CASE REPORTS

RARE CASE OF EOSINOPHILIC PNEUMONIA IN PATIENT WITH KIMURA'S DISEASE

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Abstract

Kimura's disease (KD) is a benign, multi-system chronic inflammatory condition characterized by hyperplastic lymphadenopathy and subcutaneous nodules due to inflammatory eosinophilic infiltrates. It has a predilection for the head and neck region and is almost always accompanied by peripheral eosinophilia and elevated serum IgE levels. Here we report a case of eosinophilic pneumonia (EP) as part of the clinical presentation in Kimura's disease.

Keywords: eosinophilic pneumonia, Kimura's disease, eosinophilia.

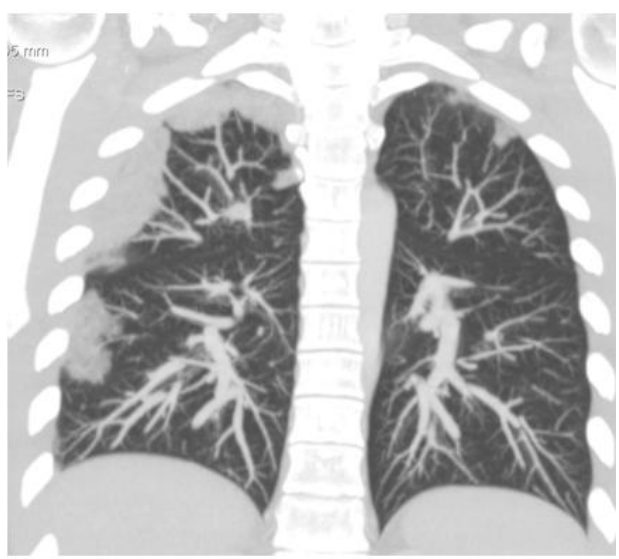
Introduction

Kimura's disease (KD) was first described in 1937 by Kimm, and its histopathology was later characterized by Kimura et al in 1948.(1) This is a rare benign chronic inflammatory condition presenting with painless lymphadenopathy with predilection for the head and neck, peripheral eosinophilia and subcutaneous nodules. Patients almost always have elevated serum IgE levels. Accurate clinicopathological diagnosis is important as it can mimic infectious, vasculitic, lymphoproliferative and neoplastic diseases.(1-3) This is the first reported case of eosinophilic pneumonia in association with Kimura's disease.

Case Report

A 17 year old Caucasian male presented with two months history of progressive right-sided cervical lymphadenopathy. He denied symptoms of fever, night sweats, and weight loss. Patient had no respiratory complaints at initial presentation. Physical examination revealed a 7 cm mass in right neck and bilateral groin adenopathy. Patient had patchy eczematous lesions involving both lower extremities. Laboratory tests revealed elevated eosinophil count at 1920/mm3 (23%) and IgE was elevated at 323 IU/ml. Computed tomography (CT) of the neck confirmed right cervical and right supraclavicular lymphadenopathy. Whole body positron emission tomography (PET) revealed bilateral mediastinal and inguinal lymphadenopathy with mild avidity for fluorodeoxyglucose-18 (FDG). Incidental note was made of a small area of ground glass opacity in the right upper lobe, despite an unremarkable chest radiograph. Bone marrow biopsy with immunohistological work-up was negative. Infectious and other inflammatory tests including EBV titers, cat scratch disease, and ANCA were unremarkable. Subsequently, the patient underwent radical right cervical lymph node dissection. Histopathological evaluation of the cervical mass revealed follicular hyperplasia, scattered giant cells, interfollicular eosinophilia, interfollicular microabscesses and proliferation of thin-walled vessels. Findings were consistent with an infectious or inflammatory process, especially KD. Extensive immunostaining were performed to rule out infectious process in the lymph nodes and all results were negative. IgE immunostaining of lymph node sample was positive, congruent with diagnosis of KD. The patient subsequently presented 9 months later with insidious onset of cough and dyspnea on exertion. Patient had not received any medical therapy in the interim. There was persistent eosinophilia of 1560 / mm3 (24%) and mild bilateral groin lymphadenopathy on exam. Chest radiograph and CT chest revealed peripheral opacities in the lung apices and mid lung zones (Figure 1). Patient underwent bronchoscopy with bronchoalveolar lavage (BAL) and biopsy. BAL revealed negative bacterial, acid-fast bacilli and fungal cultures. Biopsy showed an accumulation of eosinophils and macrophages in the alveoli and alveolar septal thickening by leukocytes. Patient was then started on a steroid therapy. Follow-up in a month revealed resolution of dyspnea and cough with near complete resolution of peripheral opacities on chest radiograph.

Figure 1 : Pulmonary infiltrates on CT chest



Discussion

KD usually occurs in the younger patients between the second and third decades of life and is endemic in East Asia. Men are more commonly affected than women. The disease follows an indolent course, with gradually increasing lymphadenopathy over months or years. The overall prognosis is good, but spontaneous involution is rare. (1,3)

The etiology of KD is unknown, but an autoimmune reaction to punitive antigens has been suggested. The most common histologic features of KD include preserved nodal architecture; follicular hyperplasia with reactive germinal centers; well-formed mantle zones; eosinophilic infiltrates involving the interfollicular areas, sinusoidal areas, perinodal soft tissue, and subcutaneous tissue; and proliferation of postcapillary venules. Immunohistochemical staining with IgE shows a characteristic reticular staining pattern of germinal centers(2,3).

Lymphadenopathy most commonly involves the posterior auricular and cervical nodes. Involvement of mediastinal, inguinal lymph nodes as well as salivary glands has been reported. KD can be associated with mesangial proliferative glomerulonephritis, minimal change disease and membrane nephropathy. This patient had normal kidney function and no evidence of microproteinuria. Skin complications such as atopic dermatitis and urticaria are common. (1,3). Patient did have eczematous changes in lower extremities that were partially responsive to topical steroids. Associations between KD and asthma has been reported. (1)

Eosinophilic pneumonia (EP) is an inflammatory pulmonary condition that can be caused by various etiologies including vasculitides, parasitic infections, medications and idiopathic conditions. (4,6) Patients may complain of shortness of breath, cough and fever. Diagnosis of EP is made when there is radiographic evidence of pulmonary infiltrates, increased eosinophils in either lung biopsy or bronchoalveolar lavage (BAL) fluid (usually >25%) with frequent association with peripheral eosinophilia. (5,6). Eosinophilic pneumonia in association with Kimura's disease is rarely seen as was seen in our patient.

To date, no standardized treatment protocol for KD has been established. Therapeutic options include surgery, radiotherapy, steroid, and other immunosuppressive agents. As there is a tendency for lesion to recur after cessation of steroid therapy, other modalities such as local irradiation and immunosuppressive agents have shown promises. (7)

Conclusion

KD should be suspected as a cause of lymphadenopathy with associated eosinophilic pneumonia in the setting of peripheral eosinophilia and elevated IgE level, following exclusion of infectious, vasculitic and neoplastic etiologies.

Authors Contribution

PJ and RCJ supplemented images and edited final manuscript, SHK supplemented pathology slides and assisted in writing pathology parts of the case, EMH was the primary scientific investigator.

Conflict of Interest

The authors of this manuscript received no funding and have no conflict of interest to report.

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