Rosai Dorfman Disease in an Isolated Cervical Node

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ABSTRACT

Lymphadenopathy associated with sinus histiocytosis is known as Rosai Dorfman Disease. Classically it presents as massive bilateral painless cervical lymphadenopathy.

We report a unique case of this disease, in which there was involvement of only a single cervical node. The diagnosis was confirmed by documenting the classical cyto-morphological features including emperipolesis.

Our case adds a new dimension to the current spectrum of Rosai Dorfman disease and illustrates that an isolated lymphadenopathy can also occur in this disease.

Keywords: Rosai Dorfman Disease; Emperipolesis; Lympadenopathy; Sinus histiocytosis

INTRODUCTION

Sinus histocytosis with massive lymphadenopathy, also known as Rosai-Dorfman Disease (RDD), is an idiopathic proliferation of histiocytes within the lymph nodes [1]. RDD generally manifests in children or young adults with massive cervical lymphadenopathy, fever, leukocytosis, and hyper-gammaglobulinemia. However, patients presenting with isolated intracranial disease tend to be older [2]. The disease is more common in males and in individuals of African descent [3]. Other lymphatic groups, such as mediastinal, axillary and inguinal lymph nodes can also be affected. Extranodal sites are affected in about 25 to 40% of cases. The concomitant involvement of one or more sites in the same individual is observed in up to 44.7% of cases [3,4].

A cytokine-mediated migration of monocytes may be involved in the accumulation and activation of histiocytes. This functional activation could be triggered by many different stimuli as RDD can coexist with autoimmune diseases, hematological malignancies and post-infectious conditions. In fact, many viruses like human herpes virus 6 (HHV-6) and Epstein-Barr Virus (EBV), have been implicated as potential causative agents [4,5].

CASE REPORT

A six-year old female child presented to our institute with a history of low-grade fever associated with enlargement of upper cervical lymph node for the last three months. Examination revealed a solitary enlarged lymph node (Left Jugulo-digastric) 3 X 3 cm, firm in consistency, discrete and freely mobile. Routine investigations showed hemoglobin of 10 g/dl, total leucocyte count of 16,000/mm^3 with neutrophilia and erythrocyte sedimentation rate of 40 mm at the end of one hour by Westergrens method. Peripheral blood film showed microcytic, hypochromic red blood cells. Chest X-ray and ultrasonography of the whole abdomen were normal.

Fine needle aspiration cytology (FNAC) of lymph node revealed presence of diffusely distributed histiocytes throughout the smears (Figure 1). These histiocytes had abundant pale cytoplasm with single or multiple nuclei with inconspicuous nucleoli. The nuclei did not show any atypia or lobation or longitudinal grooving.

The cytoplasm of these histiocytes exhibited numerous intact lymphocytes i.e emperipolesis (Figure 2). In some histiocytes, the phagocytosed cells were so numerous that they obscured the nucleus. The background had mature lymphocytes, plasma cells, neutrophils and macrophages. Based on this characteristic cyto-morphology, a diagnosis of RDD was made. The patient was treated with oral prednisolone 10 mg/day for a period of 4 weeks, which was then tapered off. She is doing well on regular follow-up.

DISCUSSION

In the year 1969, Rosai and Dorfman described 4 cases of a disease they called sinus histiocytosis with massive lymphadenopathy [1]. Later in 1972, they analyzed 30 additional cases, establishing it as a clinico-pathologic entity [2]. In its classic form, the disease presents in children and adolescents with massive painless bilateral cervical lymphadenopathy, often with associated fever and mild anemia [3]. Extra-nodal RDD has also been reported either alone or in association with lymphadenopathy. The most common extra-nodal sites include the skin, orbit, and upper respiratory tract [4]. The incidence of RDD has been estimated at 100 cases/year in the United States [6]. More than four-fifths of reported cases occur during the first and second decades of life with a 2:1 male-to-female ratio [3]. Our patient reported the occurrence of the disease during the first decade of life and was a female. Our patient was unique in the sense that she had only a solitary enlarged cervical lymph node, which has not been reported previously. Histologically, lymph nodes show pericapsular fibrosis and dilated sinuses that are heavily infiltrated with large histiocytes, lymphocytes and plasma cells. The presence of emperipolesis, or the engulfment of lymphocytes and erythrocytes by histiocytes that express S-100, is considered diagnostic of RDD [6,7]. These cells are also positive for CD68, CD163, α_1-antichymotrypsin, α_1-antitrypsin, fascin and HAM-56 while CD1a is typically negative. IL-1β and TNF-α is strongly expressed by these cells. Ultrastructurally, histiocytes in RDD lack Birbeck granules and viral particles [8].

The prognosis is excellent in most cases [9]. Complete spontaneous regression is known to occur. The course of disease however may be protracted. Complications are mostly due to pressure effects. No standard treatment for RDD is known. The treatment modalities for RDD are nonspecific and include corticosteroids, chemotherapy with a combination of vinca alkaloids and alkylating agents, low dose interferon, antibiotics therapy, radiation therapy and surgical treatment with partial or total resection [6,9,10]. Our patient responded well to low dose oral corticosteroids.

Figure 1: Giemsa staining showing diffusely distributed histiocytes in the background of mature lymphocytes, plasma cells, neutrophils and macrophages

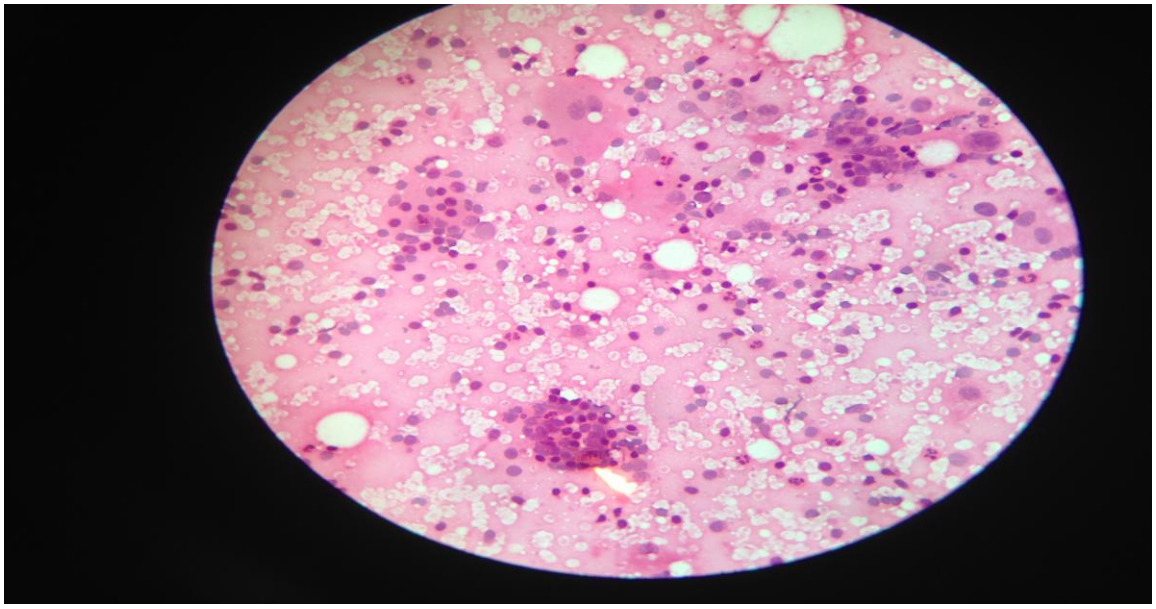
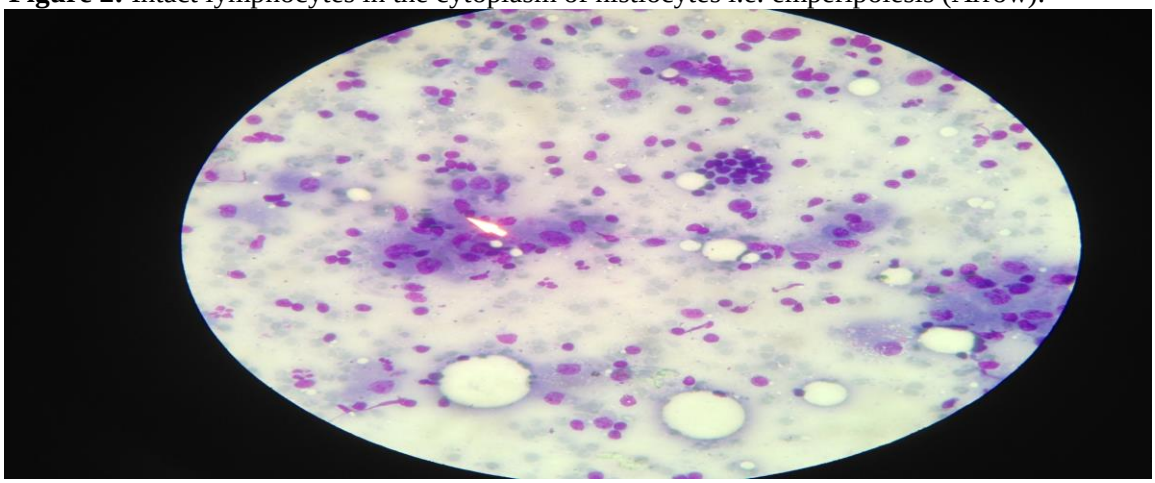


Figure 2: Intact lymphocytes in the cytoplasm of histiocytes i.e. emperipolesis (Arrow).



CONCLUSION

Although, massive cervical lymphadenopathy with enlargement of many nodes is the hallmark of RDD, but as illustrated by our case, it can also occur in solitary nodal enlargement. The diagnosis can be established by FNAC and it responds well to corticosteroids.

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