Subcutaneous sarcoidosis: a reported case

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ABSTRACT :

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Subcutaneous sarcoidosis is a rare specific lesion of sarcoidosis, which presents with multiple subcutaneous nodules. The common locations are the extremities. The histopathology shows nodular granulomas scattered in the deep dermis and subcutaneous fat. The patients usually have systemic involvement, especially hilar adenopathy.

We report a 58-year-old Caucasian male presented with multiple nodules on right forearm and dorsum of right hand. Computed tomographic (CT) scan reveals bilateral hilar adenopathy. The histopathology is compatible with subcutaneous sarcoidosis. The patient was treated with 40mg of prednisolone daily. 2-week after treatment, the nodules have reduced in size and number.

Key words: subcutaneous sarcoidosis, sarcoidosis, subcutaneous nodule, hilar adenopathy

บทคัดย่อ :

พัจนภา เวชอนุรักษ์, ปุณวิศ สุทธิกุลณเศรษฐ์ รายงานผู้ป่วยโรค SUBCUTANEOUS SARCOIDOSIS ที่บริเวณแขนและหลังมือด้านขวา วารสารโรคผิวหนัง 2557; 30: 294-298.

สถาบันโรคผิวหนัง

Subcutaneous sarcoidosis เป็นหนึ่งในอาการแสดงจำเพาะของโรค sarcoidosis ซึ่งพบไม่บ่อย ผู้ป่วยมักมาด้วยก้อนที่ผิวหนัง บริเวณที่พบบ่อยคือ แขนและขา ลักษณะทางพยาธิวิทยาจะมี granuloma กระจายอยู่ในผิวหนังแท้ชั้นลึกและชั้นไขมัน นอกจากนี้ ผู้ป่วยมักมีอาการของระบบอื่นๆ ร่วมด้วย โดยเฉพาะมีต่อมน้ำเหลืองบริเวณขั้วปอดโต

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รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยชายชาวออสเตรเลีย อายุ 58 ปี มาพบแพทย์ด้วยก้อนที่แขนขวาและหลังมือขวา ทำเอ็กซเรย์คอมพิวเตอร์พบต่อมน้ำเหลืองบริเวณขั้วปอดโตทั้งสองข้าง ลักษณะทางพยาธิวิทยาเข้าได้กับsubcutaneous sarcoidosisได้ให้การรักษาโดย prednisolone 40mg/day และหลังการรักษาสองสัปดาห์ ก้อนที่แขนและหลังมือขวามีขนาดเล็กลง

Key words: โรค subcutaneous sarcoidosis, subcutaneous nodule, ก้อนเนื้องอกบริเวณแขน

Introduction

Subcutaneous sarcoidosis was firstly described by Darier and Roussy in 1904. This lesion was classified as a specific lesion which the histopathology shows noncaseating granulomas. Subcutaneous sarcoidosis is characterized by multiple skin-colored firm nodules, predominately occur on the extremities. The lesion is usually asymptomatic or mild tender. This subcutaneous form is an uncommon presentation of sarcoidosis, but frequently has systemic involvement. Therefore, the patient must undergo investigated for systemic involvement.

In this report, we describe a patient with subcutaneous sarcoidosis on right forearm and dorsum of right hand. He also has bilateral hilar adenopathy.

Case report

A 58 year-old Australian man presented with 0.5 cm solitary asymptomatic nodule on forehead for 4 months. A biopsy was taken in private hospital revealing nodular aggregates of granulomas. AFB staining was negative. 2 months PTA, the patient has developed progressive multiple asymptomatic nodules, 0.5-3 cm in diameter, along right forearm and dorsum of right hand. The lesions have increased in size and number. No history of previous trauma in this area was noted. Fever, dyspnea, arrhythmia, abdominal pain or arthralgia were not noted. No treatment was given prior his visit to our institute. He was, then, referred to the Institute of Dermatology.

Physical examination revealed multiple firm skin-colored subcutaneous nodules on right forearm and dorsum of right hand, varying from 0.5-3cm in size. Lymphadenopathy was not detected. Complete blood counts, liver function test and renal function test are normal. Chest x-ray and EKG have no abnormality. Pulmonary function test is within normal limits. CT chest showed multiple enlarged lymph nodes at paratracheal, prevascular, precarina, subcarina and both hilar regions, sized upto 1.8 cm, without pulmonary nodule or infiltration.

Histopathologic examination from right forearm showed large packed nodular aggregates of granulomas surrounded by a numbers of lymphocytes in the lower dermis and subcutaneous fat. Multinucleated giant cells are present. The overlying epidermis appears unremarkable. Polarized light microscopy shows absence of foreign body. AFB stain is negative. Tissue

culture for tuberculosis and fungus are negative. PCR for *Mycobacterial spp*. is negative.



Figure 1 Multiple firm skin-colored subcutaneous nodules along right forearm, varying from 1-3cm in size



Figure 2 Solitary firm skin-colored subcutaneous nodule size 3 cm on right dorsum of hand

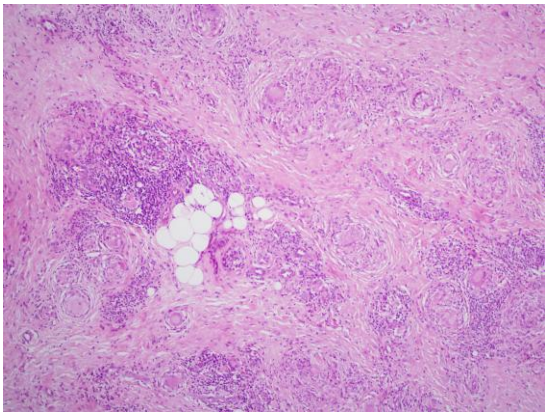


Figure 3 Diffuse nodular aggregates of granulomas surrounded by a numbers of lymphocytes in the lower dermis and subcutaneous fat. Multinucleated giant cells were presented. (H&E, original magnificantx100)

Discussion

Sarcoidosis is a noncaseating granulomatous disease which frequently involves multisystemically. The prevalence of the disease is different among race, location, and gender. The Unites States reported between 10 to 40 cases in 100,000 whereas Japan reported only one to two cases in 100,000.[1] It is more common in females than males. The onset of disease is during the third and fourth decade. The pathogenesis of sarcoidosis has remained unknown but environmental factors and infectious agents may cause dysregulation of T-cell immune response, leading to the formation of noncaseating granulomas. At least 20% of sarcoidosis cases have skin involvement and present in different morphologies, categorized into “specific”, such as lupus pernio, papule, plaque, subcutaneous sarcoidosis and infiltrated scar, or “nonspecific” lesions, such as

erythema nodosum. The histopathology of specific lesions shows noncaseating granulomas, which is not present in nonspecific lesions.

Subcutaneous sarcoidosis is a specific subtype of cutaneous sarcoidosis, firstly described by Darier and Roussy in 1904. This variant is observed in 2% of patients with systemic sarcoidosis and accounted for 12% of specific skin lesions. The peak incidence of the disease is during the fourth decade, predominantly in females.[2] It usually presents with multiple, skin-colored, firm nodules. The common locations are extremities, followed by trunk, buttock, face, head, and neck. The lesions are frequently asymptomatic-to-mild tendered and located bilaterally. Subcutaneous sarcoidosis is found significantly associated with systemic involvement. Bilateral hilar adenopathy is the most common involvement.[3] Other systemic involvements are lung infiltration, parotitis, uveitis, arthritis, mucositis, dactylitis, neurological and renal involvement and hepatosplenomegaly, respectively. Associated autoimmune diseases, including thyroiditis, vitiligo, pernicious anemia, rheumatoid arthritis, ulcerative colitis, systemic lupus erythematosus, sicca syndrome, and cushing disease have been reported.[4,5] It was noticed that this specific subcutaneous lesion appeared before other symptoms.[6] Subcutaneous sarcoidosis occurred after interferon-α therapy in chronic hepatitis C patient has also been reported.[7] The diagnosis of subcutaneous sarcoidosis is based on clinical presentation and the histopathologic finding of noncaseating granuloma involving subcutaneous fat. An infectious cause and foreign body particle must be ruled out. The imaging modalities, including MRI and sonographic study are useful for diagnosis; however, they are not specific.

First line treatment for sarcoidosis is prednisolone 0.3-0.5 mg/kg/day. The response usually occurs at 4-8 weeks after treatment. Hydroxychloroquine is also reported impressively responsive.[8] Alternative treatments such as methotrexate, allopurinol, minocycline and potassium iodide are also reported with variable outcome. No chronic or severe complication has been reported.

Our patient presented with multiple asymptomatic skin-colored nodules on right forearm and dorsum of right hand. Clinical differential diagnoses include multiple lipomas, epidermal cysts, calcinosis cutis, cutaneous metastasis and rheumatoid nodules. The less common lesions such as tuberculosis and subcutaneous mycoses have also been excluded. Based on clinical manifestation and histopathologic findings, subcutaneous sarcoidosis was diagnosed. The investigation revealed bilateral hilar adenopathy by CT chest, which is the most common systemic involvement in sarcoidosis. The patient has been treated with prednisolone 40mg/day (0.3mg/kg/day). The subcutaneous lesions are reduced in size after 2 weeks of treatment.

In conclusion, we report a patient presented with subcutaneous sarcoidosis with bilateral hilar

adenopathy. He was treated with systemic corticosteroids with a good response.

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