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Review Article

ROSAI DORFMAN SYNDROME – DIAGNOSIS ON FINE NEEDLE ASPIRATION CYTOLOGY WITH REVIEW OF LITERATURE

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ABSTRACT

Rosai-Dorfman disease, also referred to as Sinus histiocytosis with massive lymphadenopathy, is a rare non neoplastic proliferative disorder of the cells of macrophage-histiocyte family that was first described by Destombes in 1965. As per review of literature there are only few reports or small series cases on the (FNA) cytologic features of this entity. We report a case of Rosai-Dorfman disease presenting with massive bilateral cervical lymphadenopathy and conclude that FNAC is useful and reliable tool for diagnosis of Rosai-Dorfman disease avoiding the need of biopsy and proceed with accurate treatment and management of the patient.

INTRODUCTION

Rosai-Dorfman disease, referred to as Sinus histiocytosis with massive lymphadenopathy, is a rare non neoplastic proliferative disorder of the cells of macrophage-histiocyte family that was first described by Destombes in 1965[3]. It was recognized as a distinct clinic-pathological entity by Rosai and Dorfaman in1969. [4] The exact etiology of the disease is unknown [6]. It is presumed that an aberrant exaggerated immune response to an infectious agent causes proliferation of histiocytes. Although several infectious agents are suspected, none of them are documented so far [7]. The expression of HHV6 (Human Herpes Virus) specific p101k antigen was found in follicular dendritic cell in SHML.[7] As per review of literature there are only few reports or small series cases on the (FNA) cytologic features of this entity.[8]

Case Report

60 year male presented for FNAC procedure with multiple bilateral swellings in the neck since 6 months seen in submandibular, sub-mental, upper cervical and peri-auricular areas, accompanied by low grade fever with no history of weight loss. Local examination revealed size of swelling ranging in size from 1cm to 2.5cm in diameter, non-tender, soft to firm in consistency, non-matted, with normal overlying skin. Systemic examination was within normal limits with no organomegaly.

FNAC done from the multiple swellings showed highly cellular smears comprising of polymorphous population of lymphoid cells comprising of small lymphocytes, centrocytes, centroblasts, immunoblasts and abundant binucleated and multinucleated histiocytes. Occasional ill-formed epithelioid cell clusters were also seen. Few large histiocytes engulfing lymphocytes were also seen. Differential diagnosis of Rosai-Dorfman disease and? Hodgkins Lymphoma was given (as few Reed Sternberg like cells were also seen). (Figures 1 and 2)

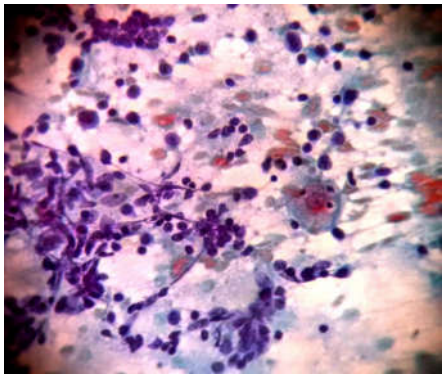

Figure 1 PAP stained smears prepared from fine needle aspirate at 10x magnification showing emperipolesis.

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Histopathological examination revealed capsulated lymphoid tissue with underlying dilated sinuses filled with histiocytes. Few lymphoid follicles with histiocytes and macrophages showing emperipolesis were also seen (Figure 2). Immunohistochemistry was diffusely positive for S-100, CD 68 (Figure 3) and negative for CD 1a thus confirming the diagnosis of Rosai- Dorfman disease.

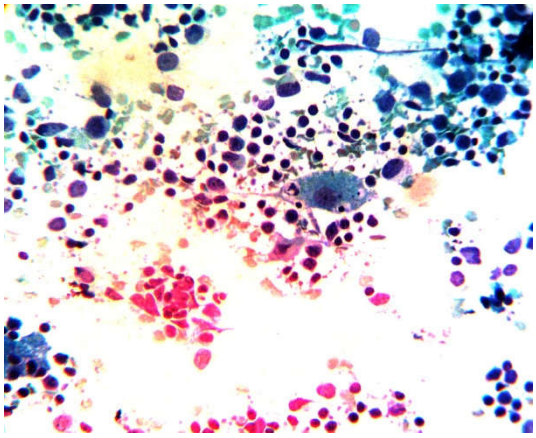

Figure 2 PAP stained smears prepared from fine needle aspirate at 10x magnification showing emperipolesis.

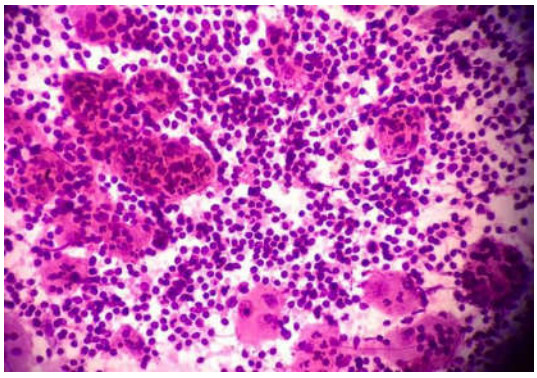

Figure 3 H and E stained smear from biopsy specimen showing emperipolesis and sinus histiocytosis at 10x magnification.

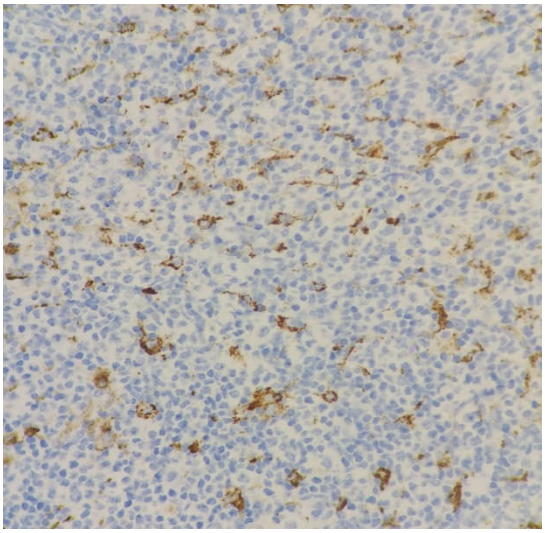

Figure 4 Immunohistochemistry- Histiocytes showing focal positivity for CD 68 at 10x magnification.

DISCUSSION

Rosai–Dorfman disease (RDD), originally described as sinus histiocytosis with massive lymphadenopathy (SHML), presents in its most typical form as massive, painless, bilateral lymph node enlargement in the neck, associated with fever, leukocytosis, elevated erythrocyte sedimentation rate, and polyclonal hypergammaglobulinemia. [(1)]

This condition was first described by Robb-Smith in 1947 in children and was termed as giant cell sinus reticulosis. Sinus histiocytosis with massive lymphadenopathy (SHML) has been recognized as a distinct clinicopathological entity, though first given this name by Rosai and Dorfman in 1969, 1972. [(5)]

Boys and men are slightly more oftenaffected than are girls and women[(2)]. Although the cervical region is by far the most common and most prominent site of involvement, other peripheral or centrallymph node groups can be affected, with or without cervical disease.[(1)] Grossly, the nodes are matted together by prominent perinodal fibrosis. Their cut surface varies from gray to golden yellow, depending on the amount of fat present [(1)]. Microscopically, there is a pronounced dilation of the lymph sinuses, resulting in partial or complete architectural effacement. These sinuses are occupied by lymphocytes, plasma cells, and – most notably – by numerous cells of histiocytic appearance with a large vesicular nucleus and abundant clear or lightly eosinophilic cytoplasm that may contain large amounts of neutral lipids. Many of these histiocytes have within their cytoplasm numerous intact lymphocytes, a feature that has been designated asemperipolesis or lymphocytophagocytosis[(1)].The sinusoidal histiocytes stain for S-100 protein and CD68, similar to the cells of Langerhans cell histiocytosis but unlike the histiocytes of reactive sinus histiocytosis[(2)].

The differential diagnosis of Rosai- Dorfman Disease includes nonspecific sinus hyperplasia (in which the cells lack emperipolesis and are S-100 protein-negative), Langerhans cell histiocytosis (in which the cells are positive for both S-100 protein and CD1a), leprosy, rhinoscleroma (with which it can apparently coexist), and metastatic malignant melanoma. Perhaps the condition that ensembles it most is the sinus histiocytosis induced by cobalt-chromium and titanium that can occur in pelvic lymph nodes after hip replacement. It should also be noted that focal RDD-like changes can sometimes be seen in lymph nodes involved by other processes, such as Hodgkinor non-Hodgkin lymphoma, a phenomenon analogous to that sometimes seen in Langerhans cell histiocytosis[(1)].

CONCLUSION

Thus the cytomorphology of Rosai-Dorfman disease is so distinctive that it can be diagnosed by FNAC. FNAC is a reliable and sensitive means to establish conclusive diagnosis, obviating the need for biopsy.

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