Rosai Dorfman Disease Involving Thyroid- An Fine Needle Aspiration Diagnosis

Ashwini Natekar[1], Jyoty Singh[2], Sweta Agarwal[3]
[1] Chittaranjan NAtional Cancer Institute, Kolkata

ABSTRACT

RDD (Rosai Dorfman Disease) also called as sinus histiocytosis with massive lymphadenopathy is a rare, self-limiting disorder. Extra nodal RDD involving the thyroid gland is very rare and only seven cases have been reported till date in the literature and ours is the second case to be diagnosed by FNAC. RDD shows female predominance. We present a case of RDD involving the thyroid in a 24 year female who presented with midline neck swelling. CT scan neck showed a large lobulated mass involving the right lobe of thyroid gland along with enlarged multiple right level II lymph nodes. FNAC from thyroid and cervical lymph node revealed histiocytes showing emperopolesis,lymphocytes and plasma cells. Hence we conclude that FNAC useful diagnostic procedure for RDD involving thyroid, as it can avoid thyroidectomy.

Key words: RDD, FNAC, Thyroid

Corresponding author address: Ashwini Arvind Natekar, Room no 651, doctors hostel, IPGMER and SSKM hospital, Kolkata WB 700020 M: 8100946471

E-Mail: dr.ashwininatekar@gmail.com

Conflict of interest: No

Case report is Original: YES

Whether case report publishes any where? NO

INTRODUCTION

RDD (Rosai Dorfman Disease) also called as sinus histiocytosis with massive lymphadenopathy was first described by Rosai and Dorfman in 19691. RDD more frequently affects children, adolescents and young man2. The most common presentation is painless cervical lymphadenopathy. Extranodal sites of involvement such as skin, brain, pericardium, scrotum, bone, eyes, upper respiratory tract, breast, heart, thyroid etc have also been described.3-9 Exact etiology and pathogenesis of RDD are not fully understood. Few RDD cases were associated with HIV infection has been mentioned in the literature. Some viruses like EBV (Epstein Barr Virus), Human Herpes Virus 6 (HHV6) have been suggested to be the causative agents of RDD but concrete evidence is still lacking10,11. Till date only seven cases of RDD involving thyroid gland has been reported in the literature to best of our knowledge with ours being the eight case.

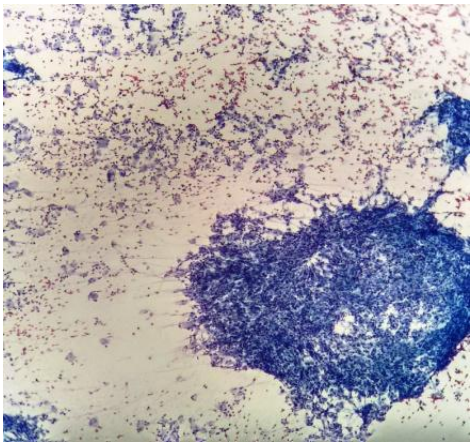
CASE REPORT

We present a case of 24 years old female who presented with midline neck swelling which suddenly increased in size. USG neck showed a SOL in right lobe of thyroid measuring 3.4x1.8mm. CT scan of neck revealed a large lobulated, non-calcified mass lesion measuring 60x46mm involving the right lobe of thyroid, suggestive of neoplastic lesion with few enlarged right sided level II lymph nodes. Hematological investigations revealed hemoglobin of 9.4 gm/dl and ESR-66mm/hr. Following which an FNAC was conducted from thyroid gland and lymph node, which showed mixed population of lymphocytes, plasma cells and histiocytes. The histiocytes showed low N:C, bilobed and trilobed nucleus with abundant cytoplasm with occasional emperopolesis. Hence the diagnosis of Sinus hstiocytosis with massive lymphadenopathy was suggested. Patient was treated with oral corticosteroids and has been showing response to the therapy.

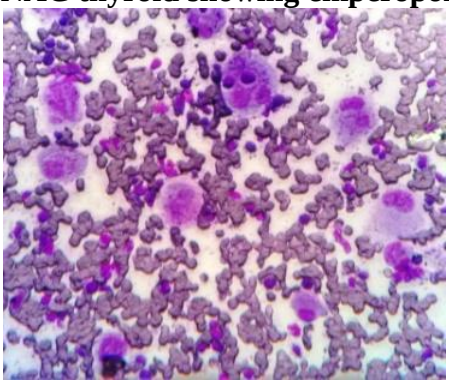
1.midline neck swelling

2. large lobulated mass involving the right lobe of thyroid gland

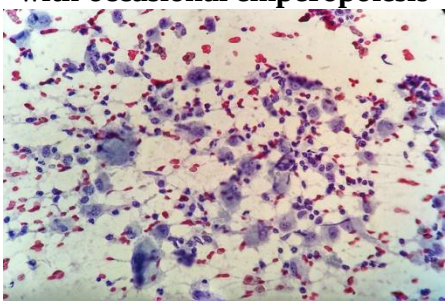
3. FNAC thyroid - histiocytes showing emperopolesis, lymphocytes and plasma cells



4. FNAC thyroid showing emperopolesis



5. The histiocytes show low N:C, bilobed and trilobed nucleus with abundant cytoplasm with occasional emperopolesis



DISCUSSION

RDD is a rare disease involving the lymph nodes. Extranodal RDD is found in 40% of the patients, thyroid involvement has been extremely rare. Only seven cases of RDD involving the thyroid gland have been reported in the literature. These cases were misdiagnosed radiologically as neoplastic in origin as was seen in our case. All the seven reported cases occurred in female with a mean age of 56.3 years.12Ours was a young female who was diagnosed radiologically as neoplastic thyroid lesion. Patient had anemia with increased erythrocyte sedimentation rate. On cytological examination of RDD shows numerous large histiocytes with abundant pale cytoplasm. Emperopolesis has been described as the diagnostic features of RDD, while lymphocytes, plasma cells and neutrophils are characteristically seen in the background.12-15 The histiocytes show positive staining for S-100, CD14, CD11c, CD23, CD33 and CD68 antigens in the cytological smears.

RDD involving the thyroid should be differentiated from undifferentiated carcinoma, Langerhans cell histiocytosis, thyroididts and lymphoma.16,17 In undifferentiated thyroid carcinoma there are highly pleomorphic cells with hyperchromatic nuclei along with neoplastic spindle cells. Lymphoma shows diagnostic Reed Sternberg cells which are absent in RDD. Thyroiditis shows multinucleated giant cells along with reactive thyroid follicular cells. Langerhan cells have grooved nuclei with eosinophils in the background.

CONCLUSION

RDD is a disease treated with corticosteroids; hence diagnosis of RDD by fine needle aspiration can avoid unnecessary surgery. To best of our knowledge only one case of RDD involving the thyroid has been diagnosed by FNAC, thus ours being the second case.

REFERENCES

1. Rosai J, Dorfman RF.Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. Arch Pathol 1969;87(1):63–70.

2. Bernácer-Borja M, Blanco-Rodríguez M, Sanchez-Granados JM, Benitez-Fuentes R, Cazorla-Jimenez A, Rivas-Manga C. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): clinico-pathological study of three cases. Eur J Pediatr 2006;165(8):536–539
3. Zimmerman LE, Hidayat AA, Grantham RL, Chavis RM, Stopak SS, Dreizen NG, et al. Atypical cases of sinus histiocytosis (Rosai-Dorfman disease) with ophthalmological manifestations. Trans. Am. Ophthalmol. Soc. 1988;86:113–135.
4. Lüdemann W, Banan R, Samii A, Koutzoglou M. Di Rocco C. Cerebral Rosai-Dorfman disease. Childs Nerv. Syst. 2015;31:529–532.
5. Lao IW, Dong Y. Wang J. Rosai-Dorfman disease of the pericardium: a case report and review of literature. Int. J. Clin. Exp. Pathol. 2014;7:3408–3412.
6. Cole AJ, Chen C, Lorsbach RB, Honnebier BM, Gardner JM. Shalin SC. Extranodal Rosai-Dorfman disease in the scrotum of a 13-month male: a unique anatomic presentation. Diagn. Cytopathol. 2015;43:40–44.
7. Orvets ND, Mayerson JL. Wakely PE., Jr Extranodal Rosai-Dorfman disease as solitary lesion of the tibia in a 56-year-old woman. Am. J. Orthop. (Belle Mead NJ) 2013;42:420–422.
8. Ioannidis I, Manolakis C, Laurini JA, Roveda KP, de Melo S, Jr AveryB, et al. Rectal extranodal Rosai-Dorfman disease diagnosed by EUS-FNA: a case report and review of the literature. Diagn. Cytopathol. 2015;43:40–44.
9. Vuong V, Moulonguet I, Cordoliani F, Crickx B, Bezier M, Vignon-Pennamen MD, et al. Cutaneous revelation of Rosai-Dorfman disease: 7 cases. Ann. Dermatol. Venereol. 2013;140:83–90.
10. Levine PH, Jahan N, Murari P, Manak M. Jaffe ES. Detection of human herpes virus 6 in tissues involved by sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) J. Infect. Dis. 1992;166:291–295.
11. Tsang WY, Yip TT. Chan JK. The Rosai-Dorfman disease histio- cytes are not infected by Epstein-Barr virus. Histopathology. 1994;25:88–90.
12. Sudhir Kumar Vujhini, Sachin S Kolte, Rahul N Satarkar, and Shastry SrikanthFine needle aspiration diagnosis of Rosai Dorfman Disease involving thyroidJ Cytol. 2012 Jan-Mar; 29(1): 83–85.
13. Powell JG, Goellner JR, Nowak LE, mciver B. Rosai-Dorfman Disease of the thyroid masquerading as anaplastic carcinoma. Thyroid. 2003;13:217–21.
14. Deshpande AH, Nayak S, Munshi MM. Cytology sinus histiocytosis with massive lymphadenopathy. Diagn Cytopathol. 2000;22:181–5.
15. Iyer VK, Handa KK, Sharma MC. Variable extent of emperipolesis in the evolution of Rosai Dorfman disease: Diagnostic and pathogenetic implipications. J Cytol. 2009;26:111–6.
16. Mrad K, Charfi L, Dhouib R, Ghorbel I, Sassi S, Abbes I, et al. Extra-nodal Rosai-Dorfman disease: A case report with thyroid involvement. Ann Pathol. 2004;24:446–9.
17. Kumar B, Karki S, Paudyal P. Diagnosis of sinus histiocytosis with massive lymphadenopathy by fine needle aspiration cytology. Diagn Cytopathol. 2008;36:691–5.