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Original Research Paper

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DIAGNOSIS OF ROSAI–DORFMAN DISEASE BY FINE NEEDLE ASPIRATION CYTOLOGY- A STUDY OF ELEVEN CASES WITH EMPHASIS ON SITE

Dr. Rinsha Surendranath — Post Graduate, Department of Pathology, Govt. Siddhartha Medical College, Andhra Pradesh ,India.

Dr.M Rajani* — Department of Pathology, Govt. Siddhartha Medical College, Andhra Pradesh ,India.*Corresponding Author

ABSTRACT Rosai Dorfman Disease also called as Sinus Histiocytosis with Massive Lymphadenopathy is a benign proliferative disorder of histiocytes which typically affects lymph nodes of cervical region ,however extranodal sites are also being reported by many authors[1].FNAC is the investigation of choice as is it less invasive and more reliable.**Aims :**To study the various sites of involvement and diagnostic significance of FNAC along with the pattern of site and age distribution .We studied a series of 11 cases for a period of one year.**Materials and methods:** Eleven cases studied in the department of Pathology Govt. Siddhartha Medical College,Vijayawada.Material collected from Govt. General Hospital,Vijayawada. **Results:**The present study showed two extranodal,one axillary lymph node,one inguinal lymph node and seven cervical lymph node involment. **Conclusion:**Rosai- dorfman disease main clinical manifestion is cervical lymph node enlargement ,but other lymph nodes and extranodal site can also be involved.Most common age of presentation was between 20-40years

KEYWORDS : Rosai -Dorfman disease,Sinus Histiocytosis with massive lymphadenopathy,Emperipolesis.

INTRODUCTION

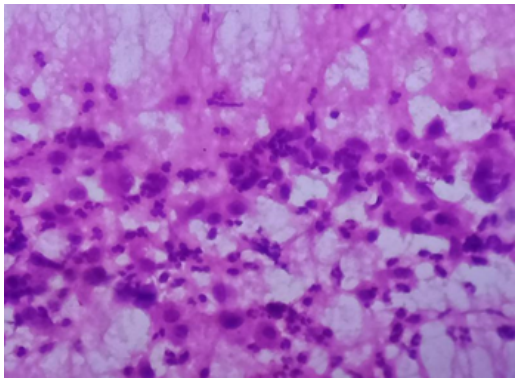
Rosai Dorfman Disease(RDD) also called as Sinus Histiocytosis of Massive Lymphadenopathy is a rare idiopathic proliferative disorder of histiocytes that has been diagnosed by FNA cytology.Typically the patients present with bilateral massive cervical lymphadenopathy, however it effects other lymph nodes and many extranodal locations . Clinically the symptoms and signs of the patient may mimic lymphoma.Despite its variable clinical presentation it is a benign disorder with self limiting nature.Hence FNAC is an important diagnostic tool as it cost effective ,less invasive and prevents unwanted surgeries, We are presenting 11 cases of nodal and extranodal Rosai Dorfman disease emphasizing the site of the involementand age disrtibution of the patients and the diagnostic significance by FNAC.

MATERIAL AND METHODS

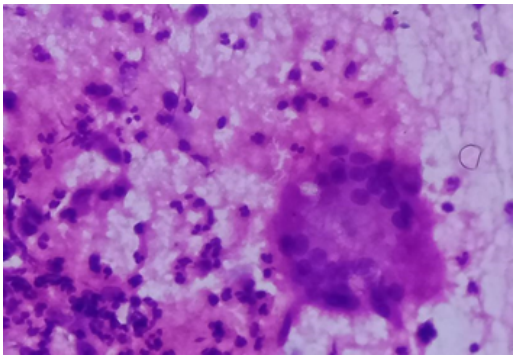
The cytologic features of 11 cases were studied in the department of Pathology Siddhartha medical College Vijayawada from July 2019 to June 2020 . After a brief history and clinical examination of the patients ,FNAC was performed using 22 gauge needle. The aspiration smears were fixed in isopropyl alcohol and stained with Hematoxylin and Eosin.Detailed cytomorphological examination of the slides were carried out.

RESULTS

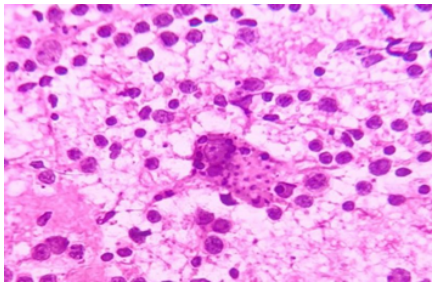
A total of 11 cases were studied .The most common age of presentation was found to be between 20-40years. The most common clinical presentation was lymphadenopathy seen in nine cases, in which cervial lymph node was involved in seven cases along with one case of axillary lymph node and one case of inguinal lymph node.Two cases of extranodal RDD presented as a unilateral breast lump and scalp nodule.Cytologically , the smears were highly cellular and showed proliferaton of large histiocytes with numerous prominent engulfed intracytoplasmic intact lymphocytes (emperipolesis). The histiocytes had abundant cytoplasm with large eccentic nucleus. The number of lymphocytes within the histiocyte varied from case to case. The backgound showed admixture of lymphocytes,neutrophils and multinucleated giant cells.



FigureA: High power magnification showing atypical histiocytes against a background of lymphocytes.



FigureB:High power magnification showing multinucleated giant cells in a smear with atypical lymphocytes

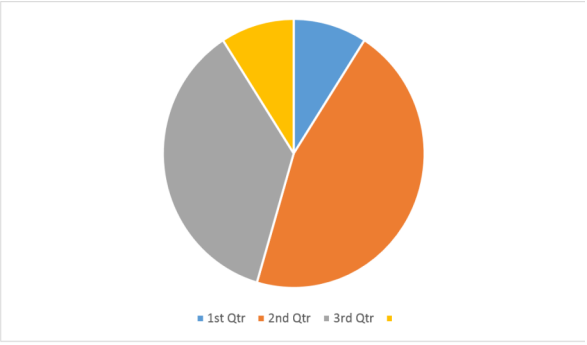


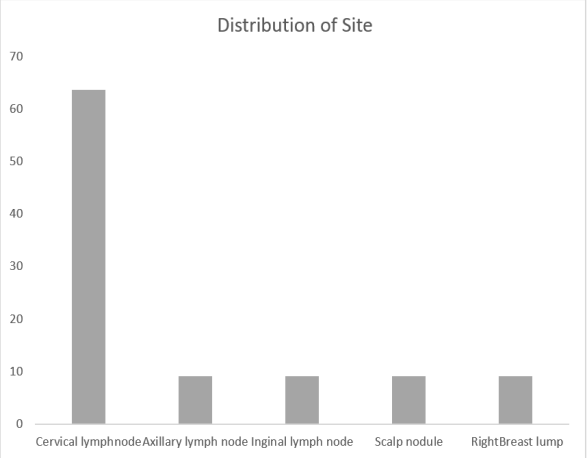
FigureC:High power magnification showing Emperipolesis.

Table1	Distribution of Site in RDD	
Serial number	Site of Involement	Percentage(%)
1	Cervical lymph node	63.63
2	Axillary Lymph node	9.09
3	Inguinal lymph node	9.09
4	Scalp nodule	9.09
5	Right breast lump	9.09

Table2:Age distribution of RDD		
Serial Number	Age of Patient in years	Percentage(%)
1	<20	9.09
2	20-40	45.45
3	40-60	36.36
4	>60	9.09

Age distribution of Rosai- Dorfman disease





DISCUSSION

Rosai Dorfman Disease also called as Sinus Histiocytosis of Massive Lymphadenopathy was first described by Pathologist Destombes[2]. Later in 1969, it was delineated as distinct clinicopathological entity ,Sinus histiocytosis of massive lymphadenopathy by Rosai and Dorfman[3].Since the original description ,several case reports and studies were published on this rare condition.However Rosai-Dorfman disease remains as an enigmatic condition with a variable clinical presentation and course. It can involve almost every organ and usually has a benign clinical course.It is commonly seen in the first and second decade of life , however it may effect the patient in any age group[3].Males are the dominant sex affected , however much studies are lacking in this area. The patient of RDD presents with painless massive lymphadenopathy ,fever, joint pain, loss of weight , leucocytosis, increased ESR and polyclonal hypergammaglobinemia. .Although the etiology of RDD is not completely known. Viral infections are most commonly implicated causative agents.Human herpes 6,[4] parvovirusB19[5], simian virus40 and polyoma virus[6] have been described by various authors as etiologic agents. An assocation with immunological disease like SLE and arthitis has also been described[7,8].

The polyclonal nature of proliferating histiocytes , also known as Rosai-dorfman cells, indicates that they are reactive rather than neoplastic[9].However, current studies identified mutation in BRAF,KRAS,NRAS and PIK3CA genes of RAS-MAPK pathway and proposed that a subset of RDD may be clonal[10,11].

Histiocytes are known for their phagocytic property. Emperipoles as defined by Humble is an active penetration of one cell by another where it remains intact [12]. It is a nondestructive phagocytosis of inflammatory cells, often lymphocytes and also plasma cells , neutrophils, and RBC.Emperipolesis is required to diagnosis RDD,the absence of which makes diagnosis difficult[13].

Immunohistochemically the histiocyte in RDD is positive for CD68 and S100 but negative for CD1a an d langerin which helps to differentiate RDD from langerhans cell histiocytosis, Histiocytes can also be seen prominently in reactive lymph nodes[14,15,].

Cytology has an important role in the diagnosis of RDD as it has a characteristic cytological features ensuring an accurate diagnosis The cellularity is mostly high with plenty of large histiocytes both with and without emperipolesis and also lymphocytes. The presence of plasma cells and neutrophils is a frequent finding[16,17] .In our present study aspirates from all the cases yielded a predominant population of lymphocytes and histiocytes. In both lymph nodal and extranodal RDD the predominant cells engulfed were lymphocytes.

Due to variable clinical presentation and histiocytic proliferation ,RDD can be confused with other disorders like Reactive lymphedinitis, Langerhans cell histicytosis,hodgkins lymphoma , granulomatous inflammation.

In our present study case number four we encountered a rare case of an adult female with a mass lesion over the occipital region with clinical suspicion of lipoma, however on cytological examination we found predominantly atypical histiocytes with prominent nuclei showing emperipolesis, along with tingible body macrophages, multinucleated giant cells ,lymphocytes and neutrophils. Presence of atypical histiocytes limited our diagnosis to histiocyte disorder on cytology.Significant emperipoles was seen by which we came to the diagnosis of RDD.

Case number eight showed another very uncommon presentation where a young adult female presented with tender swelling of right breast. On USG it was a hypoechoic lesion and clinically itwas thought to be breast abscess. On cytology the smear yielded high cellularity showing ductal epithelial cells and atypial histiocytes showing emperipolesis along with lymphocytes and necrotic debris. Breast is an uncommon site for RDD with only few cases described in literature so far. The clinical presentation is variable with single or multiple mass lesions in one or both breast, the radiology findings of RDD are non contributary and may simulate malignancy.The cytology is similar to that of nodal RDD except for less emperipolesis which make the diagnosis a challenge to the pathologist.

CONCLUSION

In conclusion Rosai -Dorfman disease is rare and distinct disorder with benign clinical course. It has a heterogeneous presentation which makes the diagnosis often difficult. FNA represents an efficient, minimally invasive , cost effective reliable techique fo the diagnosis which prevents the patients from undergoing unneccesary. The present study signifies the rare presentations of this disease and age distribution encountered for better diagnosis and thereby avoiding unneccesary surgery.

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Conflicts of interest
There are no conflicts of interest

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