



Rosai-Dorfman Disease with Involvement of the Lungs

Rosai-Dorfman Hastalığında Akciğer Tutulumu

To the Editor,

An 11-year-old boy presented with a 2-month history of painless bilateral cervical masses, low-grade fever, and weight loss. Biopsies of the cervical lymph node revealed diffuse lymphoplasmacytic infiltration, foamy histiocytes, and emperipolesis. Immunohistochemistry was performed and was found to be positive for S-100 protein and CD68, and negative for CD1a and langerin. It was diagnosed as Rosai-Dorfman disease, and corticosteroid therapy was initiated. During 6 months of follow-up, the lymph node swelling regressed, while the dry cough continued, but with less frequency. A chest computed tomography image showed interstitial pneumonitis with pulmonary nodular lesions (Figure 1). A surgical biopsy of the lung nodules was performed. Histopathology of a lung biopsy specimen revealed a polymorphous infiltrate of lymphocytes, plasma cells, and scattered multinucleated foamy histiocytes. There was a suggestion of emperipolesis. Immunohistochemistry results were positive for CD68 and S100 and negative for CD1a (Figure 2). The small lymphocytes engulfed by histiocytes included T cells. Special staining was performed, including Ziehl-Nielsen and Periodic acid-Schiff stains, which indicated no sign of acid-fast bacilli or fungi. These findings were indicative for the histopathologic diagnosis of RDD with lung involvement.



Figure 1: Chest computed tomography image showing interstitial pneumonitis with pulmonary nodular lesions

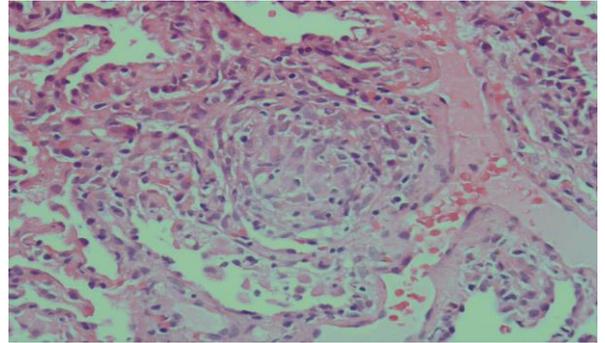


Figure 2: Histopathology of lung biopsy revealed a polymorphous infiltrate of lymphocytes, plasma cells, and scattered multinucleated foamy histiocytes (IHC, x400)

RDD with lung involvement is quite rare and to make a definitive diagnosis is a challenge. Only 2% of cases have lower respiratory tract involvement. Hilar or mediastinal lymphadenopathy, pulmonary nodules or masses, and on rare occasions, pleural effusion, interstitial lung disease, or central airway involvement might be detected intrathoracically (1,2).

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CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

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