**Pulmonary Langerhans cell histiocytosis with inguinal abscess: Unusual presentation**

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**Abstract**
We report a case of a 30-year-old man who was treated as recurrent right inguinal abscess following a 2-month history of right inguinal swelling and intermittent fever with no respiratory symptoms. Resection of his right inguinal lymph node and the histopathological analysis revealed the diagnosis of Langerhans cell histiocytosis. In addition, the CT of the thorax showed presence of bilateral cystic lung changes consistent with pulmonary Langerhans cell histiocytosis.

**Keywords**
Langerhans cell histiocytosis, inguinal abscess, pulmonary

**Introduction**
Langerhans cell histiocytosis (LCH) is a rare histiocytic disorder characterized by infiltration by histiocytes, which may infiltrate nearly every organ, including lungs and lymph nodes. The incidence is one to two cases per million adults. Pulmonary LCH (PLCH) is a rare disorder occurring in adults that is associated with cigarette smoking. The principal manifestations of PLCH depend on the disease phase and include bronchiolocentric lesions in various features with a typical distribution pattern, relatively sparing the basal region.

Our case report highlights an interestingly rare case of LCH manifesting as right inguinal abscess and the importance of recognizing this rare and unusual manifestation of LCH.

**Case report**
A 30-year-old Malay man who was a chronic smoker and had underlying poorly controlled Type-2 diabetes mellitus (HbA1C 11.8%) presented with right inguinal swelling for 2 months associated with intermittent fever. He denied any chronic cough or constitutional symptoms. On examination, the right inguinal region revealed erythematous swelling with pus discharge in keeping with inguinal abscess. His other physical examinations were unremarkable. The blood parameters were suggestive of acute bacterial infections. He was subsequently subjected for incision and drainage procedure in view of his poorly controlled diabetes and slow response towards antibiotics.

The histopathology examination (HPE) of the inguinal node showed partial effacement of lymph node with diffuse infiltrate of histiocytoid cells with kidney or bean-shaped nuclei. The cells have abundant, pale eosinophilic cytoplasm, irregular and elongated nuclei with prominent nuclear grooves and folds, fine chromatin and indistinct nucleoli. There were also eosinophils present. Mitotic figures were seen. Immunohistochemistry noted the cells were positive with CD 1a and S100 consistent with LCH (Figure 1).

In view of the HPE diagnosis, the patient was subjected for contrast-enhanced computed tomography (CT) scan of the thorax and abdomen, which revealed multiple small round and oval cystic lung lesions with nodular changes in both lungs sparing the basal regions (Figure 2). The appearances were consistent with PLCH. Note was also made of the skin ulceration at the inguinal region following recent debridement and exploration associated with enlarged nodes with necrotic

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centre (Figure 2). The skeletal survey and assessment of the included bones on CT did not reveal osseous involvement. The patient was subsequently referred to a haematology centre for further management of the disease. Upon discharge, he was advised to stop smoking, and was referred to a smoking cessation clinic. A referral to the endocrine unit was also made to optimize his diabetic control. Upon follow-up in the chest clinic few months after his discharge, he was well and had returned to his daily routine. He has also quit his smoking habit, had a good diabetic control with HbA1C 5%, and was leading a healthy lifestyle. Knowing that smoking will cause progression of his lung disease, he decided that he will never smoke again.

Discussion

Pulmonary involvement is frequently observed in adults with single-system LCH. In patients with multisystem LCH, the organs involved are mainly the bone and skin, followed by the haematopoietic system, lymph node, liver, spleen, soft tissue, lung, thymus, pituitary gland, and other organs. As presented in this case, the patient’s disease was manifested by involvement of the lymph nodes and lungs. His condition of inguinal abscess was further complicated by poorly controlled diabetes.

The exact aetiology of LCH is unknown. Compelling evidence indicates that exposure to cigarette smoke is the most important factor associated with the development of PLCH, as over 90% of adult patients who develop PLCH are cigarette smokers or were previously subjected to substantial second-hand smoke. Our patient has this significant risk factor; he was a chronic smoker for more than 20 pack years. The onset of lymphadenopathy in PLCH is rare. Here, we have demonstrated a case of LCH affecting the inguinal nodes as a rare presentation.
On CT, PLCH manifests as small nodules in early (inflammatory) stages and thin- or thick-walled cysts in more advance stages, with a characteristic distribution in the upper and middle lung fields and relative sparing of the lung bases. Cysts may appear round and of small dimension (<1 cm), but in more advance stages they are typically larger and demonstrate irregular, bilobed or bizarre shapes. This is well evidenced in our patient as his CT thorax showed numerous small cysts bilaterally with sparing of the lung bases. Lung biopsy was not necessary in the present case as the CT findings of the lungs were typical of PLCH and the confirmed concurrent inguinal involvement of LCH further supported the lung diagnosis. Lung biopsy may be needed in selected cases when the CT scan shows isolated micronodular changes and if the imaging diagnosis is still unclear.

The pathological specimen typically reveals the presence of Langerhans cells, which was subsequently confirmed by immunohistochemical staining with monoclonal antibodies directed against the membrane antigen CD1a. It is important to point out that positive staining for the intracellular S100 protein that was also seen in our patient, although widely used to identify Langerhans cells, is not specific to these cells and can be observed in neuroendocrine cells and some macrophages.

Exposure to cigarette smoke is an important factor of PLCH in our reported case. PLCH may result in permanent disability with chronic dyspnoea and cough. The disease may stabilize or even regress after quitting smoking. Persistent cigarette smoking may lead to progression of the disease process. Hence, smoking cessation as part of the treatment is crucial. As per our case, the patient responded well to smoking cessation. Multiple system LCH manifested as lymphadenopathy and bilateral lung cysts is very rare. To our best knowledge, this is the first reported case with such presentation in the Malaysian adult population.

This case represents an uncommon entity of cystic lung disease, in an unusual presentation. A multidisciplinary approach involving the radiologists, pathologists, haematologists and surgeons is crucial in managing such a complicated, rare and atypical case. Further databases of case collections are needed to identify the rare forms of this disease, its manifestation and the best mode of treatment.

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Author contributions

All authors are equally involved in planning, drafting and completing the manuscript.

Ethical approval

Non applicable.

Informed Consent

Informed consent was obtained for this case report.

Availability of data

Data is available for review.

Trial Registration

Non applicable.

Declaration of Conflicting Interests

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