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Poncet rheumatism secondary to ceco-appendicular tuberculosis in systemic lupus erythematosus

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ABSTRACT

Background. Poncet's rheumatism (RP) is an aseptic reactive arthritis secondary to active tuberculosis. Systemic lupus erythematosus (SLE) and tuberculosis are intricately related with an increase in the risk of tuberculosis in SLE. We present an original case of Poncet's disease secondary to appendicular tuberculosis in patient with lupus disease.

Case report. A 36-year-old female was followed in internal medicine for systemic lupus. In September 2023, she suffered from digestive complaints and polyarthritis. Infectious investigation and microbiologic study of joint fluid was negative. She had tenderness on pressure on the right iliac fossa. The computed tomography (CT) found an appendicular mucocele. She underwent laparotomy with right colectomy. Macroscopic aspect revealed tumorous lesion suggestive for malignancy. Histological study concluded to granulomatous inflammation with epithelioid and gigantocellular granulomas with anhist necrosis. Ceco-appendicular tuberculosis was retained. The patient received the anti-tuberculosis therapy with good progress.

Conclusions. Early recognition of atypical location of tuberculosis leads to initiate appropriate treatment especially in immune compromise statue patient.

Keywords: Poncet rheumatism, tuberculosis, appendicular tuberculosis, lupus, polyathritis

Abbreviations

CT	– computed tomography	PR	– Poncet's rheumatism
GN	– glomerular nephritis	SLE	– Systemic Lupus erythematosus
NSAIDs	– non-steroidien anti inflammatory drugs		

INTRODUCTION

Poncet's rheumatism (RP), also known as tuberculosis-associated arthritis, is an aseptic reactive arthritis secondary to active tuberculosis. It should be retained after ruling out osteoarticular tuberculosis infection [1]. This is a frequently forgotten entity. Systemic lupus erythematosus (SLE) is an autoimmune disease that is frequently treated with corticosteroids and other immunosuppressive drugs. This patients with SLE are at increased risk for infections with several pathogens including *Mycobacterium tuberculosis.* There are no established guidelines available for treatment of tuberculosis in SLE. We present a case report of Poncet's disease secondary to appendicular tuberculosis in patient with SLE. We underline the importance of early recognition of this unhabitual complication of evolutive tu-

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berculosis to avoid delayed initiation of appropriate treatment [1].

CASE REPORT

A 36-year-old female was initially admitted in our department of internal medicine in 2012 for fever and arthritis. The diagnosis of systemic lupus was established based on facial skin rash, pleural effusion, pericarditis, articular involvement, auto immune hemolytic anemia, lymphopenia, positive antinuclear antibodies with anti-DNA antibodies. She was treated with oral steroids at the dose of 1mg/kg/day started by 3 pulses of methylprednisolone in combination to hydroxychloroquine with good outcomes.

In 2015, she was hospitalized for renal flare. She was presented with positive proteinuria at 1.5g/24h without renal failure. Kidney biopsy revealed proliferative glomerular nephritis (GN) with high index activity. She was treated by pulses of steroids then relayed by high doses of oral steroids in with immunosuppressive therapy based in monthly intravenous pulses of cyclophosphamide 1g/ month received for 6 months then switched to azathioprine (150mg /day). Three years later, in 2018, she presented a second renal flare because of the poor compliance of treatment. Kidney biopsy revealed proliferative glomerulonephritis class IV. The treatment included high doses of steroids maintained for 6 weeks then progressively tapered until 10 mg /day of prednisone in association with cyclophosphamide (12 monthly pulses of 1g then relayed by 4 quarterly maintenance cures) with good progress. In May 2022, she was readmitted in our department for neurological flare. She had cerebellar syndrome with divergent strabismus and ptosis of the right eye. Magnetic resonance imaging identified neurological damage in infratentorial area and inflammatory lesion in nucleus of third nerve. Therapy was based in intravenous pulses of methylprednisolone followed by high-doses of oral steroid combined with azathioprine. In her follow up, neurological examination became normal. Four months later, in September 2023, she suffered from post prandial vomiting, fleeting abdominal pain, paroxysmal diarrhea and fever. These digestive complaints were concomitant with arthritis of the wrists, fingers knees and tenosynovitis of the thumb flexors. Infectious investigation including viral serology (HIV, HBV HCV) and microbiologic study of joint fluid was negative. Transthoracic ultrasound was normal. Biological analysis founded lymphopenia at 400 elements/mm^3 and high rates of inflammatory markers. The SLEDAI score was elevated at 6 points and the disease was considered as active. Therapeutic decision was to increase the dose of steroids at 30 mg of prednisone. Nevertheless, patient didn't get any improvement in joint involvement. One month after, in October 2023, she was presented in emergency department for acute abdominal pain. Examination showed tenderness on pressure on the right iliac fossa. The computed tomography (CT) found an appendicular mucocele with suspected abscess. (Figure 1). Then, she underwent surgery with small incision in the lower right abdomen which was transformed into an open large laparotomy with right colectomy. Macroscopic aspect revealed tu-

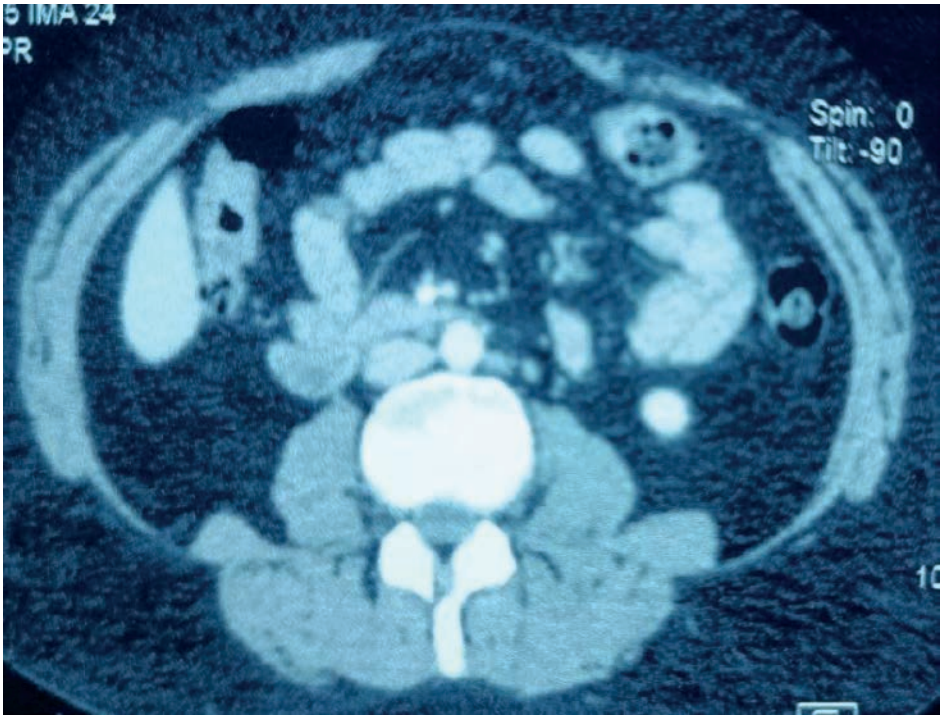

FIGURE 1. Computed tomography revealing mucocele aspect of appendix

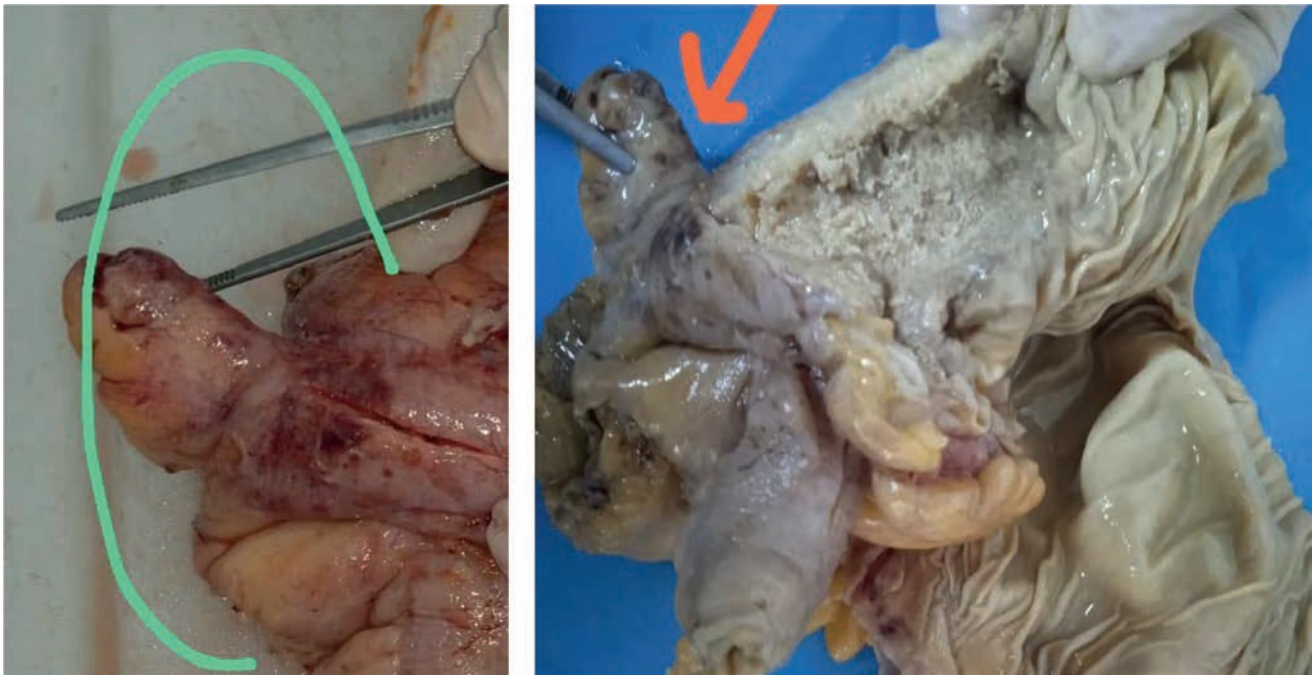

FIGURE 2. Pseudotumoral macroscopic aspect of the appendix

morous lesion suggestive of malignancy. (Figure 2) Histological study concluded to granulomatous inflammation with epithelioid and gigantocellular granulomas with anhist necrosis entitled "caseum" in coeco-appendicular wall. The colonic and ileal walls were respected. The surgical margins were either healthy. Ceco-appendicular tuberculosis was retained. Screening of other sites of infection found a crusty ulcer in the thumb which was suggestive of cutaneous involvement. (Figure 3). In addition, pulmonary involvement was retained face to the micronodules observed in the apex of the lung in computed tomography of the chest. (Figure 4)

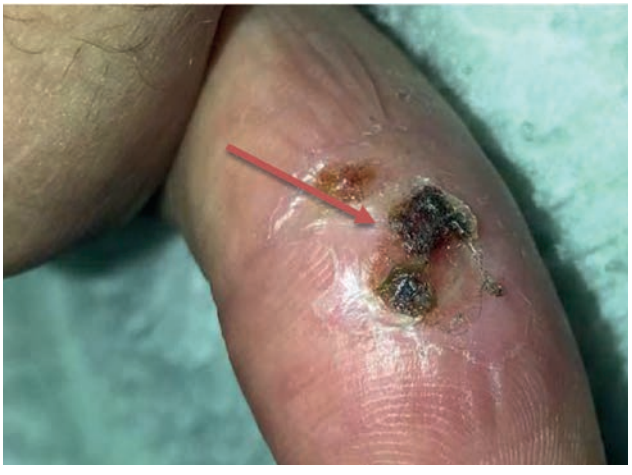

FIGURE 3. Crusty lesion of the thumb

Multifocal tuberculosis was diagnosed. The patient received anti-tuberculosis quadritherapy for two months then bitherapy. Hand radiography during the development of polyarthritis. Hand radiography showed neither bone erosion nor joint space narrowing. (Figure 5)

Arthralgia, arthritis and general statue were significantly improved after a follow up of 12 months.

DISCUSSION

Poncet's Rheumatism in a nondestructive or nonerosive inflammatory arthritis that may follow mycobacterial infection elsewhere with no direct infective agent identified in the involved joints. It was initially described in 1897 by Antonin Poncet. It's defined as an inflammatory arthritis secondary to distant mycobacterial active infection without identified infectious agent [2]. Until today, Poncet's disease remains a diagnosis of exclusion, after ruling out the known causes of polyarthritis [3]. The diagnosis criteria proposed [4-6] included the evidence of active extra-articular tuberculosis with no specific laboratory findings and no chronicity in joints, and complete resolution after antituberculosis therapy. Its frequency is considered as little known and often forgotten disease, especially in countries with low tuberculosis incidence [1,6]. Rueda and co-authors identified less than 200 cases published in the literature [5]. India was the country with the most frequent cases (70 cases), followed by Brazil (26 cases) then Mexico (20 cases). The high frequency in Asia and south America might reflect genetic predisposition. Its pathogenesis was not well understood. The hypothesis suggested was the cross-reactivity between mycobacterial antigens and cartilage proteoglycans by T-cell mediated possible mechanism. A genetic predisposition in patients with HLA-DR3 and HLA-DR4 haplotype has been proposed as being

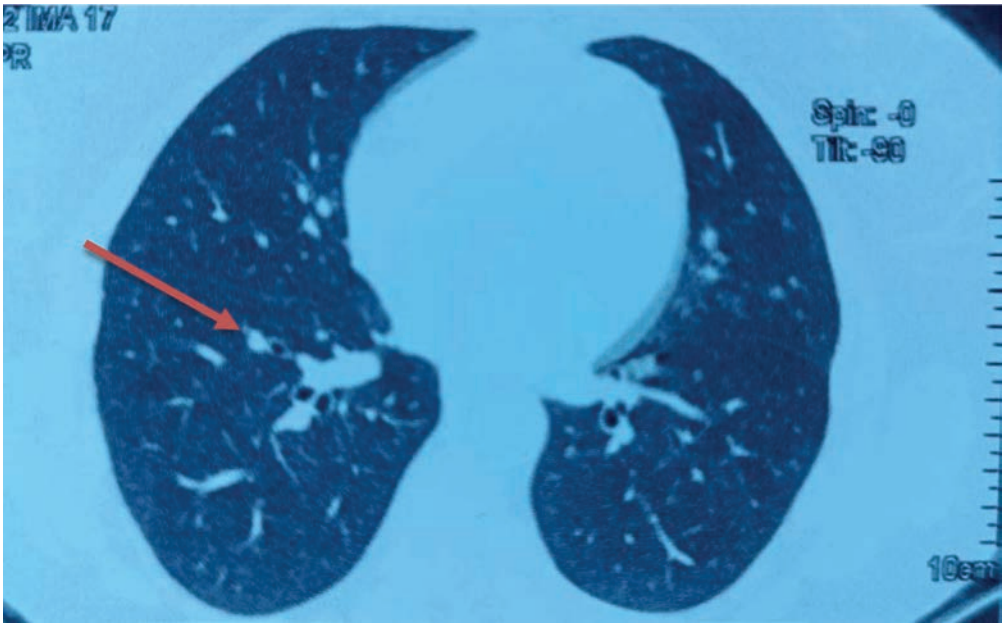

FIGURE 4. Chest computed tomography: micronodular lesions

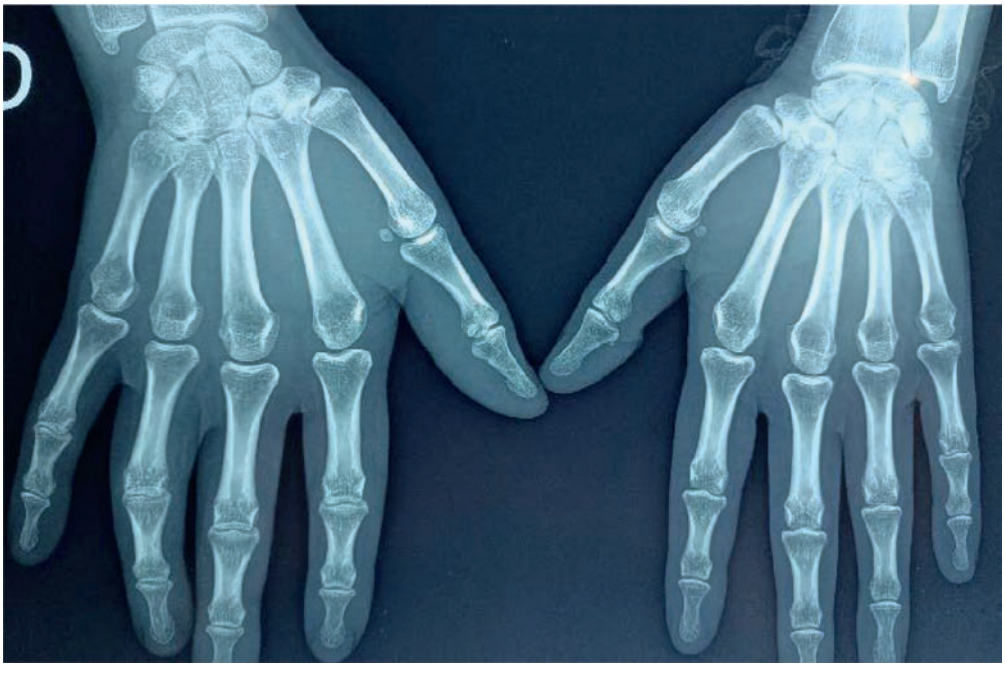

FIGURE 5. Hand X-ray demonstrating no destruction

hyper-responsive to mycobacterial antigens [5-7]. Tuberculosis was largely considered as a multisystemic disorder resulting in pulmonary and extrapulmonary signs. This infection can be presented in various forms and depends on inherent host conditions and on the properties of *Mycobacterium tuberculosis.* Skin involvement occurs in 1 to 2% of cases through direct involvement of tissues (as skin pustules, ulceration, nodular lesions) or via hypersensitivity reaction. About 10 to 11 % of extrapulmonary tuberculosis involves joints and bones [8]. Arthritis in tuberculosis infection occurs the most commonly through direct joint involvement usually as a chronic mono-arthritis [6]. Diagnosis requires often joint biopsy. Sterile reactive polyarthritis can be observed in active tuberculosis especially with immunocompromised state like HIV infection and patient with inflammatory diseases receiving or not steroids (like lupus, scleroderma or rheumatoid arthritis). This sterile reactive arthritis secondary to distant active tuberculosis is not well known in the literature and was limited to some cases [9]. In the Kroot study, 30% of patients were presented with oligoarthritis (less than 4 joints) often asymmetrical and the other patients were presented with polyarthritis. It may affect small joints of the hands, particularly the metacarpophalangeal bones [7]. In 48% of the patients, the tuberculosis was extrapulmonary.

In the Sharma's study [4] including 23 patients, pulmonary tuberculosis was found in 5 patients (21.73%) and extrapulmonary tuberculosis was found in 18 cases. 39.13% had mediastinal lymphadenopathy on imaging, 8.7% had cervical lymphadenopathy, one had abdominal lymphadenopathy and one had ileo-cecal tuberculosis.

The site of primary tuberculosis was not identified in 17.39% even after imaging with contrast-enhanced computed tomography (CT) scan of the body. The optimal treatment of Poncet's syndrome was not established. The main medication was based on anti-tuberculous therapy. Resolution of arthritis with anti-tuberculous drug ranged from 1 week to 4 months [9]. It had a good prognosis with a favorable outcome under anti bacillary treatment. Some cases require the adjunction of NSAIDs (nonsteroidal anti-inflammatory drugs) with the continuation of anti-tuberculosis therapy [9]. Previous reports have suggested steroids and methotrexate as an effective therapy [10]. Early diagnosis with initiation of therapy contribute to good prognosis. Poncet disease has good prognosis and does not progress into chronic arthritis. In our case, joint symptoms improved few days after specific anti-tuberculosis treatment. In contrary, septic arthritis can lead to erosive arthropathy, osteomyelitis and cutaneous fistula. Diagnosis difficulties represent a challenge to don't miss out diagnostic gap of almost 3 million undiagnosed tuberculosis cases globally [6]. it should be considered as a differential diagnosis for patients with fever and arthritis of unknown cause. Rheumatoid arthritis can mimicking this disease when anti-citrullinated peptide antibodies are elevated but this two diseases could be co-existent [11]. Patients with systemic lupus erythematosus are at increased risk of developing tuberculosis due to their underlying immunodeficient state and treatment with immunosuppressive drugs. The risk is higher in endemic regions and among patients with nephritis, high disease activity and high cumulative doses of corticosteroids [12]. Tuberculosis may manifest differently from immunocompetent patients. In pulmonary tuberculosis the miliary form was the most described [13]. Therefore, regular monitoring of these patients for active tuberculosis in endemic areas is necessary to treat infection in the initial stage before dissemination. It imparts a challenge to the clinical acumen especially in the setting of higher number of cases of extra pulmonary tuberculosis. The diagnosis of Mycobacterium Tuberculosis is determined on the basis of analysis of body fluids and tissues which often takes a prolonged time; thus, delaying the diagnosis. Another aspect of tuberculous presentation is a constellation of non-specific symptoms like unexplained fever, joint pain, fatigability and serositis which are also seen in patients with SLE, making the diagnosis more difficult. Previous studies have shown that, the time interval between tuberculosis onset and diagnosis may vary from one month to up to one year [14-15].

CONCLUSION

In conclusion, our present patient developed Poncet's disease and she rapidly improved with anti-tuberculous therapy. Our report highlights the strength of histopathological examination in establishing diagnosis. Moreover, we can conclude that patients with systemic lupus had a compromised immune statue requiring more attention into tuberculous infection. It should be evocated in patients receiving steroid therapy. To our knowledge, we report the first case in the literature of Poncet's rheumatism complicating appendicular tuberculosis in patient with SLE.

Patient consent: The authors undersign and certificate that they have obtained the written consent of the identified persons or their legal guardians for the presentation of the cases within the present scientific paper

Conflict of interest: The authors undersign and certificate that they do not have any financial or personal relationships that might bias the content of this work.

Author's contributions:
MS. RBS, MBH, AD: collected the data and write the manuscript
CD, ZB, SM: validate the manuscript
AZ, WF: collected the iconography.
All authors have read and agreed to the published version of the manuscript.

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