IMAGES

Isolated Intravascular Large B-cell Lymphoma Presenting with Diffuse Ground Glass Pulmonary Infiltrates

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Abstract A case of 44-year-old man with dyspnea and CT chest demonstrated bilateral infiltration. Patient was failed to improve with antibiotics and steroid. Finally, video-assisted thoracic surgery-guided lung biopsy was performed and surprisingly revealed intravascular large B cell lymphoma. The patient was subsequently started on chemotherapy with considerable clinical improvement. At the time of last follow-up (4 years from diagnosis); there was no clinical or radiologic evidence of tumor recurrence.

Keywords Intravascular large B-cell lymphoma · Video-assisted thoracic surgery · Pulmonary

Image in Hematology

A 44-year-old man, nonsmoker presented to our institution with the chief complaint of dyspnea on exertion, cough, fever, and myalgias over the preceding 1 month in conjunction with a pruritic skin rash. Physical examination was remarkable for conjunctival pallor and a papular rash over his neck and extremities. Laboratory studies were significant for hemoglobin of 10.5 g/dL (reference range 13.5–17.5) and a lactate dehydrogenase 1000 U/L (reference range 122–222). A comprehensive metabolic panel, erythrocyte sedimentation rate and C-reactive protein were all unremarkable. HIV screening, fungal serologies, anti-neutrophil cytoplasmic antibodies, antinuclear antibodies were all negative. Computerized tomography (CT) of chest

and abdomen revealed diffuse hazy ground glass infiltrates throughout both lung fields (Fig. 1) and moderate splenomegaly. Initial differential included infection, vasculitis process or malignancy. Bronchoscopy and bronchoalveolar lavage were negative for infectious pathogens and cytology was negative for malignancy. In addition, a bone-marrow biopsy demonstrated no evidence of malignancy. A skin biopsy revealed a perivascular lymphocytic infiltrate with scattered vessel dilatation extending into the mid and deep reticular dermis consistent with pressure urticaria. Empiric treatment with antibiotics and steroids were initiated without significantly clinical improvement and therefore a video-assisted thoracic surgery (VATS)-guided lung biopsy was performed. The lung biopsy surprisingly revealed large abnormal lymphoma cells within the capillaries (Fig. 2a) and immunohistochemistry was positive for CD20 (Fig. 2b) and negative for CD3. The overall picture was compatible with intravascular large B cell lymphoma

Fig. 1 Computerized tomography (CT) of chest revealed diffuse hazy ground glass infiltrates throughout the lungs bilaterally

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Fig. 2 **a** Lung biopsy hematoxylin–eosin stain showing large tumor cells with prominent nucleoli and mitosis infiltrate in small vessels. **b** Immunohistochemical stain (bottom) showing CD20+ tumor cells in lumina of small capillaries

(IVLBCL). Anemia and splenomegaly were thought to be secondary from hemolytic process secondary to intravascular lymphoma. The patient was subsequently started on cyclophosphamide, doxorubicin, vincristine, and prednisone and completed eight cycles without clinical or radiologic signs from whole body positron emission tomography–computed tomography (PET-CT) of tumor recurrence or metastasis.

IVLBCL is an extremely rare malignancy with an approximate incidence of less than one per million per year [1]. In 2008, the World Health Organization (WHO) classified IVLBCL as a type of extranodal large B-cell lymphoma. IVLBCL is characterized by proliferation of lymphoma cells within the lumen of small blood vessels, especially capillaries [1, 2]. More than 90 % of cases present as clinical stage III or stage IV disease and can be rapidly fatal if diagnosis and treatment are delayed. However, current chemotherapy regimens including rituximab have resulted in improved survival with the possibility of prolonged remission as was seen in our case [1]. Diagnosis of IVLBCL can be challenging given its nonspecific presentation and possibility for involvement of virtually any organ given its hematogenous nature [2–4]. Fever, night sweats, fatigue and weight loss are the most common constitutional symptoms in addition to organ specific symptoms which are a result of intravascular obstruction and ischemia [5]. Anemia, leukopenia, thrombocytopenia and elevated levels of CRP, LDH, ferritin, IL-2R and β 2 microglobulin are often noted. Patients with pulmonary involvement can present with cough, dyspnea, hypoxemia and pulmonary hypertension with chest CT showing focal nodules, diffuse ground glass infiltrates or consolidation

[1]. A biopsy is mandatory for the diagnosis and shows large malignant lymphoid cells (CD 20 positive, CD3 negative) in small vessels and capillaries with prominent nucleoli, frequent mitotic figures and scant cytoplasm [1].

In summary, isolated pulmonary involvement is a rare presentation of IVLBCL and can present with dyspnea and non-specific ground-glass infiltrates on CT Chest.

Compliance with Ethical Standards

Conflict of interest All of the authors have no conflict of interest.

Ethical Approval This article does not contain any studies with human participants or animals performed by any of the authors.

Informed Consent Informed consent was obtained from patient.

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