Rosai-Dorfman disease is a rare disorder of systemic histioproliferation, first described in 1969 by Juan Rosai and Ronald Dorfman. It is characterised by massive lymphadenopathy, particularly of the head and neck region, and it is sometimes associated with extranodal involvement. This is the first case to be reported from our centre. We report a case of Rosai-Dorfman disease in a 12-year-old Nigerian boy who presented at the Children's outpatient clinic of the Federal Medical Centre, Owerri, Imo State, Nigeria, with a 5-year history of bilateral neck swelling, a 2-year history of groin swelling and a recurrent fever of over a year's duration. An initial diagnosis of Hodgkin's lymphoma was made. An excisional biopsy of the cervical lymph node was undertaken and the histology showed massive histiocytosis with emperipolesis, which confirmed the diagnosis of Rosai-Dorfman disease. Rosai-Dorfman disease, although a rare clinical entity, should be suspected in patients presenting with massive lymphadenopathy located especially in the neck and groin regions.

**Case presentation**

A 12-year-old Nigerian male presented at the children's outpatient clinic of the Federal Medical Centre, Owerri, Nigeria, with a 5-year history of bilateral neck swelling, 2-year history of groin swelling and a recurrent fever of over a year's duration. An initial diagnosis of Hodgkin's lymphoma was made. An excisional biopsy of the cervical lymph node was undertaken and the histology showed massive histiocytosis with emperipolesis. A histological diagnosis of RDD was made. The laboratory investigations showed a blood cell count of 13.8 x 10⁹ L⁻¹, neutrophils 70%, lymphocytes 30%, platelet count 188 x 10⁹ L⁻¹, left shift of neutrophils, normocytic normochromic red blood cells and elevated erythrocyte sedimentation rate (ESR) (100 mm/hr). Haemoglobin was 9.8 g/dL, HIV I and II screenings were negative and haemoglobin electrophoresis had a normal AA pattern. A Mantoux test was non-reactive (0 mm). A chest X-ray showed no evidence of enlarged lymph nodes and no focal lung lesions were seen. Blood films for detecting malaria parasites showed a ring form of Plasmodium falciparum. An abdominal ultrasound scan, random blood glucose tests, urinalysis and direct Coombs tests were all normal. The patient was commenced on oral antibiotics (cefuroxime) and antimalarial therapy (Coartem) based on local sensitivity patterns and malarial tests, pending the result of the excision biopsy of the lymph node.

The histology report showed a benign, non-neoplastic lesion characterised by expanded sinuses that were filled with enlarged lymph nodes and no focal lung lesions were seen. Blood films for detecting malaria parasites showed a ring form of Plasmodium falciparum. An abdominal ultrasound scan, random blood glucose tests, urinalysis and direct Coombs tests were all normal. The patient was commenced on oral antibiotics (cefuroxime) and antimalarial therapy (Coartem) based on local sensitivity patterns and malarial tests, pending the result of the excision biopsy of the lymph node.

The histology report showed a benign, non-neoplastic lesion characterised by expanded sinuses that were filled with enlarged histiocytes containing abundant cytoplasm and plasma cells. Within some of the enlarged histiocytes were intact lymphocytes (emperipolesis). A histological diagnosis of RDD was made. The parents were counselled on the benign nature of the disease and were scheduled for monthly follow-up visits. The size of the cervical lymph node was observed to be regressing by the third monthly visit. However, the parents defaulted on further follow-up visits despite numerous telephonic requests.

**Discussion**

The term RDD was first introduced into scientific literature in 1969 when Juan Rosai and Ronald Dorfman described four cases of a rare, idiopathic, non-neoplastic lymphoproliferative disorder. They described its clinicopathological features in 1972. Typically, it is characterised by bilateral cervical lymphadenopathy with fever, leukocytosis, elevated ESR and hypergammaglobulinaemia. Our
patient had all the typical features of the disease except that we could not ascertain if he had hypergammaglobulinaemia because we lacked the facilities to test for gammaglobulin. The extranodal manifestation of the disease has been documented in about 25 - 40% of affected individuals, with the most frequent sites being the head and neck regions. The urogenital tract, breast, gastrointestinal tract, liver, pancreas and lungs have also been reported to be affected by the disease. Our patient did not have any extranodal disease.

Histologically, lymph nodes in this disease show pericapsular fibrosis and dilated sinuses, heavily infiltrated with large histiocytes, lymphocytes and plasma cells. The presence of emperipolesis, or the engulfment of lymphocytes and erythrocytes by histiocytes that express S-100, is considered diagnostic of RDD. [2] In this case, the authors lacked the facilities for testing an immunohistochemical stain for S-100 positivity in histiocytes, and therefore it was not done on this patient’s biopsy.

Although the aetiology of RDD is unknown, it is presumed to be due to an immune regulation disorder. Patients with clinically apparent immunological disorders have a higher mortality. There also appears to be a familial relationship with regard to RDD: two sets of twins, two other sets of siblings and four individuals of blood relatives were documented by Foucar [8] in 1984. RDD has also been reported following bone marrow transplants for precursor-B acute lymphoblastic leukaemia. [11] A previously unreported association with sickle cell disease. BMC Clin Pathol 2007;7:3. http://dx.doi.org/10.1186/1472-6890-7-3

As previously mentioned, the patient was lost to follow-up, probably because of emotional burn-out from repeated hospital visits and perceived slow response to treatment. Similar observations have been documented in parents of children with incurable chronic diseases in Nigeria. [13]

Conclusion

This case report highlights this rare disease entity in order to raise a high index of suspicion on diagnosis. Patients who present with lymphadenopathy and who do not respond to treatment with antibiotics or anti-TB treatment must be followed up. These patients may need a biopsy to exclude malignant diseases or other diseases such as RDD.


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