Case Report

A Rare Case of Sino-nasal Rosai Dorfman Disease

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ABSTRACT

Introduction

Rosai-Dorfman disease (RDD) is a very rare nonneoplastic lymphoproliferative disease with unknown etiology and pathogenesis. It was originally described as sinus histiocytosis with massive lymphadenopathy(SHML). Approximately 43% of Rosai-Dorfman disease show extra nodal involvement wherein the possible sites are skin, CNS, ear, orbit and rarely nose and paranasal sinuses. Definitive diagnosis can be performed by histopathological examination, which shows emperipolesis. The sinus histiocytes are strongly reactive for S-100 protein and CD68, but negative for CD1a.

Case Report

The current study presents the case of a 29 year old male patient who had complaints of nasal obstruction since 5 months. Diagnostic Nasal Endoscopy showed mass completely filling both nasal cavities. Histopathological examination concluded the diagnosis of Rosai-Dorfman Disease and Immunohistochemistry (IHC) stained positive for S-100 and CD68, while negative for CD1a, of the surgical specimen after surgical excision by Functional Endoscopic Sinus Surgery. Follow up was done after a week, monthly for 6 months and a year for recurrence.

Discussion

Extranodal RDD, although rarely seen, should be considered in nasal cavity. Even though there is no ideal protocol in the treatment, surgical excision should be considered if there is a functional disorder.

Keywords

Destombes Rosai Dorfman Syndrome; Pathology; Histiocytoses

Rosai-Dorfman disease (RDD) is a rare histiocytic disorder, commonly known as sinus histiocytosis with massive lymphadenopathy (SHML). It was described by Destombes in 1965 and later by Rosai and Dorfman in 1969 as 'sinus histiocytosis with massive lymphadenopathy' and previously classified by the Working Group of the Histiocyte Society of 1987 as a non-Langerhans cell (LC) histiocytosis.[1-3] It is a non-neoplastic diseases with unknown etiology and pathogenesis.

It 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 hypergamma-globulinemia.[4] Most commonly it has seen to affect childrens and adolescents, but any age group can be affected. Although the cervical region is by far the most common and most prominent site of involvement, other peripheral or central lymph node groups can be affected, with or without cervical disease. Extranodal involvement is identified in 43% of the patients diagnosed as RDD[(5)] .The disease albeit prolonged clinical course is benign and spontaneous remission occurs in most cases, with recurrence seen in few cases. It has a variable clinical spectrum changing from spontaneous remission to fatal vital organ involvement.

The definitive diagnosis of the disease is on histopathological examination which shows a significant proliferation of sinus histiocytes and the observation of

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lymphocytes and erythrocytes phagocytized by histiocytes, known as emperipolesis or lympho-cytophagocytosis.[6] Immunohistochemistry would further show sinus histiocytes that are strongly reactive for S-100 protein and CD68, but negative for CD1a.

Case Report

A 29-year-old male presented to the ear, nose and throat clinic with complaints of difficulty in breathing for the past 7 months. It was insidious in onset and progressive. A positive history of mouth breathing, excessive snoring was also elicited. There was no history of trauma or any surgeries. He was given antibiotics and decongestants off and on, which didn't show any relief of symptoms.

On examination, anterior rhinoscopy showed fleshy, firm, vascular mass filling both nasal cavities with mucoid secretions. Diagnostic Nasal Endoscopy was done to confirm the rhinoscopy findings, which showed mass originating from the inferior turbinate and septum completely blocking both cavities (figure 1). Systemic and ENT examination showed no other pathology or any lymphadenopathy. Computed tomography of Paranasal sinuses revealed extensive soft tissue lesion,with involvement of maxillary and ethmoid sinuses and bilateral blockage of ostiomeatal complexes, without any association with skull base (figure 2).

Functional endoscopic sinus surgery (FESS) under general anaesthesia was performed, the mass was debrided out with bilateral maxillary antrostomy and clearance of other sinuses. The specimen was sent for histopathological examination. The procedure was uneventful (figure 3). Post-operatively patient was advised nasal douching and saline drops for 2 weeks.

Histopathological examination of the specimen showed abundant histiocytes with interstial fibrosis & inflammatory infiltrate. Individual histiocytes shows enlarged round to oval hypochromatic nuclei with abundant eosinophilic cytoplasm and emperipolesis of intact lymphocytes (figure 4). There was no evidence of granuloma/ Reed Sternberg like cells/ metastatic carcinoma.

Immunohistochemistry (IHC) stained positive for S-100 and CD68 (emperipolesis is highlighted by S-100; CD68 highlights histiocytes), while immunostain for CD1a is negative. ESR was found to be raised. All the findings were consistent with expected findings in Rosai-Dorfman Disease.

Follow-up visits done after a week, monthly for 6 months and a year displayed no signs of residual or recurrent disease.

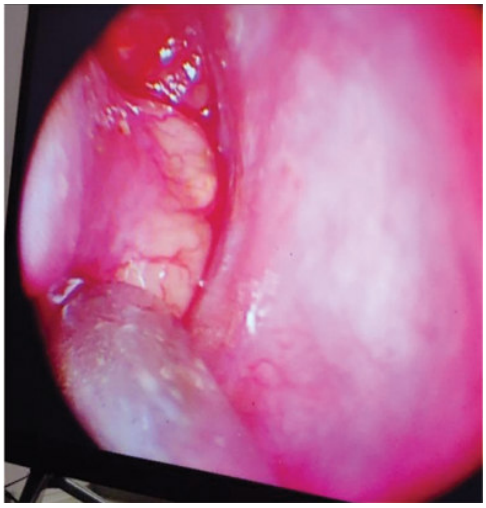

Fig. 1. Diagnostic Nasal Endoscopy showing mass with hypervascularity, arising from the septum

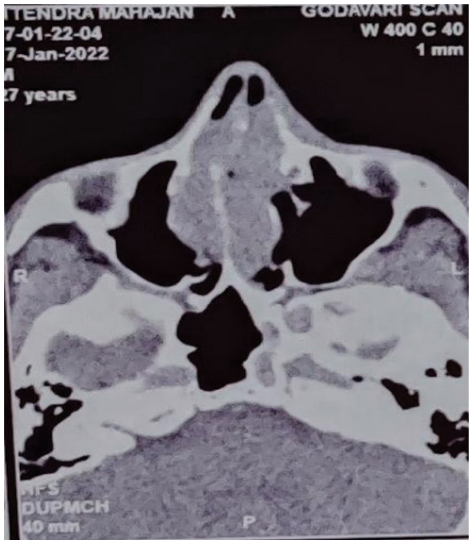

Fig. 2. CT PNS showing mass in both cavities

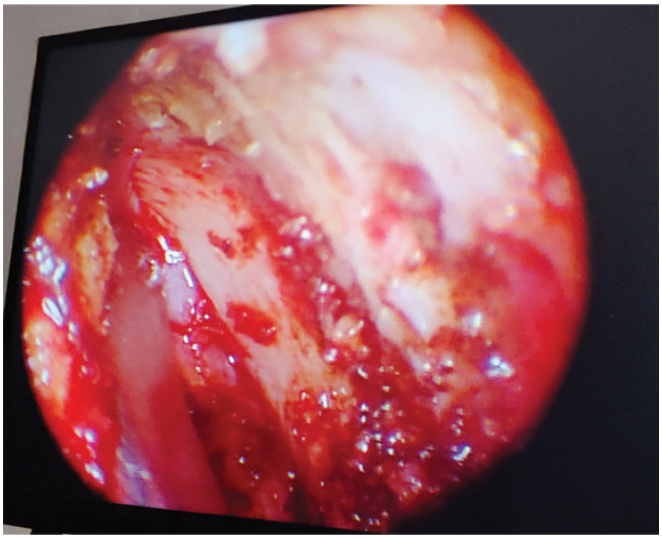

Fig. 3. Intra operative picture showing nasal mass separating from the septum

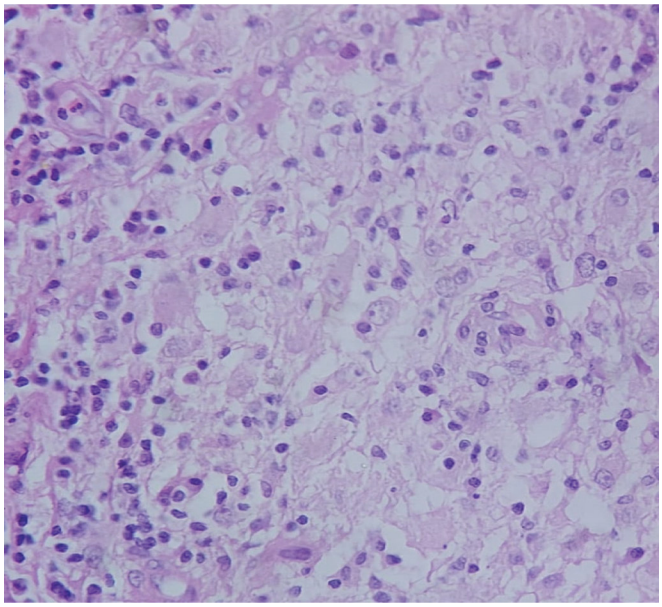

Fig. 4. Section show emperipolesis (H & E *40)

Discussion

Rosai-Dorfman disease (RDD) is a disease with unknown etiology, characterized by histiocytic proliferation of the lymphatic sinuses,[4] which primarily involves the lymphnodes, but can also demonstrate extranodal involvement. In over one-fourth of the cases, RDD involves extranodal sites, this is usually associated with massive lymphadenopathy. However, few cases of are found to be exclusive extranodal manifestations without any lymphadenopathy. The most commonly affected are eyes and occular adnexa, head and neck region, upper respiratory tract, skin and subcutaneous tissue, skeletal system and central nervous system.[4]

Suster et al[7] presented a case of RDD which involved soft tissue based on histopathological findings of emperipolesis and IHC showing S-100 positive histiocytes. Gregor et al[8] reported a case of RDD involving the paranasal sinuses. Leighton and Gallimore[9] described a case of extranodal RDD of subglottis and trachea without any lymphadenopathy.[10]

The histopathologic features of RDD in extranodal sites were found to be similar to nodal disease except fibrosis tends to be more pronounced and lymphocyto-phagocytosis less conspicuous. Microscopically, lymph sinuses are filled with mostly large number of cells of histiocytic appearance with a large vesicular nucleus and abundant clear or lightly eosinophilic cytoplasm that may contain large amounts of neutral lipids. The cytoplasm of these histiocytes have numerous intact lymphocytes and is known as emperipolesis or lymphocytophagocytosis, a constant feature of RDD and has great diagnostic significance. On Immunohistiochemistry, sinus histiocytes show cytoplasmic fat which are strongly reactive for S-100 protein and CD68, but negative for CD1a.[11] The case presented too exhibited the above described characteristic features of RDD with a raised ESR.

The disease usually has a benign course and is self-limited. However, it may be aggressive in a very few number of cases and might be fatal.[12] The ideal type of treatment regime is not yet determined since the disease is very rare and spontaneous remission is possible. It is relatively unaffected by therapy, although steroids, radiation, chemotherapy, interferon, acyclovir, monoclonal antibody treatment, and thalidomide have been used.[13]

The disease is treated surgically when a life-threatening enlargement of the lymph nodes or situations that cause functional impairment occur.[14] In our case, the mass completely filled both nasal cavities and caused

difficulty in breathing, hence was surgically removed by functional endoscopic sinus surgery.

Conclusion

Rosai-Dorfman disease is a benign condition usually presenting with massive bilateral cervical lymphadenopathy. Extra-nodal involvement, though rare can be seen, with or without any lymphadenopathy.

Involvement of Paranasal sinuses and septum without any nodal presentation as presented in our case is rare. The diagnosis is primarily based on Histopathological examination mainly eliciting emperipolesis and Immunohistochemistry showing positivity for S-100 and CD68 while being CD1a negative.

Even though there is no ideal protocol, surgical excision by functional endosopic sinus surgery should be considered if there is a functional disorder.

References

1. Destombes P. [Adenitis with lipid excess, in children or young adults, seen in the Antilles and in Mali. (4 cases)]. Bull Soc Pathol Exot Filiales 1965;58:1169–75
2. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. Arch Pathol 1969;87:63–70
3. Writing Group of the Histiocyte Society. Histiocytosis syndromes in children. Lancet 1987;1:208–9
4. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: a pseudolymphomatous benign disorder. Analysis of 34 cases. Cancer. 1972;30(5):1174-1188
5. S. Gaitonde, "Multifocal, extranodal sinus histiocytosis with massive lymphadenopathy: an overview," Archives of Pathology and Laboratory Medicine, vol. 131, no. 7, pp. 1117–1121, 2007
6. G. Pettinato, J. C. Manivel, E. S. G. d'Amore, and G. Petrella, "Fine needle aspiration cytology and immunocytochemical characterization of the histiocytes in sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman syndrome)," Acta Cytologica, vol. 34, no. 6, pp. 771–777, 1990
7. Suster S, Cartagena N, Cabello IB, Robinson MJ. Histiocytosis lymphophagocytic parmiculitis; an unusual extranodal presentation of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). Arch Dermatol 1988;124:1246-9
8. Gregor RT, Ninnin D. Rosai-Dorfman disease of the paranasal sinuses. J Laryngol Otol 1994;108:152-5
9. Leighton SE, Gallimore AP. Extranodal sinus histiocytosis with massive lymphadenopathy affecting subglottis and trachea. Histopathology 1994;24:393-4
10. Asrar L, Facharzt EH, Haque I. Rosai-Dorfman disease presenting as isolated extranodal involvement of larynx. J Otolaryngol 1998;27:85-6
11. Miettinen M, Paljakka P, Haveri P, Saxen E. Sinus histiocytosis with massive lymphadenopathy. A nodal and extranodal proliferation of S-100 protein positive histiocytes? Am J Clin Pathol. 1987;88(3):270-277
12. R. S. Cocker, J. Kang, and L. B. Kahn, "Report of a case presenting as a midline thyroid mass," Archives of Pathology & Laboratory Medicine, vol. 127, pp. 197–200, 2003
13. O. F. Unal, E. G. Koc 'an, A. Sungur, and S. Kaya, "Rosai-Dorfman disease with multi-organ involvement in head and neck region," International Journal of Pediatric Otorhino-laryngology, vol. 68, no. 5, pp. 581–584, 2004
14. A. Pulsoni, G. Anghel, P. Falcucci et al., "Treatment of sinus histiocytosis with massive lymphadenopathy (rosai-dorfman disease): report of a case and literature review," The American Journal of Hematology, vol. 69, no. 1, pp. 67–71, 2002.