Journal of Medical Practice and Review 6:5 [2022]
Everant.in/index.php/jmpr

Journal of Medical Practice and Review
Available online at http://jmpr.info/index.php/jmpr/index

Rosai- Dorfman Disease four-year follow-up: Case Report ekly voot*

Adana Numune research and training hospital department of hematology Yüregir/Adana Turkey

ARTICLE INFO

Corresponding Author: ekly voo*

Keywords:- Rosai- Dorfman Disease four-year follow-up: Case Report

ABSTRACT

Rosai Dorfman disease (RDD) is rare histiocytic disease with unknown etiology that typically presents with painless cervical lymphadenopathy. The disease is also known as sinus histiocystosis with massive lymphadenopathy (SHML). The normal functioning of lymph glands is disturbed due to significant enlargement of the lymphatic sinuses. Sinus hystiocytes contain cytoplasmic fat and strongly positive S100 protein CD68 and CD14 can be positive. Forty percent of patients have extra nodal involvement. The prognosis of the disease is usually good. However, if accompanied by immunologic abnormalities young age, extra nodal involvement (especially in the kidney or, liver), and the prognosis is poor. Regarding treatment, the disease often has spontaneous remission; therefore, observation is essential. No treatment modalities are superior to over another. In this article, we present a male patient who was diagnosed as having extra nodal RDD with skin and soft tissue involvement, who was followed up for four years.

Introduction:

Rosai–Dorfman disease (RDD) is rare histiocytic disease with unknown etiology that typically presents with painless cervical lymphadenopathy. The disease is also known as sinus histiocystosis with massive lymphadenopathy (SHML). (1) It can be seen at any age, though it is common in adolescence. Massive painless lymphadenopathy, fever, leukocytosis, and increased erythrocyte sedimentation rate are characterized by polyclonal hypergammaglobulinemia. Due to significant enlargement of the lymph sinuses, the normal regeneration of the lymph gland is disrupted. The sinuses contain large amounts of lymphocytes, plasma cells, and neutral lipids with loaded, wide, clear cytoplasms, and large vesicular nuclei in hystiocytes. Many cytoplasms of the hystiocytes contain a large number of intact lymphocytes, red blood cells, and a small amount of plasma cells. This is defined as 'emperipolesis' or 'lymphocytophagocytosis' and is considered diagnostic for SHML (2, 3). Sinus hystiocytes include cytoplasmic fat and strongly positive S100 protein. CD68 and CD14 can be positive and separated from Langerhans cells by being CD1a negative (4). Progress is almost benign but may be fatal.Forty percent of patients have extranodal involvement, sometimes without lymphadenopathy. This mostly affects the eyes, head-neck region, upper respiratory track, skin-subcutaneous tissue, and the central nervous system (5, 6, and 7). In this article, we present a male patient who had extranodal Rosai–Dorfman disease with common subcutaneous soft tissue involvement, who had been under follow up for 4 years.

Case:

A 24-year-old male patient who had type 1 diabetes mellitus (DM) presented to the polyclinic in December 2013 for common subcutaneous nodules. In his physical examination, subcutaneous painless nodules similar to lipomas were palpated.

There were no symptoms such as weight loss, loss of appetite, night sweats, or fever. His erythrocyte sedimentation rate was 110 mm/h, immunoglobulin IgG 2491 mg/dL (normal, 650-1600 mg/dL), IgA 637 mg/dL (normal, 40-350 mg/dL), IgM 72 mg/dL (normal, 500-300 mg/dL). The total blood count was normal. Computed tomography (CT) showed multiple lymphadenopathies (LAP), the largest was 12 mm in the cervical chain and 20 mm in the bilateral axillary regions. There were no LAP in other regions. LAP biopsies from the cervical and axillary regions were reported as reactive lymph node hyperplasia. A biopsy from the subcutaneous soft tissue was reported as Rosai–Dorfman's disease.

Immunohistochemically, S100 and CD68 were positive. HHV8 (-) was negative. Positron emission tomography (PET CT) showed no malignant involvement.

At superficial ultrasound (US) after 3 years, in addition to neck and axillary LAPs, multiple new LAPs were found; the largest was 37 x 9.5 mm. Reactive characteristics were detected bilaterally in the inguinal region. The patient was observed without treatment for 4 years.

No progression of the cervical, axillary, and inguinal LAPs was seen in CT with repeated imaging, but there was a little subcutaneous proliferation. IgG and IgA maintained as stable (in sequence 2382 mg/dL). The patient’s clinical condition did not worsen. We continue to observe our patient without treatment at 6-month intervals.

Discussion:

Rosai–Dorfman (RDD), massive painless lymphadenopathy, fever, leukocytosis, and increased erythrocyte sedimentation rate are characterized by polyclonal hypergammaglobulinemia. Extra nodal involvement occurs in 40% of patients and can sometimes be present without lymphadenopathy Our patient had extra nodal involvement, high ESR, polyclonal hypogammaglobinemia, lack of fever, and leukocytosis.

The etiology of the disease is unknown. The illness can occur with conditions such as polyarthralgia, and type 1 diabetes mellitus (8). Our patient had type 1 DM, which supports this view. The prognosis of the disease is generally good; however, concomitant immunologic abnormalities, young age, extra nodal involvement (especially in the kidney or liver) are poor prognostic criteria (9). In our case, the disease had benign progression for 4 years along with poor prognostic criteria such as young age, accompanying immunologic abnormality (type I DM), extranodal involvement.

In the treatment of the disease, observation is essential because of frequent spontaneous remission. No treatment modalities have any superiority. Antibiotics, acyclovir, steroids, radiotherapy, immunosuppressive drugs

(methotrexate,azathioprine,cyclosporineA,mycophenolate,mofetil,cyclophosphamide,infliximab and rituximab), and chemotherapy (vinblastine,6-mercaptorine,vincristine,cladribine,clofarabine,chlorambucil, and etoposide) have been used in the treatment of RDD (1, 10).

We observed our patient for 48 months without performing any treatment. There was no progression in that period.

Conclusion:

Rosai–Dorfman's disease is rare. However, painful cervical LAP, fever, and leukocytosis are common in the literature, like in our case, as well as extranodal involvement and atypical clinical presentation. If a biopsy from LAP provides no diagnosis but suspicion persists, a biopsy should be taken from other tissues. There is no standardization with regards treatment and follow-up. Physicians should determine treatment of patients in view of the clinical progression period.

References:

1. Mosheimer B, Oppl B, Zandieh S, Fillitz M, Keil F, Klaushofer K, and Weiss G, Zwerina J. Bone involvement in Rosai-Dorfman disease (RDD): A case report and systematic literature review. Curr Rheumatol Rep (2017) 19:29:1-5.
2. Dalia S, Sagatys E, Sokol L, Kubal T. Rosai-Dorfman disease: tumor biology, clinical features, pathology, and treatment. Cancer Control. 2014; 21(4):322-7.

3. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. Arch Pathol. 1969(1):63-70.
4. O'Malley DP, Duong A, Barry TS, Chen S, Hibbard MK, Ferry JA, et al. Co-occurrence of Langerhans cell histiocystosis and Rosai-Dorfman disease: possible relationship of two histiocytic disorders in rare cases. Mod Pathol. 2010,23(12):1616-23.
5. Juskevicious R, Finley JL. Rosai-Dorfman disease of parotid gland: cytologic and histopathologic findings with immunohistochemical correlation. Arch Pathol Lab Med 2001; 125(10):1348-50.
6. Foucar E, Rosai J, Dorfman R. Sinus histiocystosis with massive lymphadenopathy (Rosai-Dorfman disease): review of the entity. Semin Diagn Pathol.1970 (1):19-73.
7. Rodriguez-Galindo C, Helton KJ, Sanchez ND, Rieman M, Jeng M, Wang W. Extranodal Rosai-Dorfman desease in children. J Pediatr Hematol Oncol. 2004;26(1):19-24.
8. Cocker RS, Kang J, Kahn LB. Report of a case presenting as a midline thyroid mass. Arch Pathol Lab.2003; 127:197-200.
9. Unal OF, Koybasi S, Kaya S. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). Int J Pediatr Otorhinolaryngol.1998:44; 173-6.
10. Lu CI, Kuo TT, Wong WR, Hong HS. Clinical and histopathologic spectrum of cutaneous Rosai-Dorfman desease in Taiwan. J Am Acad Dermatol.2004 Dec; 51(6):931-9.