中文題目:肺外類肉瘤併發慢性腹痛:案例報告 英文題目:Unexpained Chronic Abdominal Pain related to Extrapulmonary Sarcoidosis 作者:盧律衡¹,楊曉芳²,吳秉儒³ 服務單位:¹高雄醫學大學附設醫院內科部,²高雄醫學大學附設醫院病理科, ³高雄醫學大學附設醫院胃腸內科

Introduction:

Sarcoidosis is a mutisystemic disease associated with immune dysregulation inducing inflammatory process. The exact cause of sarcoidosis is still unknown. It affects predominantly the lungs, followed by skin or other lymphatic system(1). Clinical manifestations depend on which organs it affects. The most common symptoms are chronic cough and dyspnea. Fatigue and body weight loss are also commonly seen owing to inflammatory process. Due to its clinical diversity and resemblance to other disorder, sarcoidosis is hard to diagnosed. We reported a case of a 55 year-old-female with sarcoidosis involving perigastric and celiac basin regions, presented with chronic epigastric pain.

Case Report:

A-55-year-old female had past medical history for chronic kidney disease stage 5, diabetes mellitus and hypertension. She was presented to our emergency room due to chronic epigastralgia, significant body weight loss(22 present drop) and general malaise for 4 months. She denied fever, association with food intake, bloody stool and constipation. Recent upper endoscopy only showed normal-appearing stomach and proximal duodenum. Physical examination showed lymphadenopathy over right neck. Abdomen computed tomography(CT) without contrast showed no obvious structural abnoramalty. The laboratory test showed bilirubin level of ______, lipase level of 49 U/L but ionized calcium level of 6.73 mg/dl was noted, arising the concern of malignancy or disseminated tuberculosis infection.

Positron emission tomography (PET)(Figure 1) found high uptake of F-18 fluorodeoxyglucose(FDG) over bilateral supraclavicular, paratracheal, perigastric, hepatoduodenal and celiac basins lymph nodes. Excisional biopsy over right supraclavicle lymph node was done. Pathology(Figure 2) revealed non-caseating granulomas. Acid fast stain for mycobacteria is negative.

There, glucocorticoid was given for symptomatic sarcoidosis treatment. Abdomen pain and hypercalcemia relieved in days.

Conclusion:

Extrapulmonary sarcoidosis accounts for 30% of cases and can be isolated symptoms.

Sarcoidosis presents with abdominal pain had been reported in only few cases of stomach, liver, spleen or mesenteric lymph node invasion(3). The diagnosis depend on the three criteria: clinical and radiological presentation, evidence of non-caseating granulomas, and evidence of no alternative disease(2). However, it is hard to diagnose if without typical airway symptoms or obvious chest radiograph abnormalty. PET is a useful tool in this situation not only for detecting lesion but also offer us an ideal site for biopsy. In our case, lymph nodes are mainly located at hepatoduodenal area and are eaily misdiagnosed as normal structural of intestine or pancreas under non-contrast CT.

Sarcoidosis resembles many disease such as lymphoma, tuberculosis or other autoimmune disease(4). Physicians often rely on pathology report to confirm the diagnose. As its diverse presentation, the severity of sarcoidosis is widely variable. If left untreated, half of case may resolve spontaneously within 2 years(2). However, some cases may not resolve or even exacerbate. Primary treatment for sarcoidosis is glucocorticoids. Some experts may add Disease-modifying antirheumatic drugs (DMARDs) or even biologic agents to refractory case or to avoid long term steroid use.



Figure 1. Positron emission tomography (PET)The axial section showed (A) Paratrachea, periaorta and (B) perigastric, hepatoduodenal and celiac basins lymphadenopathy. The coronal section (C) and (D) showed bilateral supraclavicular, periaorta and hepatoduodenal lymphadenopathy.



Figure 2. Histopathology of the right neck lymph node The Low-power view (left) confirmed multiple noncaseating granulomas. The High-power view (right) revealed aggregated epithelioid cells and Langhans giant cells (arrow).