Case 20

Multiple red lumps in a chubby boy

Piyatida Prasertvit, M.D.
Penpun Wattanakrai, M.D.

Patient: A 8 year-old Thai boy from Nakhon Ratchasima

Chief complaint: Multiple red papules and nodules for 2 weeks

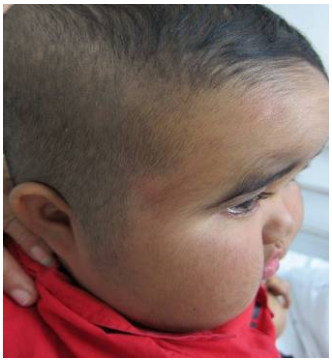
Present illness:

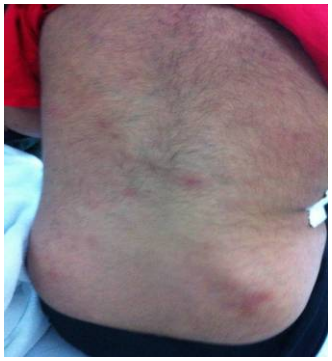
A known case of hypereosinophilic syndrome and autoimmune hepatitis was diagnosed 2 years ago. His current medications include prednisolone 10 mg/day, azathioprine 75 mg/day (3MKD), hydroxyurea 20 MKD. He was admitted to the pediatric ward for liver biopsy due to abnormal rising of liver function test profiles.

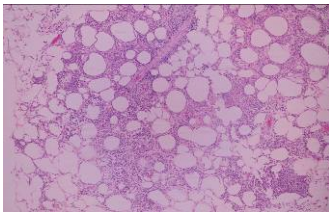
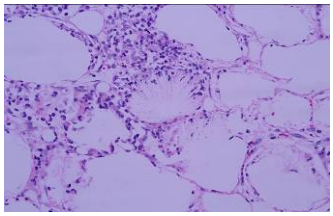
He was sent for dermatologic consultation because of multiple erythematous painful subcutaneous papules and nodules on his face, torso and extremities with gradual increasing in number and extent of disease without history of fever for 2 weeks before this admission.

Past history:

- Hypereosinophilic syndrome was diagnosed 2 years ago with WBC 43,900 (Eo 63%) (AEC 27,657 cells). Bone marrow biopsy was done twice, 1 month apart, and from both biopsies no atypical or malignant cells were detected.
- Autoimmune hepatitis was diagnosed 6 months later after the diagnosis of hypereosinophilic syndrome**.** He presented with jaundice and laboratory investigations showed ANA-fine speckle and anti smooth muscle titer 1:40. Liver biopsy result was compatible with autoimmune hepatitis (mild interface hepatitis with moderate lymphoplasmacytic cell).
- Both diseases were treated and followed up with pediatric allergist and gastroenterologist, respectively





Skin examination:

- Fifteen discrete erythematous painful subcutaneous papules and nodules, were scattered on his face, torso and extremities**,** size ranging from 2 to 4 cm.

Histopathology: (S13-27583, back)

- Dense inflammatory-cell infiltrate of lymphocytes, numerous neutrophils, histiocytes within fat lobules of subcutaneous tissue
- Fat necrosis and some needle-cleft formation in adipocyte

Diagnosis: Post-steroid panniculitis

Review history of steroid doses

20-3-13	Pred(5) 4x2	5-6-13	Pred(5) 6x1
30-7-13	Pred(5) 5x1	24-9-13	Pred(5) 4x1
10-10-13	Pred(5) 3x1	24-10-13	Pred(5) 2x1
1-11-13	erythematous subcutaneous papules present		

Treatment:

Treatment for post-steroid panniculitis: none

Treatment for hypereosinophilic syndrome and autoimmune hepatitis:

- Additional trestment with cyclosporin (100mg/ml) 0.5ml oral bid (4MKD) was started
- Continue azathioprine 75mg/day and prednisolone 10 mg/day
- Hydroxyurea was discontinued

Discussion:

Post-steroid panniculitis (PSP) is a rare complication of prolonged systemic corticosteroid treatment, first reported by Smith and Good in 1956. It has usually been described in childhood and early adulthood, and is related to a quick decrease or a sudden tapering/ withdrawal of high dose systemic corticosteroids[1,2] for the treatment of several diseases such as nephrotic syndrome[3], autoimmune enteropathy[4], COPD[5], brain tumor[6]. As previously reported, it usually appears 1 to 20 days[4] after cessation of high doses of systemic corticosteroids[1]. This entity can be identified by clinical history and characteristic histopathological findings.

Clinically, the lesions usually appear as erythematous subcutaneous nodules on the cheeks, arms or trunk. They present on areas prone to accumulation of fat during steroid therapy. Characteristic histopathological findings from the lesions help differentiate PSP from other possible causes of panniculitis[7].

The characteristic histopathological features in PSP, consist of lobular panniculitis without vasculitis, with narrow strands needle-shaped clefts in radial arrangement within the cytoplasm of histiocytes, multinucleated giant cells, and

adipocytes. These findings are only documented 2 other entities, sclerema neonatorum and subcutaneous fat necrosis of the newborn. Therefore, with consideration of age of our patient, the other 2 entities can completely be rule out.

The etiopathogenesis of PSP is not clear, but some authors suggest that the withdrawal of corticosteroids causes an abnormal lipid metabolism, resulting in an increased ratio of saturated to unsaturated fatty acids, leading crystal formation[6].

For the treatment of PSP; reinstitution of corticosteroids[4], gradual decrease of the dose[3], weight loss and topical steroid[6] has induced improvement of panniculitis. Some reports documented that they resolve spontaneously without any treatment intervention[6,8]. The average resolving time ranges from 5 days to 5 months, leaving sequelae of atrophic, hyperpigmented or normal appearing skin.

From our literature review, this case was the first case that developed PSP despite gradual tapering of oral steroids by 5mg per month for 5 months. We do not know whether the lesions spontaneously resolved or improved from the addition of cyclosporin prescribed to control his active hepatitis. Six weeks after the diagnosis of PSP, all lesions gradually decreased and resolved completely without leaving any scars or pigmentary change.

At the present time, the patient has been followed up for 8 months without any evidence of relapse.

References:

1. Torrelo A, Hernandez A. Panniculitis in children. Dermatol Clin. 2008; 26(4): 491-500.
2. Requena L, Sanchez Yus E. Panniculitis. Part II. Mostly lobular panniculitis. J Am Acad Dermatol. 2001; 45(3): 325-61.
3. Sacchidanand SA, Kanathur S, Somaiah S, Srinivas SM. Post-steroid panniculitis: A rare case report. Indian Dermatol Online J. 2013; 4(4): 318-20.
4. Peralta L, Morais P, Trindade E, Barreto F, Canelhas A, Amil J. Poststeroid panniculitis: a rare complication of systemic corticosteroid therapy. Cutan Ocul Toxicol. 2012; 31(2): 164-166.
5. Kim ST, Kim TK, Lee JW, et al. Post-steroid panniculitis in an adult. J Dermatol. 2008; 35(12): 786-8.
6. de-Andres-del-Rosario A, Verea-Hernando MM, Yebra-Pimentel MT, Rosende-Maceiras L, Pineyro-Molina F, Capdevila EF. Poststeroid panniculitis in an adult. Am J Dermatopathol. 2011; 33(7): e77-80.
7. Polcari IC, Stein SL. Panniculitis in childhood. Dermatol Ther. 2010; 23(4): 356-67.
8. Kwon EJ, Emanuel PO, Gribetz CH, Mudgil AV, Phelps RG. Poststeroid panniculitis. J Cutan Pathol. 2007; 34: 64-7.