A rare presentation of a rare disease

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Background Castleman's disease, a rare condition of uncertain etiology, is associated with lymphoproliferation. It is histologically and prognostically distinct from malignant lymph node hyperplasia.

Case presentation We report a case of a female patient who presented with interstitial lung disease and mediastinal lymphadenopathy, not responding to usual treatment.

Conclusion Definitive histological diagnosis in patients with lymphadenopathic presentation associated with systemic symptoms is important to differentiate Castleman's disease from malignant lymphoma.

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Case presentation

A 33-year-old woman, nonsmoker was referred to respiratory clinic by mild exertional dyspnea, dry cough, and fever for 2 weeks. She works as a secretary, with no significant medical, drug, or exposure history. She is married and has one child.

Physical examination revealed a cyanosed female with oxygen saturation on room air 78% at rest, and the patient was put on nasal prong 6 l/min, with oxygen saturation 93%, pulse rate 128/min, and temperature 38.5°C. Her general examination was unremarkable with no peripheral lymphadenopathy. Her chest examination revealed fine leathery crepitations all over the chest more on lung bases.

Chest radiography was performed followed by computed tomography of chest (Fig. 1).

Laboratory work-up showed normocytic normochromic anemia with hemoglobin (7 mg/dl), erythrocyte sedimentation rate (150), C-reactive protein (25), white blood cells (10 800), and platelets (500 000). Collagen markers were negative; urinalysis also yielded normal results. HIV and Epstein–Barr virus titers were not indicative of either recent or remote infection. A purified protein derivative was also placed, and no induration was seen.

Hematological consultation was performed and bone marrow aspirate was recommended, which was normal, and the patient received two packed red blood cells with improvement of hemoglobin level to 10 mg/dl.

Echocardiography was performed, which was normal apart from minimal circular pericardial effusion.

Patient received 60 mg oral prednisolone for 2 weeks without improvement of hypoxia, cough, or fever.

Histopathological examination was needed to confirm a final diagnosis; hence, PET/computed tomographic scan was performed (Fig. 2).

The image findings are of glucose avid bilateral pulmonary reticulonodular infiltrations and consolidative patches as well as bilateral hilar, subcarinal, and paratracheal lymph nodes with standard uptake value of 5 with high suspicion of malignancy.

So after a high-risk consent patient had done a mediastinoscope with no perioperative or postoperative complications happened.

Histopathology revealed enlarged lymph nodes with prominent mantle zones comprised of small, mature lymphocytes arranged in concentric layers with prominent hyalinized vessels surrounded by plasma cells, picture of Castleman's disease.

Patient initiated pulse steroid therapy in the form of 1 g methyl prednisolone for 3 days followed by rituximab (monoclonal antibodies).

With marked improvement of fever, cough, and dyspnea, the patient had improvement in oxygen saturation to 95% on room air.

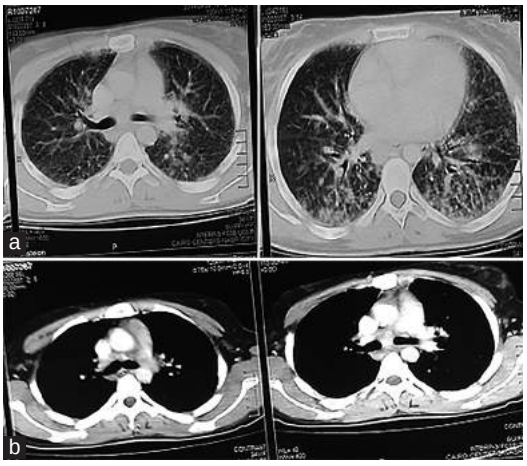
The patient was continued on 60 mg prednisolone with another rituximab after 2 weeks.

Comment

Our diagnosis was plasma cell interstitial pneumonia as a manifestation of multicentric Castleman's disease.

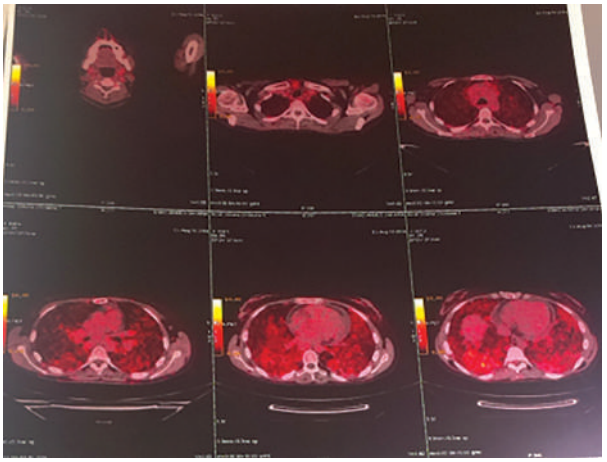
Castleman's disease is an uncommon lymphoproliferative disorder defined as 'a localized hyperplasia of lymphoid

Fig. 1



(a) Lung window shows bilateral reticulonodular infiltrations mainly at lung bases. (b) Mediastinal window shows bilateral hilar lymph nodes.

Fig. 2



PET/computed tomographic scan with diffuse glucose uptake.

follicles with and without a germinal center formation and marked capillary proliferation with endothelial hyperplasia' [1], known as the lymphoma impostor [2].

The clinical manifestations of Castleman's disease are highly dependent on the histopathology of the lesion. The plasma cell variant can be much more severe than the hyaline vascular form [3].

The diagnosis of Castleman's disease is ultimately by histology, thereby requiring either removal or biopsy of the lesion for definitive diagnosis [1].

The exact mechanism of systemic symptoms with either form of Castleman's disease is not known exactly. However, a growing body of evidence indicates that Castleman's disease is primarily a lymphoproliferative or inflammatory disorder [4]. Beck *et al.* [5] confirmed the generation of interleukin-6 (IL-6) in the germinal centers of hyperplastic lymph nodes and subsequent complex interactions with IL-1 and tumor necrosis factor-α.

There is no standard therapy for multicentric Castleman's disease. Treatment modalities include antiviral drugs such as ganciclovir for human herpesvirus type 8, chemotherapy, corticosteroids, immunomodulators, and monoclonal antibodies against IL-6 [6].

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

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