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Case Report

Rosai-Dorfman disease in a child with involvement of extra-nodal wrist joint

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ABSTRACT

Rosai-Dorfman Disease (RDD) is a benign condition primarily affecting the lymph nodes. The term sinus histiocytosis with massive lymphadenopathy was first used. There may be extra-nodal presentation of RDD with or without constitutional symptoms. A 10-year-old boy presented with massive bilateral cervical lymphadenopathy which mimicked the diagnosis of lymphoma. We describe here a case of RDD in a child with extra-nodal bone involvement of the wrist joint, its diagnosis by histopathological examination supported by immunohistochemistry and consequent initial management.

Keywords: Bone involvement, Child, Lymphadenopathy, Rosai-Dorfman disease

INTRODUCTION

Rosai-Dorfman Disease (RDD) is a rare benign disease of unknown etiology. It was first termed as a separate entity called sinus histiocytosis with massive lymphadenopathy by Rosai and Dorfman in 1969.[1] Patients of any age may be affected, but more frequently it is seen in children and young adults. The disease is more common in the males and in individuals of African descent. Etiology is idiopathic, but viruses like Herpes virus 6 and Epstein-Barr virus have been implicated as potential causative agents without any strong evidence.[2,3] Commonest clinical manifestation of RDD is massive bilateral painless cervical lymphadenopathy. Extra-nodal involvement is also seen in RDD.[4] We describe here a case of RDD in a child with extra-nodal osseous involvement of the wrist joint, its diagnosis by histopathological examination (HPE) supported by immunohistochemistry (IHC) and consequent initial management.

CASE REPORT

A 10- year- old boy reported at our institute with the chief complaint of progressive swelling on both sides of the neck (Figure 1) of 5 months duration. It was associated with fever and pain in the right wrist joint. There was no contact history with tuberculosis patient. On local examination there were multiple swellings on both sides of the neck at level II, III, IV and V which were firm, mobile, tender and discrete. The largest node was 3cm x 3cm in its maximum dimension. The wrist joint was tender without any palpable swelling. Chest skiagram showed mediastinal adenopathy. X-ray of the right wrist joint revealed osteolytic lesion on the ulna (Figure 2). There were para-aortic and retroperitoneal

nodes on ultrasonography (USG) of abdomen. Fine needle aspiration cytology (FNAC) of cervical lymph node was inconclusive, so incision biopsy was done for the HPE. It showed features favoring a diagnosis of RDD (Figure 3a). IHC study was done for CD20, CD3, CD30, epithelial membrane antigen, S100 and CD15. IHC showed positive expression of S100 in the sinusoidal histiocytes (Figure 3b) which confirmed the diagnosis of RDD. Thus, the diagnosis of RDD with extra-nodal bone of wrist joint involvement was made after clinical, radiological examination, HPE and IHC examinations. The patient was managed conservatively for the symptoms and his parents were further advised for seeking medical attention on recurrence of symptoms.

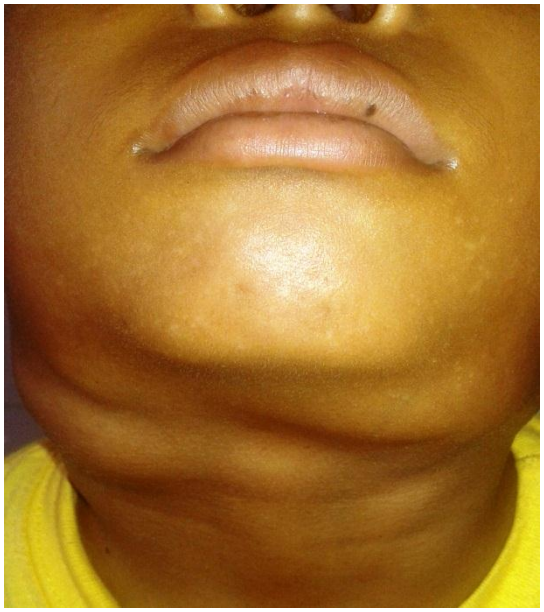

Figure 1: Showing the bilateral cervical lymphadenopathy.

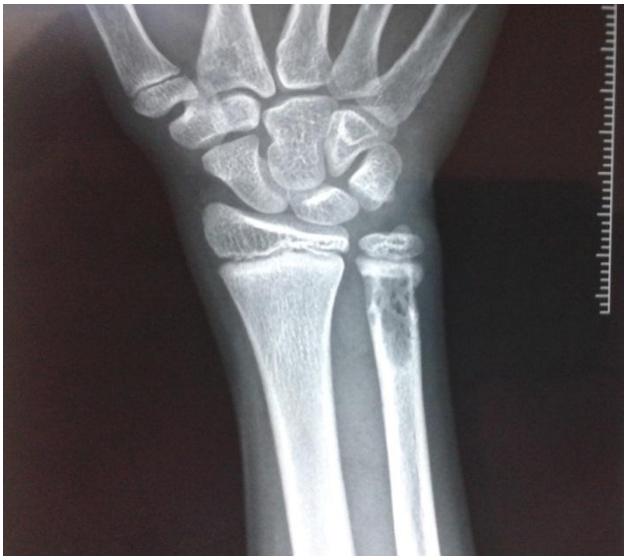

Figure 2: X-Ray showing the osteolytic lesion in the distal end of ulna.

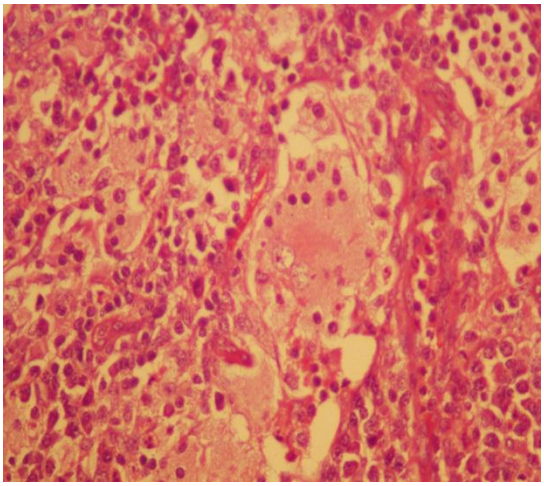

Figure 3a: Photomicrograph with H & E stain (40x) showing prominent dilatation of sinuses filled with pale looking histiocytes with abundant cytoplasm.

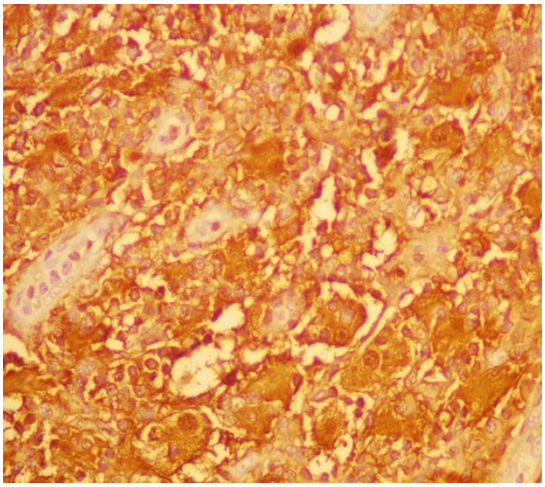

Figure 3b: Photomicrograph showing sinusoidal histiocytes with emperipolysis shows immunoreactivity to S100.

DISCUSSION

RDD is a rare, non-neoplastic, proliferative disorder of histiocytes usually affecting the lymph nodes. The usual clinical presentation is massive bilateral cervical lymphadenopathy which is painless and can involve the entire jugular chain. Also, it may involve other lymph nodal areas like the mediastinum, axilla and inguinal region. In the present case there were mediastinal and retro-peritoneal adenopathies as well. In our patient, there was associated constitutional symptom of fever and joint pain. The presence of fever warrants the exclusion of co-existence of Koch's infection in endemic areas. In this patient the presence of active tuberculosis was excluded by the following; absence of symptoms like history of weight loss and loss of appetite, absence of lung parenchymal disease on radiological examination, and typical HPE features of the node for RDD. Extra-nodal RDD frequently occurs in the skin, soft tissue, upper

respiratory tract, multifocal bone and retro-orbital tissue with lymphadenopathy. Other rare extra-nodal sites are lungs, thyroid, testis ,kidney ,breast ,pancreas and gastrointestinal tract.[5] Osseous involvement in RDD is seen in less than 8% of patients and when bony lesions are seen, it frequently involves the skull, femur, clavicle and tibia.[6-9] In the present case the extra-nodal involvement of bone was at the distal end of ulna. Isolated bone involvement is rare in RDD.[10] Our patient had single extra-nodal anatomic osseous involvement which was associated with lymphadenopathies. RDD has been shown to manifest with an unusual presentation of a spinal cord tumor.[11]

Positivity of S-100 protein is characteristic of RDD. It was confirmatory for RDD in the present case. IHC may also be positive for the expression of CD68, CD163, α1-antichymotrypsin, α1-antitrypsin, fascin, and HAM-56.[12] But, these additional IHC markers were not examined in the present study. Furthermore, the expression of CD3 and CD20 were negative, which excluded the reactive nature of the lymphadenopathy. The extra-nodal sites of involvement may demonstrate a histological pattern similar to nodal architecture mimicking nodal involvement.[13] In the present case, the diagnosis of extra nodal RDD at ulna was made only by radiological examination by the presence of a well-defined lytic lesion. Radiological examination has been shown to assist in the radiological diagnosis of Langerhans cell histiocytosis, including RDD.[14]

The overall prognosis of RDD is good. Most of the patients are kept under observation, but patients with recurrent symptoms, lymph node enlargement and fever have been successfully treated with systemic steroids. Extra-nodal disease with respiratory and compressive symptoms and which are non-responsive to steroids will necessitate surgical intervention. Chemotherapeutic agents like vinca alkaloids, vinblastin, methotrexate, etoposide and 6 mercaptopurine were used in some selected cases with good results. 2-chlorodeoxyadenosine (2-CdA,cladribine) have been used when other approaches had failed. [15] Radiotherapy and rituximub are also used with satisfactory results in steroid resistant extra-nodal RDD.[16] Mortality in RDD is extremely rare and occurs in cases with disseminated disease, involvement of rare extra-nodal site and with systemic immunologic disorders.[17] Our patient had remission of symptoms of fever and pain with conservative management.

CONCLUSION

RDD is a non-malignant disease affecting the lymph nodes and extra-nodal solitary involvement of ulna in a child is rare. Patients with RDD can be managed conservatively to begin with. Systemic steroids should be used in patients with recurrence of symptoms after excluding the presence of Koch's infection to avoid a flare up of tuberculosis.

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