*Journal of Pediatric Disease* (2021) Volume
4 doi: 10.24294/jpd.v4i1.124

REVIEW ARTICLE

# Sinus histiocytosis with massive lymphadenopathy

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## ABSTRACT

Sinus histiocytosis with massive lymphadenopathy is a benign lymphoproliferative disorder.In 1969, Rosai and Dorfman[1] made detailed research on it, so it was also called Rosai DorfnlRn disease (rosai.dorfmandisease, RDD).The clinical manifestations were fever, neck lymph node enlargement, leukocytosis and high gamma globulin.Histopathological findings showed that lymph node involvement was present in group RDD, and the infiltration of the cells was predominant, especially the phagocytosis of the histiocytic cells.About 43% of RDD patients have lymph node involvement in[2], in which the skin is the most common extranodal organs involved.About 10% of patients with skin damage, skin rash and morphological diversity, is easy to be misdiagnosed.In this paper, through the analysis of a case of RDD and EB virus infection, in clinical in patients with special infection can be early detection and treatment.

***Keywords***: *Sinus； histiocytosis ； lymphadenopathy*

## Case report

Children, female, 3 years old, children with recurrent fever for 20 days, no cough and asthma, no vomiting and diarrhea.The pharyngeal hyperemia, no obvious oral mucosa and conjunctival congestion, bilateral cervical lymph nodes obvious swelling, no tenderness.Blood routine: WBC:19.83 x 109/L N:0.81 L:0.12 M:0.05, RBC:4.7 x 1012/L; HGB:119g/L PLT:436 x 109/L,ESR 47mm/h; CRP36mg/L.Mycoplasma pneumoniae antibody, liver and kidney function ions, myocardial enzymes, blood culture, tuberculosis series, rheumatoid factor, EB virus, ASO, immunoglobulin were normal. Ultrasound showed: cervical lymph nodes multiple bilateral neck, left about 3.1 * 27mm, 3.4 * 26mm on the right side of the chest and abdomen, CT showed bilateral supraclavicular fossa, mediastinal and retroperitoneal multiple nodules, consider the lymph nodes. The initial anti infection treatment was ineffective, and then 6.3 and 4.2 x 106 were cultured again in the EB throat swab, and 1 cases of cervical lymph nodes were taken out to exclude malignant lymphoma.

Pathological examination:

Appearance: a lymph node of 3 x 2.5 x 1.6cm, the surface is roughly smooth, one side has peeling marks, slightly rough, cut section of gray red, delicate, crisp, fish shape.

Under the microscope: low magnification lesions showed indifferent area and anachromasis structure zone alternate distribution, high magnification, indifferent area into a piece of tissue cells, cells, oval or polygonal, the size of small lymphocytes more than 10 times, abundant cytoplasm, lightly eosinophilic or fine granular, see the swallow a plurality of intact lymphocytes and a few plasma cells in the cytoplasm, nucleus round or oval nuclei, prominent nucleoli and mitotic rare; dark staining area for more lymphocytes, plasma cells and a small amount of eosinophils, scattered in lymphoid follicles; infiltration of inflammatory cells may be associated with many different areas the fibrosis, fibrous tissue around the tissue cells showed nodular image.

Chemical staining: tissue S-100 (+), SMA (-).

Corrected diagnosis: sinus cell hyperplasia with giant lymphadenopathy.

Intravenous drip of ganciclovir 5 mg /kg/ times, 2 times / day, oral prednisone 1 mg /kg/ day, treatment for 5 days, the body temperature gradually dropped to normal, maintenance treatment for 9 days, EB virus culture turned negative, blood routine, ESR, CRP normal discharge. In March after the follow-up, neck lymph node with the front position, size is reduced and left about 16 * 14mm, the right side of 27 * 13mm, degree of movement.


doi: 10.24294/jpd.v4i1.124

## Discussion

Sinus histiocytosis with massive lymphadenopathy (SHML) is a rare benign lymphoproliferative disease, most people think that SHML is mainly due to antigen presenting cell proliferation induced by monocyte macrophage system and bone marrow, the specific etiology and pathogenesis has not been fully elucidated, and individual genetic susceptibility, infection, immune imbalance and dysfunction of the 3. Infection may be associated with human herpesvirus 6 (HHV -6), HHV- 8, EB virus, human papilloma virus and cytomegalovirus infection[4-5]. The patients later detected by EB virus, antiviral and immune suppression in the treatment of disease control, nonspecific inflammatory disease related indicators of negative, suggesting that Rosai-Dorfman may occur and EB virus infection, but too few cases, nonspecific clinical manifestation and the disease is easily missed and misdiagnosed, future in clinical practice in further exploring and summing up.

## Summary points

Sinus cell hyperplasia with giant lymph node disease (SHML), also known as Rosai-Dorfman's disease (RDD), is a special clinical and pathological characteristics of idiopathic histiocytic proliferative diseases. It can be seen in all age groups, from infants to the elderly can occur, but better in children and adolescents, it has been reported that the predilection age of RDD is 0~20 years old, 62% of which are less than 10 years old[6]. The main clinical features are more common in children and young adults; showed bilateral painless cervical lymph node enlargement accompanied by fever, neutrophils increased, erythrocyte sedimentation rate, hypergammaglobulinemia, more than 92% of the SHML patients with painless cervical lymph node enlargement as the first symptom, lymph node hard mutual confluence,

Other lymph nodes could be involved, such as axillary, inguinal and mediastinal lymph nodes, accounting for 87.3%, 25.7%, 23.7%, respectively. Nearly 30%SHML patients have extranodal organ involvement, skin and meninges are the main sites of extranodal, only 5%SHML patients simply involve extranodal organs[7]. Its pathological features are: the whole dermis visible foam cells formation of nodular or diffuse infiltration; cytoplasmic cells seen in intact lymphocytes and plasma cells or neutrophils, called into motion, is useful in the diagnosis of the lymphocyte emperipolesis.

In immunohistochemistry, S-100 protein, CD68 and vimentin were expressed in tissue cells, but no CD1, SMA and CD34 were expressed. There were no Birbeek particles under electron microscope. The distribution characteristics of nodular distribution of tissue cells and inflammatory cells infiltration in dense zone formation of deep and shallow phase at low magnification is an important clue in the diagnosis of pathology doctor SHML, at high magnification to determine olistherozone for hyperplasia of large tissue cells and often accompanied by inflammatory cells, especially lymphocytes into the phenomenon, you can consider the diagnosis of Rosai-Dorfman disease. Immunohistochemistry S-100 protein positive and CD1 alpha negative are helpful for the diagnosis.

Sinus histiocytosis with massive lymphadenopathy is a disease, not cancer, clinicians should be paid attention to and lymphoma, metastatic malignant melanoma, infectious granuloma, Hans Langerhans cell histiocytosis, tuberculosis, histiocytic necrotizing lymphadenitis and phase identification. SHML has a better prognosis, mostly self limiting, several months to years to resolve, lasting a few cases of disease, disease and great important organ involvement, deteriorated or transformed into lymphoma, or accompanied by other immune diseases and infection and death. At present, RDD has not yet formulated the corresponding treatment guidelines, treatment methods are diverse, including surgery, corticosteroids, radiotherapy and chemotherapy, such as[8]. If the lesion is limited, such as a single nodular lesion area, surgical resection is the preferred treatment, there are a few recurrence after surgery, but most cases can obtain a long-term remission[9,10]. The lesions are extensive or involved in important organs or systems, and the first choice is corticosteroids, which has significant curative effect on SHML which only invades lymph nodes. If the extra organs are involved, adjuvant radiotherapy and chemotherapy are needed.

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