Case Report

A Report of Rosai-Dorfman Syndrome in a 6-year-old

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Abstract

Rosai–Dorfman disease is a rare subtype of non-Langerhans cell histiocytosis with very little data recorded in the paediatric surgical literature. This condition is benign and usually self-limiting but can sometimes be misdiagnosed as lymphoma. While this disease has been historically considered a benign entity, a subset of patients endures an aggressive course necessitating the use of systemic therapies. We describe the clinical management of a 6-year-old male who presents with a long-standing history of progressively increasing posterior neck swelling.

Keywords: Lymphoma, lymphadenopathy, Rosai–Dorfman

Introduction

Rosai–Dorfman disease (RDD) also known as sinus histiocytosis is a benign systemic histioproliferative disease. Patients with RDD usually present with fever, leucocytosis, weight loss, night sweats and lymphadenopathy usually affecting the cervical lymph nodes.[1,2] It is common in children and young adults. RDD is a differential for cervical lymphadenopathy. It can occur in other sites of the body as some case reports have described lesions in extranodal sites including the skin, soft tissues, upper airways, bones and brain. There have also been reported cases of spinal involvement. The disease is usually self-limiting. However, in severe disease and atypical presentations, various treatment modalities may become necessary. Surgical excision is the main treatment of choice if treatment becomes necessary. Histopathological examination showed pericapsular fibrosis with dilated sinuses, large histiocytes with intact phagocytized lymphocytes heavily infiltrated with lymphocytes and plasma cells in the background. Due to the common presentation of cervical lymphadenopathy, the disease is initially diagnosed as lymphomas until histological report excludes otherwise.

Case Report

A 6-year-old boy presented to the paediatric surgical clinic with a 1-year history of progressively increasing posterior neck swelling. He had been previously treated with antibiotics at several peripheral facilities with no response. On further enquiry, he had no other systemic symptoms. Physical examination showed a posterior triangle swelling which was firm in consistency, freely mobile, nontender and not attached to any underlying muscles or skin. The lesion measured about 2 cm × 4 cm [Figure 1]. All other systems were normal. Laboratory investigations revealed normal haemoglobin levels and white blood count. An ultrasound scan of the neck showed an enlarged lymph node measuring 3.2 cm × 1.5 cm with multiple other right-sided lymph nodes with no necroses. A diagnosis of reactive lymphadenopathy to rule out lymphoma was made.

Based on the suspicion of lymphoma, an excisional biopsy of the right cervical lymph node was done. His post-operative period was uneventful. Histopathology revealed partial architectural distortion due to expanded parafollicular sinuses encroaching the follicles. Sinuses were filled with a prominent population of histiocytes with emperipolesis and admixture of plasma cells. These features are in keeping with RDD. Further immunohistochemistry was positive for S-100 confirming the diagnosis of RDD [Figure 2].

The only treatment offered was an excisional biopsy of the mass. He has been regular at follow-up clinic for the past 6 months post-excision biopsy and is currently stable [Figure 3].

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Figure 1: The Posterior Cervical Swelling lesion measured about 2cm x 4cm (black arrow)

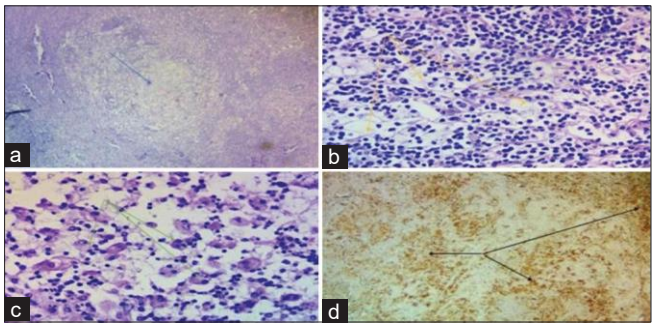


Figure 2: (a) Lower pole (×40) with a large collection of foam cells (blue arrow), (b) medium power (×100) of foam cells expanding nodal sinuses (yellow arrow), (c) medium power (×100) of lymphocytes engulfed by macrophages (green arrow), (d) low power (×40) of S-100 stained foamy histiocytes (black arrow)

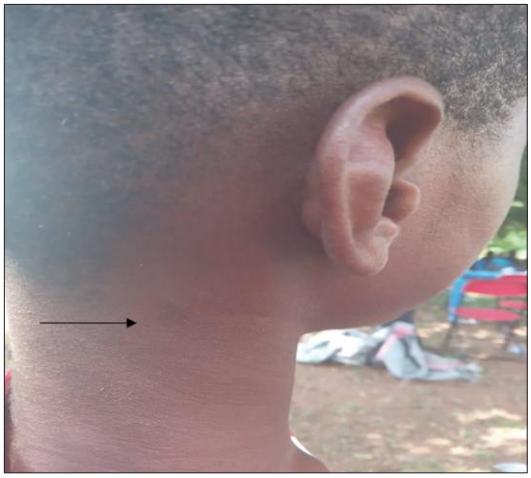


Figure 3: Post-operative picture (black arrow)

Discussion

In our case, the child was 6 years old which is consistent with the age of presentation in many published literature. RDD is a rare disease with an unknown aetiology. The condition was first described by Rosai and Dorfman in 1969.[1] Various infectious agents and viruses have been postulated to cause this disease but there is no concluding evidence in the literature.[2]

Our case is one of the few documented cases seen in Ghana, however, in a recent study, an uncharacteristically high incidence of RDD was seen in such a short period with five cases that occurred at the same institution within a 12-month period which proposes the possibility of a novel environmental predisposing factor.[3]

The disease is characterised by the accumulation of activated histiocytes in the sinusoids of lymph nodes and extranodal sites.[4] Extranodal disease occurs in a significant proportion of patients.[5] In most cases of lymphadenopathy, reactive lymphadenitis constituted the majority of the diagnoses, followed by tuberculosis, acute suppurative, Bacillus Calmette–Guerin-induced lymphadenitis, Kimura disease, RDD, and Kikuchi–Fujimoto disease.[6] Patients typically present with painless asymmetrical cervical lymphadenopathy similar to the presentation in this case report. The only other case report on RDD in Ghana occurred in a 12-year-old adolescent with the patient presenting with painful cervical lymphadenopathy.[4]

Cervical lymphadenopathy may sometimes be symmetrical and associated with other features of inflammation such as fever, leucocytosis and elevated sedimentation rate. In our case, there was no associated fever, cervical lymphadenopathy was unilateral, and there was no associated anaemia, elevated erythrocyte sedimentation rate, leucocytosis or hyperglobulinaemia.

The disease commonly affects lymph nodes, however, any organ of the body may be involved. There have been reported cases of RDD affecting extranodal sites such as the skin, central nervous system, upper respiratory system, long bones and soft tissue.[7,8]

Histological findings show the typical Rosai–Dorfman cells which are characteristically large, pale and histiocytic cells. These cells exhibit cytophagocytosis. Confirmatory immunohistochemistry shows histiocytes which are positive for S-100 protein and CD68, but stain negatively for CD1a.[9]

RDD is significantly more common amongst Caucasians and blacks than amongst Asians.[10] The disease can involve more than one site at the same time in the same patient. In the Indian subcontinent, two cases have been reported with extranodal, sinonasal mucosal involvement and cervical lymphadenopathy.[11]

A number of diseases can mimic RDD both clinically and histologically. These include infectious lymphadenitis, reactive lymph node hyperplasia, histiocytosis of Langerhans cells, histiocytic sarcoma, lysosomal storage diseases (e.g., Gaucher disease), classical Hodgkin lymphoma, melanoma and metastatic carcinomas and infections caused by Histoplasma and mycobacteria involving lymph nodes.[4,12]

It is important to also note that in Hodgkin and non-Hodgkin lymphomas, the malignancy and RDD can either precede or follow each other and occur in the same node.[13]

Fine-needle aspiration cytology (FNAC) coupled by histopathology has been described in helping make a diagnosis. In this case, excision biopsy alone was done without prior FNAC. It is important to note that FNAC is a reliable, minimally invasive, cost-effective, efficient and sensitive means to establish a conclusive diagnosis of RDD.[14] Immunohistochemistry is helpful in differentiating the different conditions.

Rosai–Dorfman is not very common, therefore, standardised management protocols are not yet fully available. This is also in part because the majority of patients with this condition have a self-limiting course, however, in most reports, an excisional biopsy was done just as in this case. When vital organs are involved, interventions proposed by previous literature include corticosteroid administration, chemotherapy, radiotherapy and surgical resection, but their efficacy remains uncertain.[15]

Conclusion

RDD is a rare occurrence and can sometimes be a concern to parents and guardians. The progressive and long history can be a diagnostic dilemma. This case report aims at creating knowledge of this rare condition and creates awareness of RDD in making a differential diagnosis of cervical lymphadenopathy. When in doubt, a lymph node biopsy with immunohistochemistry is helpful in establishing a diagnosis and ruling out sinister diseases such as lymphoma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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