Rare disease

Primary pulmonary classic Hodgkin's lymphoma

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Primary pulmonary Hodgkin's lymphoma (HL) is very uncommon. The diagnosis is only made if nodal involvement is absent and disease elsewhere has been excluded. Here, the authors present a 54-year-old female with fever, chills, sweating, non-productive cough, anorexia and weight loss. Chest x-ray and CT scan revealed bilateral and multiple nodules especially in upper pulmonary lobes without mediastinal lymphadenopathy. The biopsy from lung nodule revealed classic HL which was primary.

BACKGROUND

Pulmonary lymphoma is rare and represent less than 1% of primary lung cancer. Lung involvement is usually

secondary. Primary pulmonary lymphoma encompass 10% of extranodal lymphoma.¹ The majority are non-Hodgkin lymphoma (non-HL) and primary pulmonary

Figure 1 Chest x-ray showed diffuse bilateral reticulonodular pattern.

Figure 2 CT scan revealed bilateral and multiple nodules especially in upper pulmonary lobes; there was no mediastinal lymphadenopathy and pleural effusion.

Figure 3 Classic Reed–sternberg cells in the inflammatory background ($\times 100$).

Figure 4 Reed–sternberg cells with other inflammatory cells in the lung biopsy ($\times 400$).

HL is very uncommon.² The diagnosis is only justified if hilar lymph nodes are uninvolved and disease elsewhere has been excluded. Microscopically, the presence of classic Reed–Sternberg cells among immunoreactive cells and stroma is diagnostic and adequate specimen usually achieved by open-lung biopsy.³ Recognition of HL in lung, although based on well-established morphologic criteria, may represent a source of interpretative problems because of the unusual clinical presentation as well as the peculiar histologic changes induced within the pulmonary microenvironment.⁴

CASE PRESENTATION

A 54-year-old female presented with fever, chills, sweating, non-productive cough, anorexia and weight loss (10 kg) from 6 months ago. During this period, antibiotic therapy was ineffective, and hence she was admitted. She had diabetes mellitus, hypertension and hyperlipoproteinemia from 4 years ago. She was housewife and did not give any history of contact with environmental pollutant. On physical examination, body temperature was 38.5°C and had coarse crackle in her chest. X-ray and CT scan revealed bilateral and multiple nodules especially in upper pulmonary lobes, and there was no mediastinal lymphadenopathy and pleural effusion (figures 1 and 2). Abdominal and pelvic sonography and CT scan were normal. Laboratory investigation revealed a haemoglobin of 9 g/dl, total leucocyte count of 17 000/mm³ with neutrophilic leukocytosis and erythrocyte sedimentation rate of 105 mm at the end of first hour. Transbronchial lung biopsy was inconclusive

and open-lung biopsy was performed. Microscopic examination showed characteristic Reed–Sternberg cells and its variants in a background of lymphocytes, plasma cells, neutrophils and eosinophils (figures 3 and 4). Adjacent lung parenchyma revealed nodular aggregates of lymphoid tissue in the peribronchiolar and interstitial tissue. The diagnosis was classic HL. Immunohistochemical (IHC) stains also confirmed this diagnosis. CD30, CD20, LCA and pan-cytokeratin were negative.

INVESTIGATIONS

X-ray and CT scan revealed bilateral and multiple nodules especially in upper pulmonary lobes, and there was no mediastinal lymphadenopathy and pleural effusion. Abdominal and pelvic sonography and CT scan were normal. Laboratory investigation revealed a haemoglobin of 9 g/dl, total leucocyte count of 17 000/mm³ with neutrophilic leukocytosis and erythrocyte sedimentation rate of 105 mm at the end of first hour.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of primary pulmonary lymphoma includes necrotising granulomatous inflammation particularly Wegener's granulomatosis, Langerhans cell histiocytosis, undifferentiated carcinoma and other from of malignant lymphoma. Primary pulmonary HL rich in neoplastic cells may resemble anaplastic large cell lymphoma and T cell/histiocyte-rich large cell B cell lymphoma. IHC stains are helpful.

TREATMENT

Our patient received ABVD (adriamycin, bleomycin, vinblastine and dacarbazine).

OUTCOME AND FOLLOW-UP

She is well now after 5 years' follow-up.

DISCUSSION

Primary pulmonary HL is rare but distinct entity, to be distinguished from nodal Hodgkin's disease and from lymphomas involving the lung secondarily.^{5–6} Fewer than 100 cases have been reported.⁷ Kern *et al*⁸ proposed the following criteria for establishing a diagnosis of primary pulmonary HL: (1) anatomic documentation of typical histologic features of HL; (2) disease restricted to the lung without or minimal hilar lymph node involvement and (3) adequate clinical and/or pathologic exclusion of disease at other sites. Our case met all of these criteria as extensive investigation did not reveal any extrapulmonary disease at the time of presentation. Reported cases cover a wide age range (18–82, mean=43), and there is a slight female predominance. The presented case was female, and she was 54 years old. Most patients have respiratory symptoms and about half also have B symptoms.^{1–8} Our case also had B symptoms and a lot of time investigated for fever of unknown origin (FUO). The pulmonary lesions are single nodule in one third of the patients and multiple nodules in one half. Occasionally, bilateral reticulonodular infiltrates or pulmonary consolidation are seen. Upper lobes involvement are more common, and there may be cavitations in the larger ones.^{9–10} Since the clinical presentation in primary pulmonary HL is non-specific, open-lung biopsy is needed in the majority of cases.¹¹ Occasionally, FNA coupled with IHC stains is helpful.^{12–13} The microscopic appearances are typical of classic HL usually of either nodular sclerosis or mixed cellularity type, and IHC stainings are confirmatory.¹³ The differential diagnosis of primary pulmonary lymphoma includes necrotising granulomatous inflammation particularly Wegener's granulomatosis, Langerhans cell histiocytosis, undifferentiated carcinoma and other from of malignant lymphoma. Primary pulmonary HL rich in Reed–Sternberg cells may resemble anaplastic large cell lymphoma and T cell/histiocyte-rich large B cell lymphoma. IHC stains are helpful. For appropriate treatment, clinical staging is needed. In patient with solitary pulmonary nodule, surgery and

radiotherapy are effective but in diffuse and bilateral nodular lesions, chemotherapy is the treatment of choice. Our case was treated by chemotherapy agents, and she is well after 5 years. To conclude, although primary pulmonary HL is a rare condition, it is in differential diagnosis of a group of more common pulmonary disease, such as infection, bronchogenic carcinoma, pulmonary metastasis and vasculitis and should be considered in the differential diagnosis list.

Learning points

- ▶ Primary pulmonary classic HL is a rare distinct entity.
- ▶ Open-lung biopsy is necessary for a definite diagnosis.
- ▶ It is in differential diagnosis of a group of more common pulmonary diseases and should be considered in the differential diagnosis list.

Competing interests None.

Patient consent Obtained.

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Binesh F, Halvani H, Taghipour S, Navabii H. Primary pulmonary classic Hodgkin's lymphoma. *BMJ Case Reports* 2011;10.1136/bcr.03.2011.3955, date of publication

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