Rosai-Dorfman Disease: A Case Report

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Citation

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Abstract

A 10-year-old Bidayu girl (DL) presented at the Sarawak General Hospital in 2004 with bilateral neck swellings. She had asymptomatic bilateral lympadenopathy of three months duration. She had fever initially. The enlarged lymph nodes clustered in the upper neck especially in the posterior triangles and were of the sizes up to 5 cm. in diameter. She was otherwise found to be healthy on examination. Her full blood count was normal, and ESR raised. Sputum examination for acid-fast bacilli was negative. Chest Xray was clear. Enodoscopy of the nasopharynx showed lymphoid mass in the roof, which was biopsied, and the result confirmed lymphoid tissue and no evidence of malignancy. Fine needle aspiration cytology of the enlarged lymph nodes on the right posterior triangle showed lymphadenitis and biopsy to exclude lymphomas was suggested. An isolated lymph node in right side of her neck was excised and the histology revealed Rosai-Dorfman disease. She was not given any form of treatment. Her neck nodes had decreased in size over the follow-up period and no nodes well clinically palpable 14 months after the initial presentation. Brief review of the literature of the disease and discussion in the case were presented.

INTRODUCTION

Rosai-Dorfman disease[1,2] (RDD or sinus histiocytosis with massive lymphadenopathy, SHML) is a rare, benign proliferative disorder of histiocytes, sometimes showing familiar incidence. It is characterized in the majority of cases by painless bilateral lymph node enlargement in the neck, often associated with fever and leucocytosis[3]. It presents mainly in the first or second decades of life, but any age group can be affected[4]. Without the awareness of RDD, the diagnosis of RDD is unexpected and surprising. Especially in South East Asian countries where certain lymphadenopathies such as tuberculosis, metatatatic nasopharyngeal carcinoma and lymphomas are common, RDD is unnoticeably missed, and its revision would seem worthwhile with this case illustrating the disappearing of the lymphadenopathy without treatment

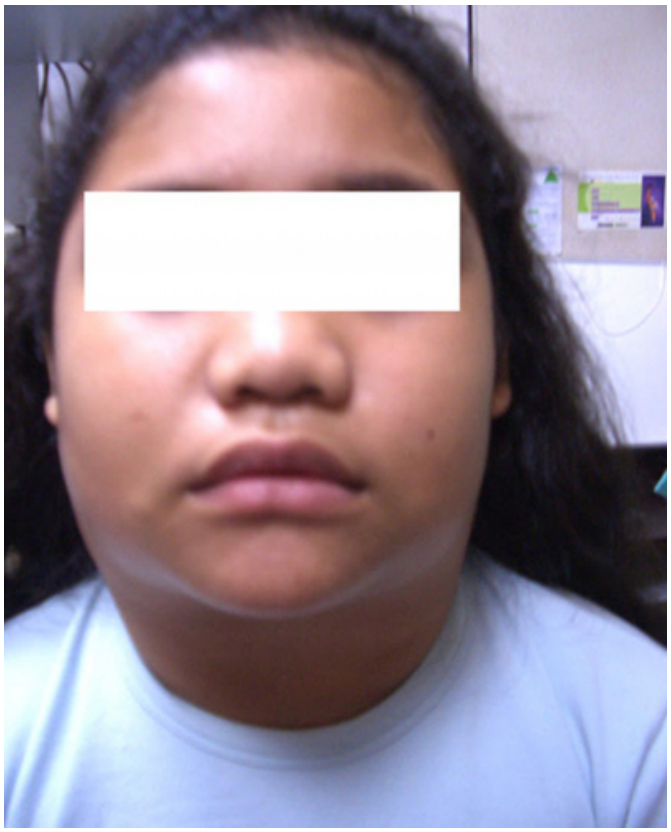
THE CASE

A Bidayu girl, aged 10, was referred by a local Polyclinic to the Out-patient Clinic of Sarawak General Hospital. Her mother was worrying over the increasing sizes of her neck swelling over a period of three months. She had fever for a short time when the neck swellings started and the local doctor thought the likelihood of mumps. The neck swellings were noted initially in her right side and then her left side. She had no history of pain, respiratory tract infection, and no symptoms related to her ears, nose and throat. She had no loss of appetite or weight. She had been healthy and had no past history of tuberculosis contact or family history of malignancy. There was no family history of immunologic diseases.

Clinical examinations of her ears, nose, throat, and head were normal. She had bilateral neck swellings found to be lymph nodes in the upper part of her neck (Fig 1). Most of the nodes had clustered mainly in the upper posterior triangles up to size of 5cm and a few smaller isolated ones lower in mid neck areas. The nodes were matted and felt firm to hard. They were not tender and had reduced mobility. The cranial nerve examination was normal.

Figure 1

Figure 1: Patient DL AP view at initial presentation



Endoscopy of the nose and nasopharynx showed lymphoid mass in the roof of nasopharynx slightly bigger on the right and this was biospied. There was on abnormality in the nose and throat. No abnormal findings were noted in examinations of the chest, abdomen and limbs.

Fine needle aspiration cytology (FNAC) was done on the right upper lymph nodes mass and bloodstained aspirate was obtained.

Chest Xray was done on her first visit and was normal.

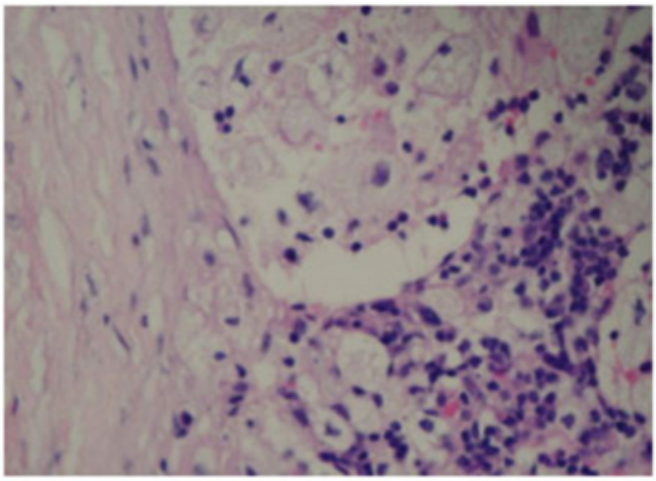
Full blood count and ESR showed Hb 12g/dL, WCC 8.5x109/L and ESR 35 mm/hr.

The nasopharyngeal mass biopsy report stated benigh lymphoid tissue with no evidence of malignancy. FNAC showed polymorphous lymph node aspirate, likely to be lympadenitis and Hodgkins' lymphoma could not be excluded. If lymph node enlargement persisted, a biopsy was needed.

Then the excisional biopsy was done on an isolated enlarged lymph nose in the posterior triangle. The histology report was: sections showed lymphoid tissue with prominent dilatation of lymph sinuses resulting in partial architecture effacement. The primary lymphoid follicles were present mostly at the cortical areas. The sinuses were occupied by numerous histiocytes and fewer lymphocytes and plasma cells. In addition, there were frequent emperipolesis phenomena seen. No lymphomatous change was noted. The interpretation was Rosai-Dorfman's disease. (Fig. 2)

Figure 2

Figure 2: Histopathology patient DL



She was not given any form of treatment. Her neck nodes had decreased in size over the follow-up period and no nodes well clinically palpable 14 months after the initial presentation.

DISCUSSION

Rosai-Dorfman disease is a benign condition characterized frequently by spontaneous remission. Its aetiology is uncertain. It manifests mainly with an asymptomatic latero-cervical lymphadenopathy, occasionally with extranodal locations. The illness usually takes on an insidious and self-limiting course, unless there happens to be an important involvement of vital organs[3].

Literature reviews show about 600 cases of RDD had been reported till 2004, in all races but mainly in the whites (43%), in any age group but mainly in first and second decades (81%), and in more male than female (2:1)[4].

RDD can present in extra-nodal form that usually affects an older patients. In this patient DL no extra nodal disease was seen

Macrosocopic and microscopic features had been well described[6]. Briefly the gross examination of the removed lymph nodes shows 1 or more enlarged lymph nodes, which

often are matted together by a prominent perinodal and capsular fibrosis. A polymorphous cellular infiltrate composed of neutrophils, lymphocytes, plasma cells, medium-sized mononuclear cells with indented nuclei, and, typically, numerous large and distinctive Rosai-Dorfman (RD) cells expand lymph node sinuses. RD cells have round-to-oval, medium-to-large nuclei with a vesicular chromatin pattern. Most nuclei contain a centrally located, single nucleolus. It is eosinophilic and small, but distinct and sometimes prominent. Nuclear atypia is unusual. Cytoplasm is abundant, and with hematoxylin and eosin staining, it appears lightly eosinophilic or, less frequently, amphophilic or clear. Phagocytized intact-appearing lymphocytes are present in the cytoplasm of most RD cells (lymphocytophagocytosis or emperipolesis: : “wandering about within”), which is a diagnostic feature. Internalized lymphocytes are usually located within cytoplasmic vacuoles.The other diagnostic feature is that RD cells are S-100 protein-positive on immunostaining, so differentiating from ordinary histiocytes.

In the early stages, the lymph node architecture is usually preserved, whereas in RD disease of long duration, proliferation of RD cells may massively dilate the sinuses, with complete effacement of normal nodal architecture[7]. This was seen in our case as enlarged lymph nodes with architecture effacement.

In light of the uncertain aetiological origin of the disease and of its frequent spontaneous remissions, treatment still does not have any set guidelines. Most aymptomatic cases progress to resolution with time of several months and no therapy is needed.. Symptomatic cases are treated symptomatically including corticosteroids, cytotoxics, radiotherapy and surgery, singly or in combination, in according to the complications resulted from the spread of the disease[3].

In general, RDD is diagnosed on biopsy, and rarely suspected by clinical evaluation. Patients with extranodal RDD however should have a diagnostic workup for immunologic abnormalities, since almost half of these patients with extranodal head and neck manifestations have immune dysfunction[8]. No immunologic workup was done in our case, as there was no related history, no symptoms and no extranodal features.

Differential diagnoses in asymptomatic cervical lmphadenopthy are many. Commonly in Malaysia three major groups are usually considered first, namely metastatic malignancy (especially nasopharyngeal carcinoma in the male patients), tuberculosis and lymphomas.

Pathological differential diagnoses of RDD include reactive sinus hyperplasia, Langerhans cell histiocytosis, Hodgkin's disease, metastatic carcinoma, malignant melanoma, and lymphoma. The differentiation of these various diseases had been well-described [6].

In the case that we observed, most of the histological features were seen and a conservative approach was taken. After 14 months of follow-up, the patient had spontaneous full recovery. The only symptom that this patient had was fever of short time at the onset of the disease thought by the examining doctor to be a viral illness of mumps at the time.

SUMMARY

Rosai-Dorfman disease is a rare, idiopathic, benign histiocytic proliferation, most commonly involving cervical lymph nodes. Its aetioloy and pathogenesis is unknown. The diagnosis is by biospy. When patients are asymptomatic, conservative observing approach is started with regular follow-up as in our case. Most of these patients would recover fully. Our case is presented to emphasize this general approach and the need to be aware of a differential diagnosis of RDD in asymptomatic cervical lymphadenopathy.

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