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Prolonged Fever: Case Report

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ABSTRACT
Background: Sarcoidosis is a granulomatous disease with unknown cause that can vary from an asymptomatic condition. Almost half of the patients with sarcoidosis have no symptoms.
Case report: 8 year-old female, referred from Abha Hospital as a case of prolonged fever. The fever was low grade, lasted for months, mostly at night, and was associated with fatigue and no weight gain. Systemic review showed abdominal distension, history of knee pain, mouth ulcers and temporary rash on feet.
Conclusion: Sarcoidosis is a disease that mostly affects the lung, but sometimes it is outside the lung, as in our cases. Doctors should pay attention to this so that the wrong diagnosis does not lead to complications or death of the patient.
Keywords: Prolonged fever, Sarcoidosis.

INTRODUCTION

Sarcoidosis is an idiopathic multisystem granulomatous disease that affects patients of all races and ethnic groups however predilection for women and African Americans is apparent. Extrapulmonary manifestations of sarcoidosis occur in up to 50% of cases. The most common sites of extrapulmonary involvement are the skin, eyes, liver, and reticuloendothelial system followed by renal, cardiac, and neurological involvement [1-6]. Only 8% of patients present with isolated extrapulmonary disease in the absence of pulmonary involvement with the most common presentation in this group being isolated cutaneous sarcoidosis [4, 6].

Our case being presented is a unique manifestation of nonpulmonary sarcoidosis with abdominal distension with hepatomegaly on examination, and palpable cervical lymph nodes.

CASE REPORT

8 year-old female, referred from Abha Hospital as a case of prolonged fever. The fever was low grade, lasted for months, mostly at night, and was associated with fatigue and no weight gain. She had abdominal distension with hepatomegaly on examination, and palpable cervical lymph nodes, history of knee pain but no current swelling, history of mouth ulcers and history of temporary rash on feet. Family history was significant for lymphoma in the maternal uncle.

She was admitted for total of 9 days and treated with antibiotics. She improved but then fever recurred. Bone marrow biopsy was done: was reported as unremarkable. She was followed up in OPD: suspected to have familial Mediterranean fever. Colchicine was prescribed for around 2 months, with which the fever improved. Yet, her ESR remained elevated, so an alternative diagnosis was suspected, and therefore she was referred her to our hospital (Children's Specialized Hospital, King Fahad Medical City).

History of Presenting Illness:
Fever:
- Low grade (at home: 38°C).
- Occurred mostly at night.
- Progressed later on and lasted full days.
- Responded to antipyretics temporarily.
- Associated with lethargy and decreased activity.
- Lasted a total of 7-8 months.

Past Medical and Surgical History:
- At 6 months of age: had BCG abscess (required drainage) with axillary lymph node enlargement.
- Not on any medications.
- No similar complaints before.
- Was admitted in Abha Hospital for 9 days then discharged.
- Not known to have allergies.
- No surgical history.
- Vaccinations were up to date.

Family History:
- Mother and father are healthy
- Siblings are healthy
- Maternal uncle had lymphoma at 12 years, treated with chemotherapy
- No history of sibling deaths
- No history of congenital or metabolic diseases in the family

Investigations:
CT of the chest abdomen and pelvis showed:

Enlarged lymphadenopathy throughout the body, the largest in the neck is seen at the lower cervical region on the left side approximately measuring 0 7 cm,

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the largest in the chest is pericardiac lymph node that measures up to 1 2 cm, the largest in the abdomen is at the level of the para-aortic region measuring almost 1 1 cm in addition to multiple variable size mediastinal, gastrosplenic, porta hepatis, mesenteric, iliac and inguinal prominent lymph nodes.

Noted multiple right lung upper and middle lobe pulmonary nodules with the largest measuring 0.6 x 0.6 cm. Also in multiple tiny penpheral pleural based lung nodules noted through the left lung measuring about to 0.4 cm.

There is a large fibro- atelectatic band seen at the anterior segment of the right lower lobe Major airways are patent.

Heart and great vessels of the thorax vessels are within normal limits. No pleural, pericardial effusions or pneumothoraces.

Liver and spleen are enlarged, they are measuring 19.3 cm and 12.5 cm respectively.

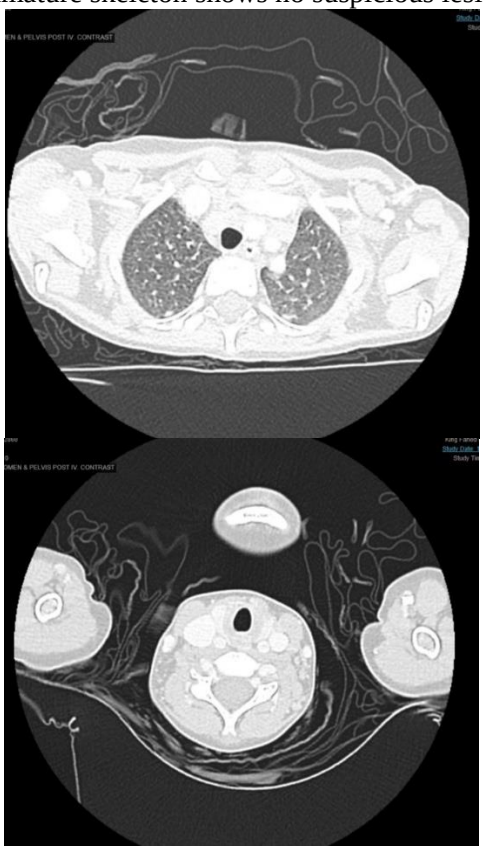
No focal hepatic or splenic lesions, portal hepatic veins are patent: gallbladder is present, no intra or extrahepatic biliary ducts dilatation. Adrenals and kidneys are intact Pancreas appears unremarkable the major abdominal and thoracic vasculatures are patent

There is a horizontal oriented blunting in the tubular structure with air foci within it, it is measure up to 0.58 cm, likely relating the fusiform appendix with no adjacent inflammatory signs or fat stranding. Otherwise, the unprepared large and small bowels appear grossly unremarkable

No peritoneal disease or ascites.

Pelvic organs are within normal limits.

The immature skeleton shows no suspicious lesions.



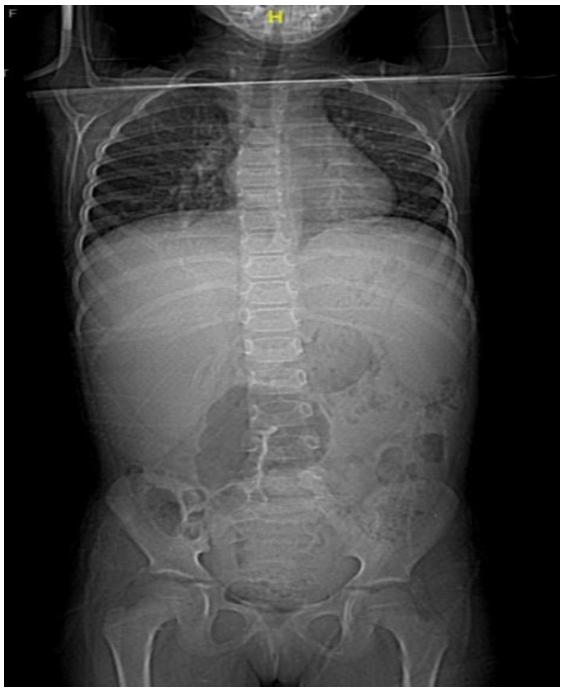


Figure (1): CT of the chest abdomen and pelvis.

Diagnosis: Sarcoidosis**.**

Treatment plan:

She received multiple courses of antibiotics but all the cultures were negative. Ophthalmological exam did not show any uveitis. She received I.V methylprednisolone pulse therapy for a single dose and was shifted to oral prednisolone. She received subcutaneous methotrexate at a dose of 7.5 mg once weekly.

Medications:

1. Prednisolone syrup - weaning doses weekly schedule provided to the family.
2. Methotrexate 7.5 mg once weekly subcutaneous injection.
3. Folic acid 1 mg PO daily except the day of methotrexate.
4. Vitamin D 1000 IU daily
5. Multivitamin Syrup 5 ml daily.

Follow up appointment after 4 weeks in OPD

Declaration of patient consent:

An approval of the study was obtained from Children's Specialized Hospital, King Fahad Medical City (Saudi Arabia) Academic and Ethical Committee.

The patient and her relatives were informed that the case would be taken as case report for publishing and they accepted. This work has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

DISCUSSION

Sarcoidosis is a chronic, granulomatous condition with unknown cause [(7)]. Diagnosis is based on tissue biopsies, demonstrating granulomas without caseous necrosis in the presence of clinical and radiological findings, consistent with sarcoidosis, where other causes which might be associated with a similar manifestation are ruled out [(8)].

Presence of granulomas alone is not diagnostic. From the histological perspective, same lesions may also be observed in chronic berylliosis, tuberculosis, histoplasmosis, coccidioidomycosis, lymphoma, Hodgkin's disease, bronchogenic carcinoma, foreign body granuloma, schistosomiasis, syphilis, and leprosy [(9,10)].

A considerable percentage of patients are diagnosed after mediastinal lymphadenopathy is incidentally discovered on chest radiography [(6)]. The most common clinical manifestations are pulmonary in nature and include dry cough, dyspnea, and chest pain. Constitutional symptoms, such as fever, unintentional weight loss, and fatigue, are also seen in about one-third of patients [(3)].

Although hepatic involvement is seen in the cases in sarcoidosis, clinical symptoms and impairment of the hepatic function are not common [(10, 11)]. Corticosteroids (e.g. prednisone) remain as the main therapeutic choice [(12)].

CONCLUSION

Sarcoidosis is a disease that mostly affects the lung, but sometimes it is outside the lung, as in our case. Doctors should pay attention to this so that the wrong diagnosis does not lead to complications or death of the patient.

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