A Case Report: Rosai Dorfman Disease

*Yasmin F¹, Al-Amin ANM², Begum F³, Ghosh AK⁴, Kabir MRZ⁵, Begum M⁶

Abstract

Rosai-Dorfman Disease (RDD) is a rare disorder which usually associated with enlargement various superficial or deep lymph nodes. Clinical course of this disease is prolonged with exacerbation and remission phases occasionally. Over-production and accumulation of a specific type of white blood cell called histiocyte occurred in the lymph nodes of the body, mostly on the the neck. Other lymph node groups and in some cases, extra nodal abnormal accumulation of histiocytes may occur in other areas of the body that is more common in the head and neck region. Here we discussed a patient with extra-nodal involvement who presented with bilateral orbital mass.

Key words: Rosai Dorfman Disease, Sinus histiocytosis, massive lymphadenopathy.

Introduction

Rosai-Dorfman Disease (RDD) also known as Sinus Histiocytosis With Massive Lymphadenopathy (SHML) is a rare and benign histiocytic disease.¹ This disease first described by Destombes in 1965 later on by Rosai and Dorfman in 1969.² RDD usually develops in first and second decades of life. This disease is microscopically characterized by benign histiocytic proliferation with frequent lympho-phagocytosis (emperipolesis).³ A chronic massive enlargement of cervical lymph nodes occurred in this disorder which is accompanied by fever, leucocytosis and other lymphatic groups, such as mediastinal, axillary and inguinal lymph nodes.^{4,5,6}

Extra nodal involvement also occurred in skin, orbit, eyelid, liver, spleen, testis, CNS, salivary glands, bones, respiratory tract.^{7,8} The cause of the disease is remains unknown, but an altered immune responses and infectious agents may play a role. SHML is a self-limited requiring only observation and seldom life-threatening disease which commonly does not require therapy.⁹ In some cases, the condition can be severe. Several treatments have been used with varying efficiency.

Case Report

A 17 years old boy of a non-consanguineous parents attended pediatric hematology & oncology department first time with the complaints of swelling of the left eye and eye lids for 10 years, loss of vision of left eye for 5 years and swelling of the right eye for 6 years. Gradually the swelling was increased in size, developed nasal voice and snoring. Swelling of the left eye-lid first developed when he was at 10 years of his age, excisional biopsy done from left eye and histopathological report revealed Rosai-Dorfman disease. After this he remained well for about 2 years. Then again pain and subconjunctival hemorrhage developed in left eye with gradual loss of vision. At that time, he also developed swelling of the right eye which gradually increasing day by day but he did not take any treatment.

On examination, the patient was afebrile and in a good general condition. His weight was of 40 Kg. He had a pulse rate of 90/min, blood pressure of 120/80 mmHg, respiratory rate of 18/min, and mildly pale. Multiple

Author's

1. *Farida Yasmin, Department of Paediatric Haematology and Oncology, National Institute of Cancer Research and Hospital, Mohakhali, Dhaka, Bangladesh.
2. Abu Naser Mohammad Al-Amin, Department of Paediatric Haematology and Oncology, National Institute of Cancer Research and Hospital, Mohakhali, Dhaka, Bangladesh.
3. Ferdousi Begum, Department of Paediatric Haematology and Oncology, National Institute of Cancer Research and Hospital, Mohakhali, Dhaka, Bangladesh.
4. Ashis kumar Ghosh, Department of Paediatric Haematology and Oncology, National Institute of Cancer Research and Hospital, Mohakhali, Dhaka, Bangladesh.
5. M. Rashed Z. Kabir, Department of Paediatric Haematology and Oncology, National Institute of Cancer Research and Hospital, Mohakhali, Dhaka, Bangladesh.
6. Mamta Begum, Department of Paediatric Haematology and Oncology, National Institute of Cancer Research and Hospital, Mohakhali, Dhaka, Bangladesh.

Correspondence: Farida Yasmin, Department of Paediatric Haematology and Oncology, National Institute of Cancer Research and Hospital, Mohakhali, Dhaka, Bangladesh,
e-mail: faridayasmin71@yahoo.com, Contact: +880 1719-672785.

nodular swelling over both upper and lower eye lids, the larger one measuring about 5 cm x 3.5 cm, mobile, firm in consistency, non-tender. There was proptosis of left eye with ulceration of sclera and congestion of conjunctiva. Right upper eye covers part of the sclera. Vision in right eye was - 6/9, left eye loss of vision. There was no lymphadenopathy. Cardiac and respiratory examinations were normal. His abdomen was soft and there was no hepatomegaly or splenomegaly. Musculoskeletal and neurological examinations were also normal.

CT Scan of paranasal sinuses, orbits and brain showed huge soft tissue mass occupying bilateral nasal fossa, all paranasal sinuses with extension to bilateral facial muscles, soft palate, left infratemporal fossa, left orbit with involvement of orbital contents with bony erosion suggestive of neoplasm.

Histopathological report showed a benign lesion composed of polymorphous cellular elements. These are consisting of numerous lymphoid cells, histiocytes, plasma cells and few eosinophils. Most of the macrophage are highly reactive containing abundant cytoplasm. Some of these present small number of lymphocytes within the cytoplasm (lymph phagocytosis). Features are compatible with Rosai-Dorfman Disease (extra nodal type). He was treated by oral prednisolone and inj. Vinblastine 6mg/m².

Discussion

Rosai-Dorfman disease (RD) is a rare histiocytic disorder which involves the over-production of a type of white blood cell called non-Langerhans sinus histiocyte. These cells are accumulated most often in the lymph nodes, but may occur in other areas of the body which lead to organ damage. The exact reason of these cells over-production is unknown, but many

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possibilities have been considered such as viral bacterial infection, autoimmunity, environmental and genetic causes may also consider.

The symptoms and on examination findings associated with Rosai-Dorfman disease vary person to person greatly depending on the extent of the disorder and the specific organ systems affected. Some cases may only affect the lymph nodes and may not cause any serious complications. Less often, some cases may affect various organ systems of the body and may potentially cause serious complications. Any organ of the body can be affected and extra nodal involvement also common. In some cases, the lymph nodes may not be affected, instead of a specific area of the body such as the skin, a solitary bone, orbit, or the central nervous system may be affected. In a case series study, it was shown that out of 13 patients 11 patients had infiltrate in the orbital soft tissues, and five of these patients also had eyelid disease and one patient had infiltrates only within the eyelid, and one without disease in the orbit or eyelid had extensive infiltrates in the uveal tract.¹⁰

There is no specific treatment for the Rosai-Dorfman syndrome but in many cases, spontaneous remission occurred without treatment within in months or a few years. But whenever possible clinical observation is preferred for individuals with Rosai-Dorfman disease without treatment.

But when the condition is organ threatening or life threatening then treatment is required for those patients. Various treatment options may become necessary for the patient of Rosai-Dorfman disease which is the management of the specific symptoms that are apparent in each individual. Several different treatment options include therapy with certain drugs including steroids (e.g., prednisone), chemotherapy with a combination of vinca alkaloids and alkylating agents, alfa-interferon, and radiation therapy and surgery. In some cases, affected individuals have shown improvement of symptoms with these treatments.¹¹

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