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Case Report

Rosai-Dorfman disease mimicking gastrointestinal tuberculosis and fungal sinusitis: A case report [☆]

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ABSTRACT

Every organ can be affected by Rosai-Dorfman disease, the most common being skin and soft tissue, bone and upper respiratory tract. Here we present a Rosai-Dorfman disease patient who manifested with multiple organ involvement. A 33-year-old male presented with multiple joint pain, diarrhea, a purulent nasal discharge and sinus pain. After having serials of investigations he was misdiagnosed as having tuberculosis and fungal sinusitis. Finally, cytology from lymph node aspiration suggested Rosai-Dorfman disease. The sino nasal presentation of Rosai-Dorfman disease can be misdiagnosed as fungal sinusitis and the gastrointestinal manifestation can mimic gastrointestinal tuberculosis. Diffuse joint involvement could also be an initial manifestation of Rosai-Dorfman disease.

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Introduction

Sinus histiocytosis with massive lymphadenopathy (also known as Rosai-Dorfman disease (RDD) was initially described by Lampert and Lennert in 1961 and then characterized as a benign entity with marked, painless cervical lymphadenopathy with associated fever and weight loss by Rosai and Dorfman in 1969 [1]. However, every organ can be affected by RDD. Common extranodal sites of involvement include the skin, nasal cavity, bone, orbital tissue and the central nervous system. It commonly affects children and young adults [2]. The typical pathologic finding of RDD is an enlarged

lymph node with a low power appearance of extensive sinusoidal expansion [1].

RDD usually has a good prognosis, with spontaneous remission reported in up to 50% of cases [3]. Surgery may be indicated for single extranodal disease or for symptomatic airway, central nervous system or sinus disease. Those with multifocal extranodal disease may require systemic treatment [4].

In developing countries, RDD can mimic some infectious diseases and malignancies. Here, we present a patient who initially manifested with multiple joint pain, later on developed diarrhea and abdominal pain, sinus pain and a purulent nasal discharge.

Abbreviations: CT, computed tomography; CNS, central nervous system; GI TB, gastrointestinal tuberculosis; RDD, Rosai-Dorfman disease.

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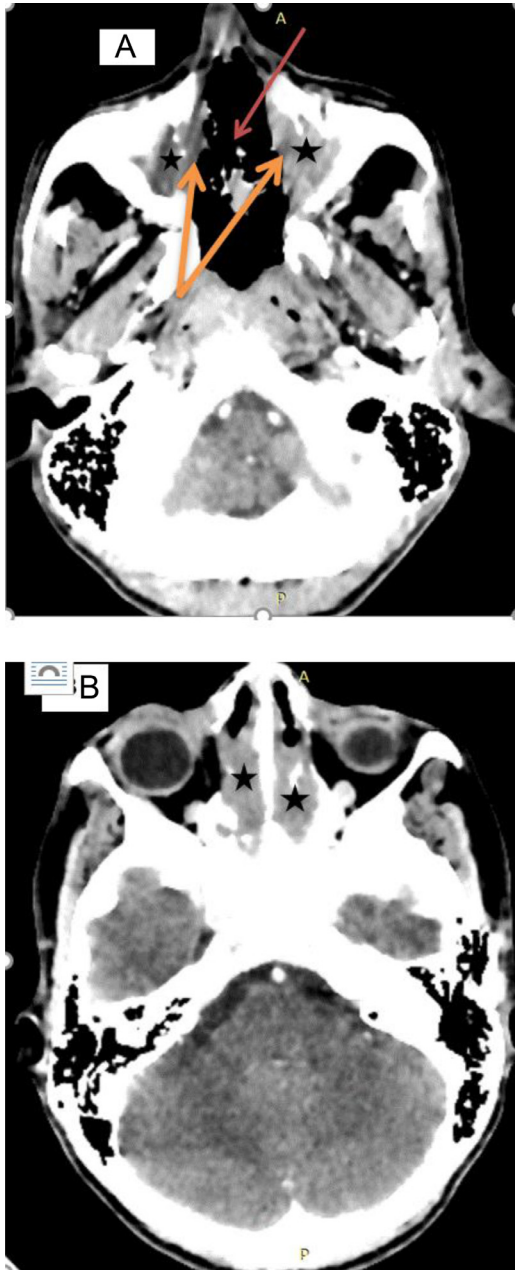


Fig. 1 – (A and B) Axial postcontrast PNS CT showing opacification of the bilateral maxillary and ethmoid sinuses with a soft tissue lesion (asterisk) with erosion of the maxillary sinus wall and nasal septum (arrows).

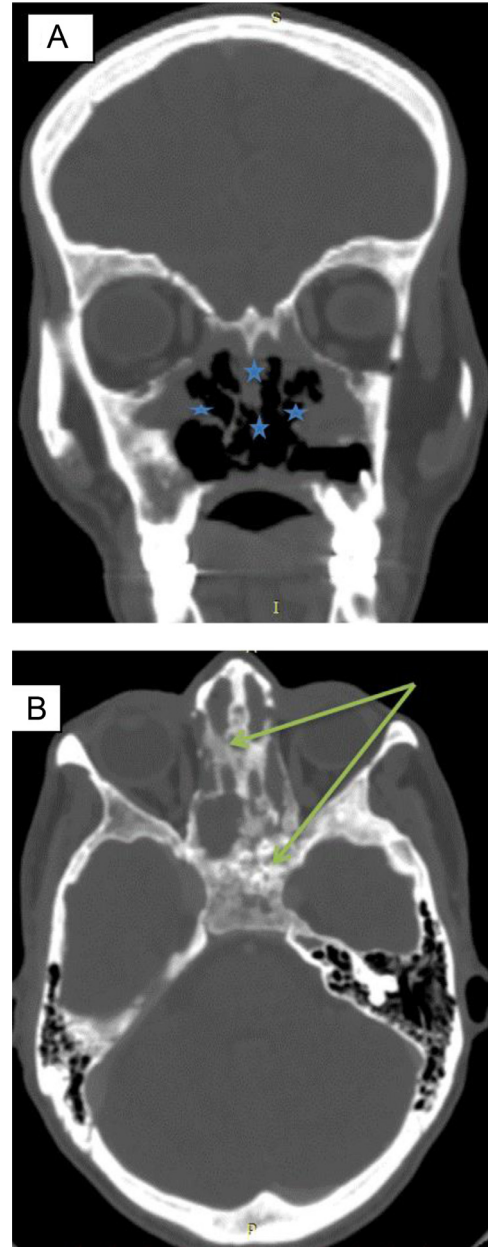


Fig. 2 – (A and B) CT scan of PNS coronal and axial section in bone window showing extensive destruction of bilateral medial wall of maxilla, ethmoid septa, nasal cavity with septal perforation (asterisk) and associated sclerotic changes (arrows).

Case presentation

We report a case of a 33-year-old male who initially presented with diffuse joint pain which started 8 years before his current presentation. The joint pain was associated with swelling of multiple joints and limited movement. For these complaints he visited several clinics and took non-steroidal anti-inflammatory drugs with short term improvement. A year before his last visit he developed diarrhea and abdom-

inal pain and was also diagnosed with gastrointestinal (GI TB) after an abdomino-pelvic ultrasound. He took anti tuberculosis (Anti TB) (2-month intensive phase with 4 drugs, isoniazid, rifampin, ethambutol, and pyrazinamide and 4 months continuation phase consists of 2 drugs (isoniazid and rifampin) administered for 4 months). He took the Anti TB with good adherence without showing any improvement. Few months before his last visit, he developed a purulent nasal discharge and facial pain, for which he was diagnosed with fungal sinusitis

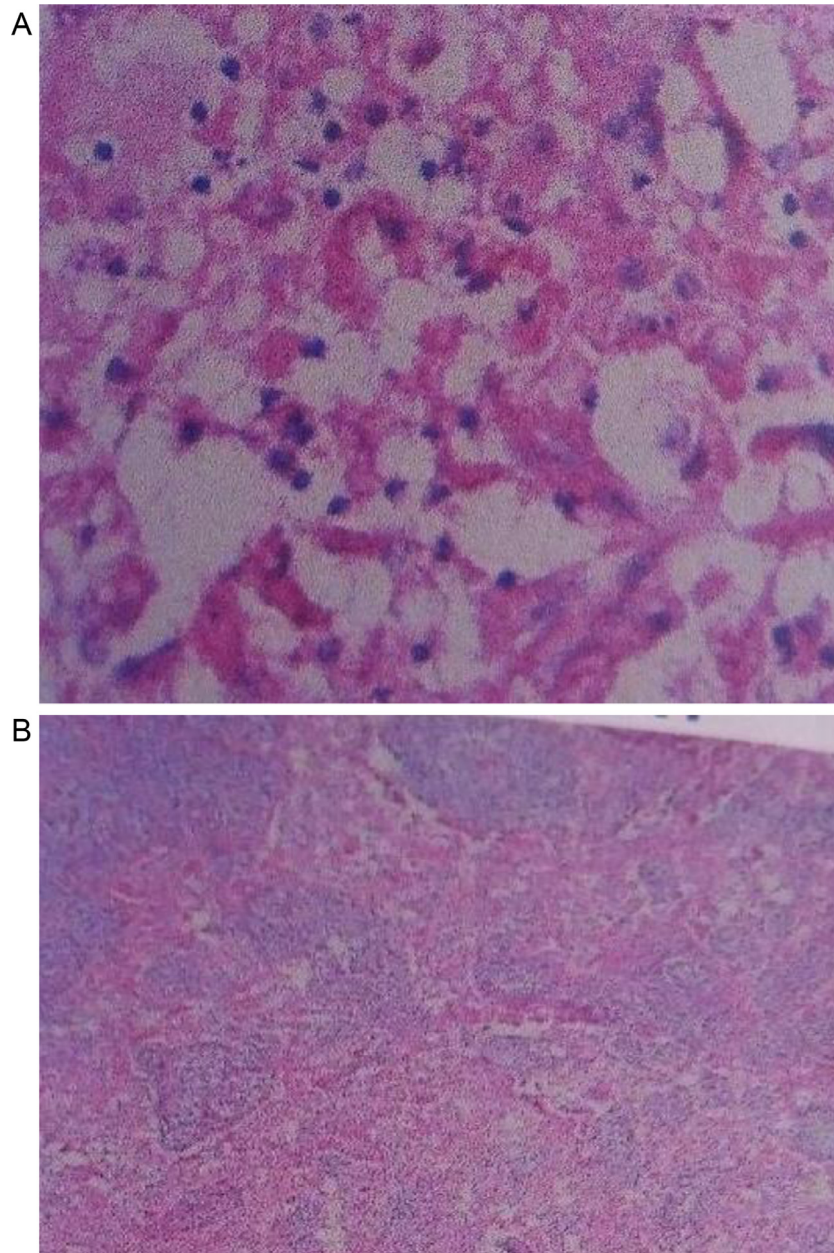


Fig. 3. (A and B) – Microscopic images (Hematoxylin and Eosin stain, magnification: A, 40x and B, 10x) sections show capsulated lymph nodes with partly effaced architecture having large infiltrates of histiocytes with emperipolesis. Dx—right cervical lymph node—consistent with Rosai-Dorfman syndrome.

after having a computed tomography (CT) scan and received anti-fungal. He had no family history of similar problem.

On examination, he had signs of anemia (pale conjunctivae, palmar pallor, third heart sound), multiple significantly enlarged lymph nodes in the neck, axilla and groin with no organ enlargement.

His blood examination revealed anemia with Hemoglobin of 6.6mg/dl (13-16.5) and leukocytosis of 12,000/ μ l (4000-1000) (88.4% (50%-70%) neutrophil) with normal platelet counts. Abdomino-pelvic ultrasound showed multiple intra-abdominal lymphadenopathies with no bowel matting. A CT scan of the head revealed areas of homogenous non-

enhancing soft tissue density lesions opacifying the whole paranasal sinuses; and associated extensive destruction of the nasal cavity with septal perforation (Fig. 1A and Figs. 2A and B). In addition, there were sclerotic bony changes involving the skull base. A cervical lymph node fine needle aspirate showed capsulated lymph nodes with partly effaced architecture having large infiltrates of histiocytes with emperipolesis consistent with RDD (Figs. 3A and B).

Finally, the histologic diagnosis of RDD was made. The patient was started on Prednisolone 0.5 mg /kg and a review was scheduled after 2 weeks. Unfortunately, the patient disappeared from follow-up. We finally heard from his wife that

there was marked improvement in the first week of his treatment. However, he developed abdominal pain and vomiting during the second week after starting prednisolone treatment and he died of unknown cause at his home.

Discussion and conclusion

This is the first RDD case reported from Ethiopia. Several case reports from other countries discussed various manifestations of this disease. Lymph node involvement is present in about 43% of cases with 75% of these being in the head and neck. Subsites of extra nodal involvement in the head and neck include 73% in the upper respiratory tract, 50% orbital, and 25% salivary glands [5]. However, initial joint manifestation, sinus involvement with nasal septum perforation were not previously described. Another interesting feature of this case is the significant diagnostic delay and treatment for other conditions such as nonspecific arthritis, fungal sinusitis and GI TB.

Mono articular involvement with RDD was reported [6,7]. However, diffuse joint involvement as initial manifestation was not described before. For the joint pain, he was taking different anti-inflammatory medications which may have hampered symptoms of RDD and contributed to the late presentation.

RDD involving the upper respiratory tract has been reported, typically with polypoid masses, mucosal thickening, or soft-tissue opacification of the nasal cavity or paranasal sinuses [8]. However, there was extensive destruction of the paranasal sinuses and nasal septum perforation in our case mimicking destructive sino nasal pathologies such as inflammatory granulomatous diseases (like fungal sinusitis, granulomatosis with polyangitis) and malignancies.

GI involvement with RDD was reported in few studies [9–11], but misdiagnosis and treatment as GI TB was not reported. Differentiating GI TB from RDD lymphadenopathy was the real challenge in an area highly endemic for TB, and this forced the treating clinician to start him on anti TB medicines. The nature of lymphadenopathy and additional findings may help to differentiate these 2 problems. Discreet, bulky homogeneously enhancing lymph nodes favor RDD, while small, matted lymph nodes favor tuberculosis. Luminal involvement can happen with both cases. While every segment of bowel can be involved with RDD, the most frequent site of GI TB is the ileocecal region.

While diagnostic delay of infections like TB is causing huge problems, the reverse is also common particularly in developing countries where diagnostic facilities for etiologic diagnosis are not adequate. This patient took a 6-month course of anti TB treatment and several months of anti-fungal medication without showing any improvement. Considering alternative

diagnoses are crucial and should be considered early when a patient fails to improve after taking anti-infective therapies.

The main limitation of this case report is the demise of the patient from unknown cause and lack of follow-up to document resolution of symptoms and lymphadenopathy.

The sino nasal presentation of RDD can mimic fungal sinusitis and the GI manifestation can mimic GI TB. Diffuse joint involvement could also be an initial manifestation of RDD.

Patient consent

Written informed consent was obtained from the patient's next of kin for publication of this case report and any accompanying images. Verbal and written consent was taken before his death.

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