Rosai-Dorfman disease mimicking mediastinal lymphoma

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Abstract Rosai-Dorfman disease is rare and typically presents with cervical lymphadenopathy. The disease is generally indolent and self-limited, but it carries a poor or fatal prognosis when it is advanced or when it involves and compresses vital structures. We herein report a rare case of Rosai-Dorfman disease affecting only the mediastinal-hilar region in a 66-year-old woman.

Key words Rosai-Dorfman disease · Lymphadenopathy · Mediastinal lymph nodes

Introduction

Rosai-Dorfman disease (RDD) is a benign histiocytic proliferative disease, with approximately 600 reported cases. RDD affecting only the mediastinohilar region is rare. Here, a case of RDD that was histopathologically diagnosed in a Japanese woman with chief complaints of bilateral scleritis, fever, and weight loss, who underwent thoracoscopic biopsy of the mediastinal lymph nodes, is reported.

Case

A 66-year-old woman visited another hospital on March 2007, complaining of conjunctival congestion, low-grade fever, and weight loss. The symptoms were diagnosed as bilateral scleritis. Subsequently, blood tests showed persistent elevation of C-reactive protein (CRP) (about 1–3 mg/dl), as a result of which detailed tests were performed. Computed tomography (CT) showed enlargement of mediastinal-hilar lymph nodes, and positron emission tomography (PET) showed high fluorodeoxyglucose (FDG) accumulation at the same sites. Thoracoscopic biopsy of the mediastinal lymph nodes was scheduled for a definitive diagnosis, and the patient was referred to our institution in July 2007.

The low-grade fever was not present when she was admitted to our hospital. Superficial lymph nodes could not be palpated, and there was neither hepatosplenomegaly nor rash.

Laboratory data showed slight CRP elevation, but lactate dehydrogenase, soluble interleukin-2 receptor, and angiotensin-converting enzyme were not elevated. The values for other tumor markers were also normal. However, the antinuclear antibody assay was weakly positive.

Chest radiography showed slightly mediastinal enlargement and no abnormal shadows in the lung fields. CT showed enlargement of mediastinal-hilar lymph nodes (Fig. 1). There were no intrapulmonary lesions or enlarged lymph nodes in the supraclavicular region. FDG accumulation was seen at the same sites as previously.

Thoracoscopic biopsy of the mediastinal lymph nodes was performed for a definitive diagnosis on March 2008. The mediastinal-hilar lymph nodes appeared to be
wrapped around the bilateral main bronchi from the lower trachea. They were enlarged and formed a mass that showed no clear borders with the surrounding tissues. The anterior mediastinal lymph node was isolated and enlarged, but it showed no tendency to infiltrate the surrounding tissue. It was removed. The tracheal bifurcation lymph node was subjected to incisional biopsy.

Histological examination showed disruption of the lymphoid follicles, proliferation of large histiocytes that had clear, eosinophilic cytoplasm, and marked dilation of the lymphatic sinuses. Emperipolesis was seen in the cytoplasm of the histiocytes, with embedding of morphologically intact lymphocytes and erythrocytes (Fig. 2). Immunostaining showed the proliferating histiocytes to be positive for S-100 protein. A pathological diagnosis of RDD was made on the basis of the above histological and immunochemical staining results.

The postoperative course was good, and the patient was discharged on the 4th postoperative day. The patient received no treatment after surgery but remained under observation. Two years later, imaging revealed that the lesion showed no change.

Discussion

Rosai-Dorfman disease was first reported as a disease entity by the pathologists Rosai and Dorfman in 1969. RDD is a nonneoplastic, benign, proliferative disorder of histiocytes. There have been reports that RDD is associated with an immune disorder, Epstein-Barr virus, and human herpesvirus 6. The incidence of RDD is low. Many aspects regarding the appearance of histiocytes have yet to be elucidated, and the etiology of the disease is unclear.

The age at onset of RDD ranges from childhood to young adulthood (mean age 20.6 years). Most reports of RDD have originated in the United States (39%), Europe (20%), and Africa (15%); that is, it appears primarily in Caucasians and Blacks. Few cases have been reported in Asians.

Various clinical symptoms are associated with RDD. Painless cervical lymphadenopathy is frequently seen, sometimes accompanied by generalized symptoms such as fever, sore throat, night sweats, fatigue, and weight loss. More than 90% of affected lymph nodes are cervical lymph nodes, although axillary, mediastinal, and inguinal node involvement has been reported. On rare occasions, the lymph nodes enlarge into large masses that can press on contiguous organs. In addition, leukocytosis, anemia, an elevated erythrocyte sedimentation rate, hypergammaglobulinemia, and other findings of chronic inflammation are sometimes observed. Extranodal lesions are seen in approximately 40% of cases, with reports documenting almost all possible body sites.

Diseases that are characterized by abnormal proliferation of histiocytes, which is a collective name for immune cells other than lymphocytes, are referred to