Rosai-Dorfman Disease with Extranodal Lesions
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Abstract

Objectives: Rosai-Dorfman disease is a rare condition of marrow hemopoietic stem-cell origin. Most patients present with lymphatic involvement, whereas less than one third of cases are extranodal presentations. Only about 5% of the extranodal cases involve bone. The purpose of this study is to present a case of Rosai-Dorfman disease with lymphadenopathy, intracranial tumors and bone lesions mimicking malignant neoplasm.

Methods: Case report concerning the diagnosis and treatment of Rosai-Dorfman disease with extranodal presentation including intracranial tumors and multifocal bone lesions.

Results: A 53 year-old man was admitted to our hospital because of three-month history of bilateral cervical lymphadenopathy. He also had a five-year history of dizziness. Computed tomography, magnetic resonance imaging and positron emission tomography showed cervical, mediastinal and inguinal lymphadenopathy. Intracranial tumors and pelvic bone lesions were also detected. The cervical lymph node biopsy confirmed the diagnosis of Rosai-Dorfman disease. The patient received oral prednisolone, and his neck pain was improved. The massive cervical lymph nodes and bone lesions didn't progress.

Conclusion: Rosai-Dorfman disease should be considered in the differential diagnosis of lymphadenopathy, even though the disease is rare. The predominant clinical manifestation of the disease is painless cervical lymphadenopathy. Rosai-Dorfman disease is considered a benign disease, however the disease can be fatal because of cellular infiltration and mass forming. The treatment for Rosai-Dorfman disease has not been established.

Introduction

Rosai-Dorfman disease is a rare, benign, idiopathic, and non-neoplastic histiocytic disorder. It's characterized by massive but painless cervical lymphadenopathy, fever, leukocytosis, increased erythrocyte sedimentation rate, and polyclonal hypergammaglobulinemia [1]. Most patients present with lymphatic involvement, whereas 43% of cases are extranodal presentations: eyelids, orbit, respiratory tract, salivary glands, skin, bone, testis, lung, kidney, central nervous system, thyroid and gastrointestinal tract [2].

Case Report

A 53 year- old man was admitted to our hospital because of three-month history of bilateral massive painless cervical lymphadenopathy and five-year history of dizziness. On physical examination, he had enlarged bilateral cervical lymph nodes. He had normal vital signs and no neurological abnormality. Hematological and biochemical examinations were normal without leukocytosis and hypergammaglobulinemia. Computed tomography, magnetic resonance imaging and positron emission tomography showed cervical, mediastinal and inguinal lymphadenopathy. Intracranial tumors and multiple lytic bone lesions of the skull and pelvic bone were also detected (Figure 1-3). The patient underwent cervical lymph node biopsy and intracranial tumor resection. The histological examinations of the cervical lymph node biopsy showed atypical histiocytes engulfing multiple lymphocytes (Figure 4A). The immunohistochemical staining revealed positive S-100 staining of histiocytes (Figure 4B). Histopathological and immunohistochemical studies established the diagnosis of Rosai-Dorfman disease. The neck pain appeared, and the oral administration of prednisolone improved his symptoms. No evidence of residual or recurrent tumor in the intracranial lesion was identified. The size of bone lesions and lymph nodes were stable.

Discussion

Sinus histiocytosis with massive lymphadenopathy, also known as Rosai-Dorfman disease, is an idiopathic histiocytic proliferation affecting lymph nodes. Histologically, the lesions consist of variable numbers of pale-staining histiocytes with emperipolesis. In this case, the patient had multiple lymphadenopathy, central nervous system involvements and bone lesions. The differential diagnosis of Rosai-Dorfman disease is broad and includes malignant glioma, malignant lymphoma, histiocytosis X (Langerhans cell histiocytosis), sarcoidosis, metastatic tumors, granulomatous disease and meningioma [3][4]. Rosai-Dorfman disease has a long natural history with mostly benign course, and special treatment is not required. However, massive lymphadenopathy, extranodal involvement of multiple organs (kidney, lungs and liver), immunologic abnormalities and anemia can lead to a poor prognosis. The treatment modalities for Rosai-Dorfman disease include corticosteroids, chemotherapy with a combination of vinca alkaloids and alkylating agents, low-dose interferon, antibiotic therapy, radiation therapy and surgical treatment [5]. However, there is no established treatment for Rosai-Dorfman disease.

Conclusions

Rosai-Dorfman disease is considered a benign disease, however the disease can be fatal because of combination of cellular infiltration and mass forming. Very careful management is required.

References

1) Rosai J. Dorfman RF. Arch Pathol. 1969 Jan;87(1):63-70.