilia. Erythrocyte sedimentation rate (ESR) was raised with very high serum ferritin levels (3850 ng/ml), which was repeated twice showing similar values. Except for mildly elevated transaminase levels rest of the liver and renal function tests, chest radiograph and urine examination was normal. Abdominal sonogram revealed mild hepatospleenomegaly with bilateral minimal pleural effusion and ascitis. CT chest and abdomen revealed the same findings. Tests for other infectious and autoimmune diseases were negative. Sputum smear for acid fast bacilli (AFB), TB-PCR (polymerase chain reaction) and adenosine deaminase (ADA) of ultrasound-guided aspirated ascitic fluid were all negative. Hand radiographs showed mild soft tissue swelling without any destruction [Figure 2]. She was started with injectable pipracillin-tazobactam 4.5 gm q*uarter in die* (QID) with metronidazole 200 mg *ter in die* (TID) and doxycycline 200 mg *bis in die* (BID) along with supportive treatment. Repeated blood counts during her stay in the hospital after starting antibiotics showed no decline in the levels. Keeping in view the very high serum ferritin levels along with fever, arthralgias, macular rash, elevated liver enzymes and erythrocyte sedimentation rate with neutrophilia, a presumptive diagnosis of AOSD was made fulfilling Yamaguchi criteria [Table 2]. After 14 days of antibiotics and no response she was started with oral prednisolone 1 mg/kg/day. Initially patient responded dramatically and arthralgias disappeared within 2 days. Seven days later, patient developed severe abdominal pain and vomiting when repeat CT abdomen showed multiple enlarged and matted mesenteric and periportal lymph-nodes with mid ascitis [Figure 3]. Laparoscopy guided lymph-node biopsy revealed caseating granulomas without AFB [Figure 4] and the tissue was sent for mycobacterial culture. As CT-chest showed no evidence of any pulmonary parenchymal or thoracic lymph node involvement, a fungal etiology was unlikely. Considering the fact that the most common cause of caseating granuloma in India is tuberculosis, and that abdominal tuberculosis is paucibacillary, we kept the provisional diagnosis of abdominal tuberculosis. Patient improved significantly with four drugs anti-tubercular therapy for 6 months, and is asymptomatic under our regular follow-up over the last 8 months. Culture reports received after 10 weeks were positive for *Mycobacterium tuberculosis*. Repeat ferritin levels after 6 months of ATT were 190 ng/ml. Patient was advised repeat laparoscopy, but did not consent for the same as she had improved.

DISCUSSION

AOSD is a rare multi-systemic inflammatory disorder of unknown etiology first described in 1971. It is characterized by fever, arthritis, evanescent skin rash, sore throat and occasional lymphadenopathy, hepatospleenomegaly and serositis. It is associated with marked leucocytosis (neutrophilia), raised serum ferritin levels and abnormal liver function tests. Diagnosis of AOSD is based on Yamaguchi's criteria[2] [Table 2]. Before diagnosing AOSD, infectious, malignant, autoimmune and vasculitic disorders should be ruled out. Ferritin is an intracellular iron binding protein and acute phase reactant whose levels in the body are regulated mainly by levels of iron, cytokine release and chemokine production. It is raised in several acute and chronic inflammatory conditions, however very high

Table 1: Table showing laboratory investigations

Hemoglobin		9.2 g/dL	
Total leucocyte count (N, L, M, E)		18,480/cumm (80%, 17%, 01%, 02%)	
Platelet counts		3.7 lakh/cumm	
ESR and CRP		130 mm and 80 mg/dL respectively	
Liver Function Tests	ALT/AST/ALP/GGT	340 IU/L, 82 IU/L, 250 IU/L and 20 U/L respectively	
	Serum albumin and globulin	2.5 gm/dl and 3.2 gm/dl respectively	
PT-INR and APTT		Normal	
Serum creatinine/Blood urea		0.8 mg/dl and 45 mg/dl respectively	
Serum Ferritin levels		3850 ng/ml (normal 50-200 ng/ml)	
Radiographs	Chest X-ray	Normal	
	Hand X-ray	Mild soft tissue swelling.	
Sputum AFB		Negative	
Ultrasound abdomen		Hepatospleenomegaly with mild ascitis and mild B/L pleural effusion	
Ascitic fluid examination		WBC-200/mm^3 (N-50%, L-45%, M-5%), albumin 2.0 gm/dL	
TB-PCR and ADA of ascitic fluid		Negative and ADA 12U/L respectively	
Rheumatoid factor(RA) and Anti-CCP		4.5 IU/ml (0-20 IU/ml normal) and Negative respectively	
HIV ELISA/ Anti-HCV IgM andIgG/HbsAg and Anti-Hbc IgM/VDRL		All Non reactive	
ANA		8.86 (normal <20)	
APLA		Negative	
ASMA and Anti-LKM-1		Negative	
p-ANCA and c-ANCA		Negative	
TSH		1.42 μIU/mL	
Transesophageal echocardiography		Normal	
Blood cultures for bacteria(done thrice)		Negative	
CT chest and abdomen		Initial suggestive of mild hepatospleenomegaly with bilateral mild pleural effusion and ascitis	Done 10 days later suggested multiple enlarged and matted portocaval, celiac, par aortic and retrocaval lymph nodes with mild ascitis and bilateral pleural effusion.
Bone marrow aspiration, biopsy and culture		Myeloid hyperplasia with no infiltration, granuloma and no organism isolated on culture	
Mycobacterial culture (from tissue)		Positive	

ESR: Erythrocyte sedimentation rate at end of 1st hour, CRP: C-reactive protein, ALT: Alanine transaminase, AST: Aspartate transaminase, ALP: Alkaline Phosphatase, GGT: Gamma glutamyl transferase, AFB: Acid fast bacilli, TSH: Thyroid stimulating hormone, HIV ELISA: Human immunodeficiency virus enzyme linked immunosorbent assay, Anti-HCV: Anti hepatitis C virus antibody, HBsAg and Anti-HBc IgM: Hepatitis B surface antigen and hepatitis B core antibody respectively, VDRL: Venereal disease research laboratory, APLA: Anti-phospholipid antibody, ASMA: Anti smooth muscle antibody, Anti-LKM: Anti liver kidney microsome antibody, p-ANCA and c-ANCA: Perinuclear and cytoplasmic anti-neutrophilic cytoplasmic antibody respectively, ADA: Adenosine deaminase.

Table 2: Table showing Yamaguchi's criteria

Yamaguchi's criteria	
Major criteria	**Minor criteria**
Arthralgias >2 weeks	Sore throat
Fever >39° intermittent, >1 week	Lymphadenopathy and /or Spleenomegaly
Typical rash*	Abnormal liver function tests
WBC >10,000/cumm (>80% granulocytes)	Negative rheumatoid factor and ANA
Diagnostic combination	
Exclusion criteria	Diagnosis
Infections	5 criteria with (at least 2 major)
malignancy	
Rheumatic disease	

*Evanescent, salmon-pink, maculopapular eruption, predominant on the proximal limbs and trunk

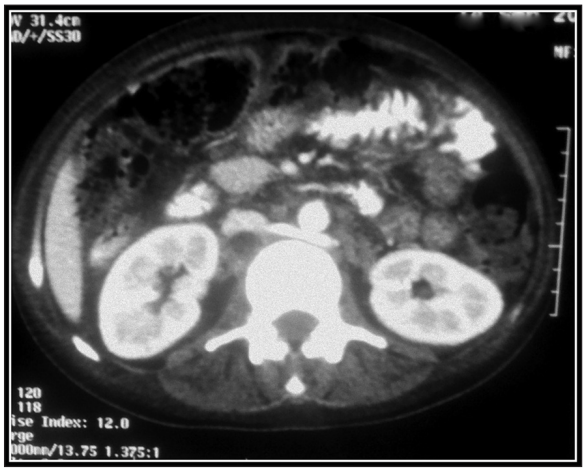

Figure 3: Contrast Enhanced Computed Tomography (CECT) abdomen showing multiple enlarged and necrotic retroperitoneal lymph nodes

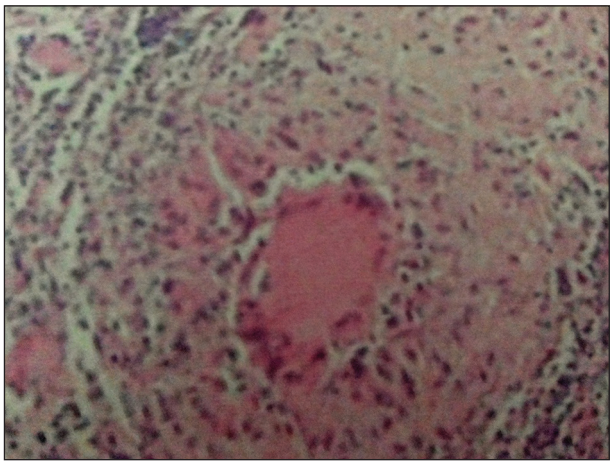

Figure 4: Histopathology of abdominal lymph node showing granuloma with central caseation

serum ferritin levels [> five times normal (>1000 ng/ml)] are associated with four uncommon disorders namely, the macrophage activation syndrome, AOSD, catastrophic antiphospholipid syndrome and septic shock.[3] Serum ferritin levels are marginally increased in isolated TB infection however levels upto 10,000 ng/ml have been reported in HIV and TB coinfection.[4] The diagnosis of abdominal and milliary TB can be challenging due to varied clinical presentations. Rai *et al*.,[5] in their study concluded that laboratory studies, tuberculin skin tests, Ziehl-Neelsen (ZN) staining and CT scans can give false negative results in cases of abdominal tuberculosis. Although ascitic fluid ADA levels more than 32 IU/L (sensitivity >90%) are considered confirmatory in TB, false negative results increases in patients with decreased ascitic fluid protein concentration.[6] Similarly sensitivity of TB-PCR in ascitic fluid is around 93% but with negative AFB staining, sensitivity can drop to as low as 33%.[7] Acid-fast bacilli can be seen on histological examination by ZN staining in only 6-8% of patients.[8] Tissue and fluid cultures are positive in only 20% of cases of Abdominal TB.[1]

Abdominal tuberculosis is considered as great mimicker and is difficult to diagnose. Since abdominal tuberculosis is paucibacillary, histopathological evidence of caseating necrosis can be taken as diagnostic for starting AKT till culture reports are awaited in cases of strong clinical suspicion especially in endemic areas. In our patient of abdominal tuberculosis none of the above mentioned investigations were helpful and patient's clinical findings mimicked AOSD. Also marked non-scarring alopecia, painful oral ulcers and symmetrical arthralgias associated with elevated ESR and quick response to steroids pointed towards an autoimmune etiology. Since the incubation period of TB is around 2-12 weeks, chances of patient getting secondarily infected during course of steroids are less. Arthralgias in this patient could be a manifestation of TB. Poncet's disease, a rare form of aseptic arthritis can develop in a case of active tuberculosis. Clinically it resembles reactive arthritis and can involve any joint but large joints are involved more often. The mechanism behind it is thought to be immunological.[9]

As per our literature search, there has been only one case report of tuberculosis with extreme hyperferritinemia mimicking AOSD in which the patient had hepatic tuberculosis.[10] Such atypical manifestation of not so easily diagnosed abdominal tuberculosis in endemic areas can pose a major diagnostic challenge and delay the necessary treatment.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Debi U, Ravisankar V, Prasad KK, Sinha SK, Sharma AK. Abdominal tuberculosis of the gastrointestinal tract: Revisited. World J Gastroenterol 2014;20:14831-40.
2. Yamaguchi M, Ohta A, Tsunematsu T, Kasukawa R, Mizushima Y, Kashiwagi H, *et al*. Preliminary criteria for classification of adult Still's disease. J Rheumatol 1992; 19:424-30.
3. Rosário C, Zandman-Goddard G, Meyron-Holtz EG, D'Cruz DP, Shoenfeld Y. The hyperferritinemic syndrome: Macrophage activation syndrome, Still's disease, septic shock and catastrophic antiphospholipid syndrome. BMC Med 2013;11:185.
4. Visser A, van de Vyver A. Severe hyperferritinemia in Mycobacteria tuberculosis infection. Clin Infect Dis 2011; 52:273-4.
5. Rai S, Thomas WM. Diagnosis of abdominal tuberculosis: The importance of laparoscopy. J R Soc Med 2003;96:586-8.
6. Fernandez-Rodriguez CM, Perez-Arguelles BS, Ledo L, Garcia-Vila LM, Pereira S Rodriguez-Martinez D. Ascites adenosine deaminase activity is decreased in tuberculous ascites with low protein content. Am J Gastroenterol 1991;86:1500-3.
7. Chau TN, Leung VK, Wong S, Law ST, Chan WH, Luk IS, *et al*. Diagnostic challenges of tuberculosis peritonitis in patients with and without end-stage renal failure. Clin Infect Dis 2007;45:e141-6.
8. Prakash A. Ulcero-constrictive tuberculosis of the bowel. Int Surg 1978;63:23-9.
9. Malaviya AN, Kotwal PP. Arthritis associated with tuberculosis. Best Pract Res Clin Rheumatol 2003;17: 319-43.
10. Manoj EM, Srigrishna R, Ragunathan MK. Hepatic tuberculosis presenting with extreme hyperferritinemia masquerading as adult-onset Still's disease: A case report. J Med Case Rep 2012;6:195.