ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 7, Issue, 05, pp.16484-16486, May, 2015

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

CASE STUDY

A CASE OF EXTRA NODAL ROSAI – DORFMAN DISEASE of LUMBO SACRAL SPINE

[1]Sawsan A. H. Aldeaf, [2]Ahmed M. Elhassan, [4]Lamyaa A. M . Elhassan, [*,1]Alsadig Gassoum [1]Mohamed Saad A. Saad and [3]Mohamed A. Arbab

[1]National Center of Neurological Sciences, Sudan
[2]Institute of endemic diseases, University of Khartoum Sudan
[3]Faculty of Medicine Khartoum University
[4]Faculty of Medicine, Ahfaad University of Women

ARTICLE INFO

Article History:
Received 20th February, 2015
Received in revised form
02nd March, 2015
Accepted 25th April, 2015
Published online 31st May, 2015

Key words:

Rosi-Dorfman disease,
Lumbo sacral spine.

ABSTRACT

Background - Rosai – Dorfman disease, or sinus histiocytosis with massive lymphadennopathy was first described as clinical distinct clinicopathlogical entity in 1969, in 43% of cases, extranodal sites are involved. The disease is most common in children and young adult. It has morphological features of greatly exaggerated reactive process. The etiology of the disease is uncertain, possible causes include, abnormal immune response and infections.

Case history- A 20 years young man, born of consanguineous marriage, presented to our outpatient neurosurgical clinic with a history of rabidly progressing backache over the course of 6 months, he had nocturnal low grade fever and lower limbs pain for the same duration. Peripheral blood picture showed, Neutrophils 71%, lympocytes 18%, monocytes 9%and essinophils 2%, ESR 50 mm/hr. biochemical profile of renal and liver functions was normal. CT chest revealed no abnormality. MRI lumbo sacral spine was done and revealed, Straight lumber spine, there is an extra dural mass lesion noted occupying the posterior aspect of the dural sac, extending from the level of L1 lower border down to the upper border of L5. The patient underwent partial laminectomy L3 L5 and excision of the tumor. MRI in our case revealed that , diffuse enhancing lobulated masses without bony destruction this findings may be a diagnostic challenges for radiologist, although the diagnosis of Rosai dorfman disease depend on histopathological features and immno stain.

Citation: Sawsan A. H. Aldeaf, Ahmed M. Elhassan, , Lamyaa A. M . Elhassan, Alsadig Gassoum, Mohamed Saad A. Saad, and Mohamed A. Arbab. 2015. "A case of extra nodal Rosai – Dorfman Disease of Lumbo Sacral Spine", *International Journal of Current Research,* 7, (5), 16484-16486.

INTRODUCTION

Rosai – Dorfman disease , or sinus histiocytosis with massive lymphadennopathy, was first described as clinical distinct clinicopathlogical entity in 1969 –(Rosai and Dorfman). The disease is most common in children and young adult (Rosai and Dorfman). It has morphological features of greatly exaggerated reactive process. It usually presents with painless cervical lymphadenenopthy. In 43% of cases, extranodal sites are involved simultaneously and in only 23% isolated Rosai-dorfman disease occurs. (Foucar, Rosai, and Dorfman) The etiology of the disease is uncertain, possible causes include abnormal immune response and infections by Varicella Zoster and other herpetic viruses, Epstien Barr, cytomegalo virus, Brucella and Klebisiela. (Levine *et al.*) Since it is considered as a benign or reactive proliferation with self limiting course, however, the treatment option range from surgery, radiotherapy and steroid or chemotherapy.

Corresponding author: Alsadig Gassoum,
National Center of Neurological Sciences, Sudan.

Case history

A 20 years young man, born of consanguineous marriage, presented to our out patient neurosurgical clinic with a history of rabidly progressing backache over the course of 6 months, he had nocturnal low grade fever and lower limbs pain for the same duration. He developed gradual lower limbs numbness with unsteady gait few days before his presentation. There was history of chronic rhinitis but no cough and there was also history of anorexia and weigh loss. No significant family history of the same condition.

Physical examination of the patient revealed unwell thin under weigh patient, with tenderness over the lumbo sacral area and brisk reflexes in his lower limbs. Laboratory investigations showed hemoglobin of 11.5 g/dl and total leucocytes count 16200/cu mm and platelets 480000 cu/mm. Peripheral blood picture showed, Neutrophils 71%, lympocytes 18%, monocytes 9%and essinophils 2%, ESR 50 mm/hr. biochemical profile of renal and liver functions was normal. CT chest revealed no abnormality. Ultra sonography of the whole abdomen showed no organomegally or lymph node enlargement. In addition to that, serum screening of EBV and HIV was negative. MRI lumbo sacral spine was done and revealed, Straight lumber spine, there is an extra dural mass lesion noted occupying the posterior aspect of the dural sac, extending from the level of L1 lower border down to the upper border of L5. An other similar lesion was noted involving the sacral region. The lesion modulate it self to fill the configuration of the interlacing ligamentum flavum without effecting the adjacent bone, this is indicating its soft consistency. The lesion of the lumber region compresses the cauda equine and the tip of the conus medullaris. At multiple levels, the lesion extends through neuroforamina and compressing the existing nerve roots.

The one in the sacral region shows encasement of the descending roots, ie obliterating the thecal sac circumferentially with the clustering nerve roots. In addition through the existing neuroforamina, the mass is noted in Para vertebral spaces bilaterally. In the sacral region posterior scalloping is noted. The mass showed iso intense signal to the cord parenchyma on T1W and relatively high signal on T2W, homogenous post contrast enhancement noted. No marrow signals changes, no spondylolithesis, discs are unremarkable and normal facet joints. (Fig. 1)

The patient underwent partial laminectomy L3 L5 and excision of the tumor, it was soft bluish mass lobulated compressing the spinal theca, the part that over the sacral segment is left behind,the patient recover smoothly from anesthesia with no neurological deficit. Histopathology showed nodular mass of soft tissue consisting of a mixture of cells. The most conspicuous cells in some nodules are large cells with pale or vacuolated cytoplasm and vesicular nuclei. The cells are phagocytosing lymphocyte, plasma cells and occasional red cells. The other areas of section consist of lymphocytes and plasma cells. Some of the latter contain Russell bodies. The large cells are positive for S- 100 protein and macrophages marker CD 68. They are negative for the Langerhans cell marker CD1a.The patient was given steroid for the treatment of the remnant part left over the sacral area. Fig. (2, 3, 4, 5 and 6)

DISCUSSION

Extra nodal type of Rosai dorfman disease is seen in around 43% of patients, and the common site are skin, upper respiratory tract, soft tissue and bone ((Hsiao et al.). More over other site include: breast gastrointestinal tract, head and neck and central nervous system and the later may be intra cranial or spinal. In cranial, utually the lesion attaches to the dura and this may lead to radiological feature of meningioma. Rosai Dorfman disease has certain distinctive histological feature which characterize by lymphoplasmacytic infiltration, and the histocytes in this disease are positive to immune marker CD 68, S100 protein and negative for CD1a (Lopez and Estes). Our findings for these markers showed similar results. The most common clinical features of the disease are bilateral cervical lymphadonopathy and constitutional symptoms such as fever weight loss, in Sino nasal type nasal discharge, epistaxis and upper respiratory tract infections like tonsillitis pharyngitis are the cardinal symptoms, but in our patient CT nasal sinuses was negative for sinus obstruction. The spine MRI of the patient, was suggestive of plexiform Neurofibroma, however the anatomical location and in the absence of scalloping of the vertebral body and in the absence of cutaneous lesion this makes the diagnosis is unlikely. The differential diagnosis of Lymphoma and secondary metastasis was made but no bone invasion.

MRI in our case revealed that, diffuse enhancing lobulated masses without bony destruction may be diagnostic challenges of radiologist although the diagnosis of Rosai dorfman disease depend on histopathological feature and immno stain. Other laboratory screening are non specific, elevation of sedimentation rate have been reported in many cases as well as in our patient. The benign course of the disease and being self limited, definite treatment is still questionable but if the patient having extra nodal type with involvement of vital structure like brain or spinal cord, in this circumstances, surgical treatment is limited,(Deodhare, Ang, and Bilbao). More over systemic steroid may be beneficial in decreasing the nodal being immunosuppressive.

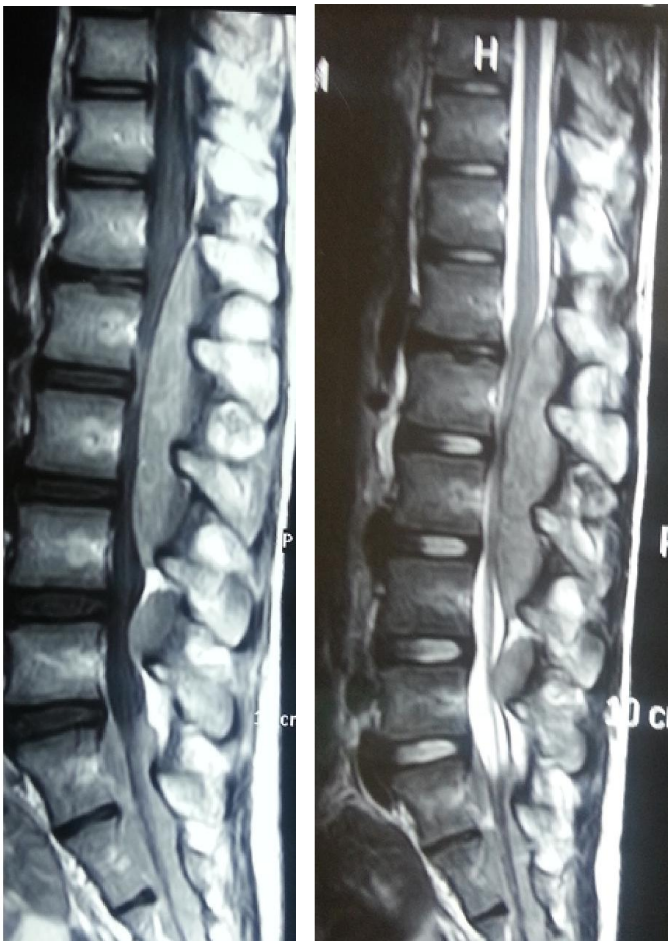

Fig. 1. Shows on the right site sagittal cut of T1 with contrast and on left site T2

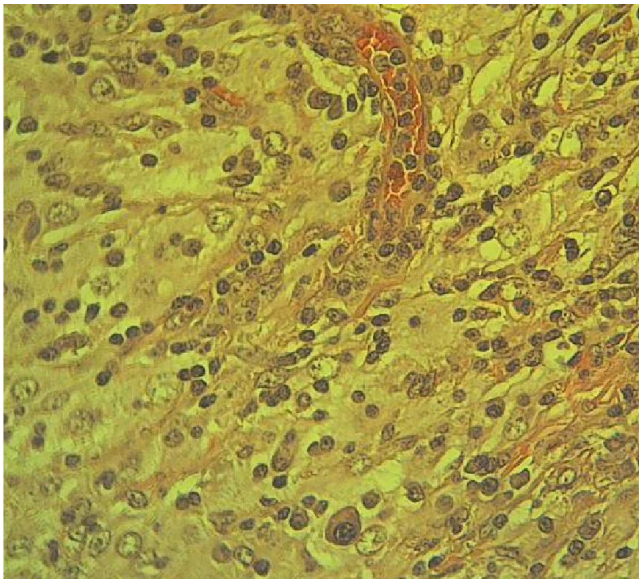

Fig. 2. Nodular mass of soft tissue shows abundant of histiocytes, plasma cells and lymphocytes

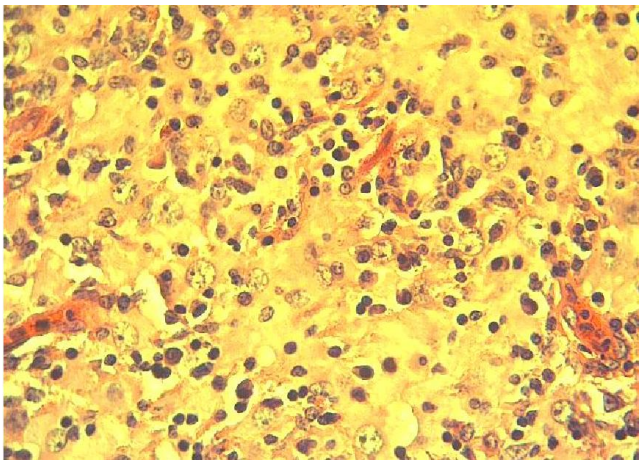

Fig. 3. Shows plasma cells, lymphocytes and histiocytes

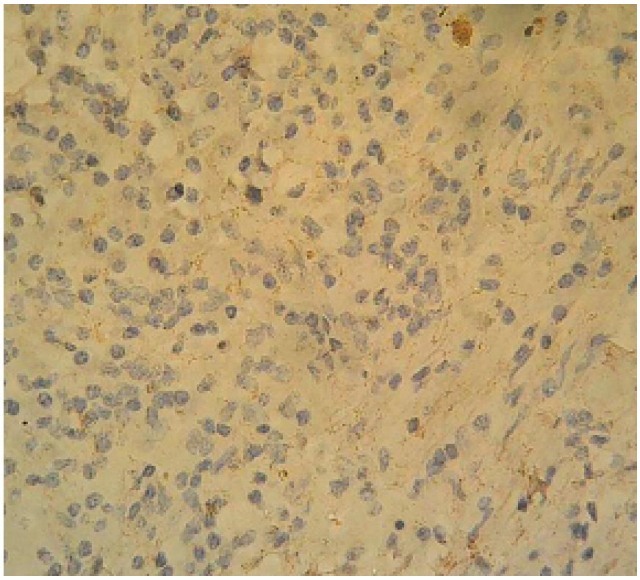

Fig. 4. Shows negative immunostaining for CD1a marker

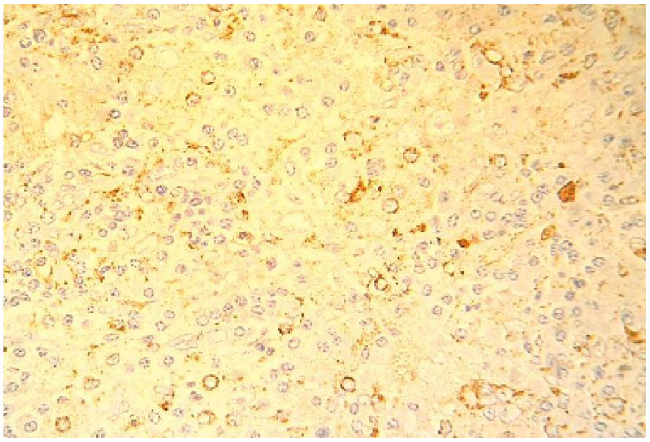

Fig. 5. Shows positive immunostaining for CD68 marker

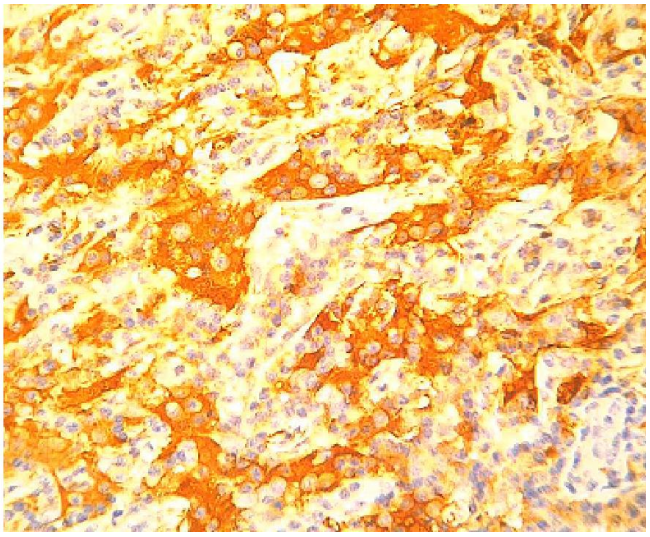

Fig. 6. Shows positive immunostaining for S100 protein marker

REFERENCES

Deodhare, S. S., Ang, L. C. and Bilbao, J. M. 1998. "Isolated intracranial involvement in Rosai-Dorfman disease: a report of two cases and review of the literature." *Arch .Pathol. Lab Med.* 122.2 161-65.

Foucar, E., Rosai, J. and Dorfman, R. F. 1978. "Sinus histiocytosis with massive lymphadenopathy." *Arch. Otolaryngol.,* 104.12 687-93.

Hsiao, C. H., *et al.* 2006. "Clinicopathologic characteristics of Rosai-Dorfman disease in a medical center in northern taiwan." J.Formos.Med.Assoc. 105.9 701-07.

Levine, P. H., *et al.* 1992. "Detection of human herpesvirus 6 in tissues involved by sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease)." *J. Infect. Dis.,* 166.2 291-95.

Lopez, P. and Estes, M. L. 1989. "Immunohistochemical characterization of the histiocytes in sinus histiocytosis with massive lymphadenopathy: analysis of an extranodal case." *Hum. Pathol.,* 20.7711-15.

Rosai, J. and Dorfman, R. F. 1969. "Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity." *Arch. Pathol.,* 87.163-70.

Sinus histiocytosis with massive lymphadenopathy: a pseudolymphomatous benign disorder. 1972. Analysis of 34 cases. *Cancer,* 30.5 1174-88.

****