CASE REPORT

Sinus Histiocytosis with Splenomegaly in Children-A Rare Case Report

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ABSTRACT

Sinus Histiocytosis/Rosai-Dorfman Disease (RDD) are benign, rare proliferative disorder caused by over production and accumulation of specific type of white blood cell (Phagocytic Histiocyte) in the lymph nodes of the body. Here, lymphadenopathy mostly painless and commonly found in the neck (cervical) but may occur in other areas of the body such as skin, lung, central nervous system, kidney (less than 5%). Predominantly it affects the young age group of children, adolescents or young adults. In spite of spontaneous remissions, treatment strategies can be different according to involvement and severity (RDD-Seldom life threatening disease).[1,2]

Key Words: Sinus Histiocytosis/RDD, Splenomegaly, Children.

Introduction

In 1969, Sinus Histiocytosis was first narrated by Rosai and Dorfman. It is commonly presented as massive, painless, bilateral, symmetric cervical lymphadenopathy with fever, leukocytosis, and elevated sedimentation rate. It is a rare inflammatory non-neoplastic process where differentiation of extra nodal RDD from soft tissue generally difficult without histo-pathological examination. Remaining as a poorly elucidated disease, it has nodal and also extra-nodal involvement with episodes of exacerbation and remissions.[3,4] It is more common in men than women and is significantly more common among white & blacks than Asian.[3,10] It is highly variable in its clinical presentation and response to treatment. Its treatment is poorly defined but the prognosis is usually favorable. Here we are reporting a rare, unusual clinical presentation of abdominal mass with splenomegaly diagnosed as RDD with histo-pathological correlation. Only a few such case of RDD have been reported in the literature, so there are no such report as this case we are going to report. We emphasize that clinicians and pathologists should always be aware of RDD in making a differential diagnosis of lymphadenopathy and splenomegaly.

Case Presentation

The case of RDD reported here is a 7 year 10 months old female child, first issue of non-consanguineous parents with occasional fever for 2.5 months which was high grade, intermittent in nature (highest recorded 103°F) with no association with chills and rigor, subsided by antipyretics. She also complaints of progressive pallor and abdominal mass for same duration. She had no history of any bleeding manifestation, convulsion, respiratory distress, and contact with TB patient. With these above complaints she had visited several registered physicians and taken oral drug medications. Then she admitted on Anwer Khan Modern Medical

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Date of submission: 26.02.2019 Date of acceptance: 23.03.2019

AKMMC J 2019; 10(2) : 176-178

College Hospital (AKMMCH) on 28th may 2018. At the time of admission she was severely pale, (Hb-5.1 gm/dl), dyspnoic, febrile (T- 102°F), no regional lymphadenopathy, BCG mark present, no bony tenderness but huge splenomegaly was present (spleen is palpable which is 10 centimeter, hard in consistency, non-tender). Patient was treated with 2 units PRBC, IV antibiotics. But suddenly patient took DORB without any significant improvement before doing any investigations. Then, after 1 month 6 days, she re-admitted here on AKMMCH with the same complaints and she was ill-looking, febrile (T-100°F), moderately pale, no bleeding manifestation. On general examination, pulse-88/minute, BP 110/70 mm Hg, mildly pale , no cyanosis, no jaundice, lymph node is not palpable. On systemic examination of gastrointestinal system, abdomen is soft, liver was just palpable, spleen was palpable which is 12 centimeter, hard in consistency, non-tender. Then series of investigations revealed persistent anemia even after blood transfusion on Complete Blood Count (Hb:5.1>9.1>10 mg/dl) ESR 85 in 1st hour, Myeloid Hyperplasia on bone marrow. S. Ferritin 1439 ng/ml and TIBC 193 microgram/l, S. LDH-300 U/L, CRP-202.1 mg/L, PT-13.2 Sec, APTT-27.2 Sec, on Chest X-ray suggestive of right paratracheal and right hilar-lymhadenopathy, on USG of Whole Abdomen hepato-splenomegaly. CT Chest and Whole Abdomen reveals hilar & mediastinal lymhadenopathy, massive abdominal lymphadenopathy, moderate to severe splenomegaly with multiple lesions, mild hepatomegaly, suggesting lymphoma, USG guided FNAC from abdominal Lymph Node found- No malignant cell identified. Then an excisional lymph node biopsy was done and histopathology revealed that it's a case of Sinus Histiocytosis.

Before diagnosis, she was treated by infusion and transfusion to ensure symptomatic treatment and antibiotics were also given (Ceftriaxone, Meropenem) as per the clinician's judgment. After availability of report she was supposed to receive appropriate treatment with the protocol available for Sinus Histiocytosis but parents wanted to continue treatment in abroad. So, patient discharged without any particular treatment for the disease.

Discussion

In our patient the most significant clinical finding was splenomegaly and lymphadenopathy in hillar region and mediastinal region and also in abdomen. Though cervical nodes are most commonly involved but axillary, mediastinal, and inguinal nodes involvement are also found in association with RDD. It has been noted that the disease is a reactive rather than a neoplastic disesase. The final diagnosis of RDD is made based on clinical and histological findings and histiocytes positive with massive lymphadenopathy. Until now, no>1000 RDD cases have been reported in English journals. It is often accumulated in extranodal sites including the orbit, eyelid, skin, bone, central nervous system, and soft tissues. However, simple soft tissue manifestation of RDD (without lymphadenopathy or other systemic symptoms) is rarely seen, which occurs in <3% of patient. RDD is primarily found in trunk and proximal extremities as a rapidly evolutional entity. On occasion, it manifests as a multifocal and persistent disease. Although RDD is slightly more common in men but RDD has a female sex predominance of nearly 3:1. To our knowledge, this is the first case report of RDD presented with splenomegaly in children.[5, 7, 8]

Laboratory tests and radiograph results were unremarkable. Noguchi et al reported that patients of RDD might show slight elevation of CRP and ESR. However, such results were also observed in our case(very high ESR and CRP). Laboratory parameters may show nonspecific increase in RDD, which was reported by a previous literature.[9]

The diagnosis of RDD is mainly confirmed by pathological examinations. Specimens are mainly obtained by open surgical biopsy or fine needle aspiration. In general, histopathological inspection markedly shows a large number of mixed cell population, including mature plasma cells and lymphocytes. The most typical cells are histiocytes of accentuated phagocytic appearance. The most useful markers of histiocytes in RDD are positive for S-100 protein and CD68, and negative for CD1a

Differential diagnosis of RDD includes histiocytosis of Langerhans cells, histiocytic sarcoma, lysosomal

storage diseases (eg, Gaucher disease), classical Hodgkin lymphoma, melanoma and metastatic carcinomas, and infections caused by Histoplasma and mycobacteria involving lymph nodes.

Owing to its low incidence, no ideal or standard treatment has been defined for RDD. The predilection sites of the lesion and its self-limiting nature also make the majority of RDD patients not necessary to be intervened. Nevertheless, the course of RDD is still unpredictable. When vital organs are involved, interventions proposed by previous literatures include corticosteroids administration, chemotherapy, radiotherapy, and surgical resection, but their efficacy remains uncertain. In our case, the girl and her family had refused our treatment after the surgery and no recurrence had been observed.

Conclusion

Managing a case of RDD is a challenge specially when complicated. We managed this patient with close observation and continuous risk benefit assessments of management decisions. Unfortunately, we could not complete the treatment in spite of our diagnosis but we tried to manage the patient as per as her parents approved. However, experience with one patient cannot be generalized to others. Therefore, it is important that physicians write their difficulty and experiences in managing such complicated patients.

Conflict of interest: None.

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