2. Szücs G, Szekanecz Z, Zilahi E, Kapitány A, Baráth S, Szamosi S, et al. Systemic sclerosis-rheumatoid arthritis overlap syndrome: a unique combination of features suggests a distinct genetic, serological and clinical entity. Rheumatology (Oxford) 2007;46:989-993.
3. Rosenbach M, English JC 3rd. Reactive granulomatous dermatitis: a review of palisaded neutrophilic and granulomatous dermatitis, interstitial granulomatous dermatitis, interstitial granulomatous drug reaction, and a proposed reclassification. Dermatol Clin 2015;33:373-387.
4. Kim YS, Lee JH, Lee JY, Park YM. Interstitial granulomatous dermatitis associated with rheumatoid arthritis. Ann Dermatol 2016;28:395-397.
5. Moinzadeh P, Aberer E, Ahmadi-Simab K, Blank N, Distler JH, Fierlbeck G, et al. Disease progression in systemic sclerosis-overlap syndrome is significantly different from limited and diffuse cutaneous systemic sclerosis. Ann Rheum Dis 2015;74:730-737.

https://doi.org/10.5021/ad.2017.29.6.806

A Case of Segmental (Zosteriform) Juvenile Xanthogranuloma

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Dear Editor:

A 14-year-old boy presented with asymptomatic skin nodules. Clinical examination revealed multiple, 0.3～0.5 cm-sized, brown to skin-colored nodules in a band-like fashion along the left side of the waist (Fig. 1). The lesions were said to have appeared 6 months ago and have been increasing in size and number. He denied the history of trauma or other cutaneous inflammation. There were no systemic symptoms such as fever and he had no family history of any skin diseases. There were no evidence of systemic organ involvement, including eyes and bones. The clinical differential diagnoses included prurigo nodularis, steatocystoma multiplex, segmental leiomyoma and juvenile xanthogranuloma (JXG). A 4-mm punch biopsy was taken from lesions on the back and left flank. Histopathologic examination showed dense lymphohistiocytic infiltration in the dermis. Touton-type giant cells with foamy cytoplasm were present. The overlying epidermis was normal. Histiocytic cells were stained with CD68 (Fig. 2). S-100 stain was negative. Laboratory tests were normal including lipid profile. Based on these findings, a final diagnosis of segmental distribution of JXG was made. Patient was lost for follow-up after the initial visit.

JXG is the most common form of the non-langerhans cell histiocytosis. It is a self-limiting disorder which typically occurs during infancy or childhood. It is known to disappear within months to years without any treatment. JXG typically presents as a solitary, yellow-brown papule or nodule commonly affecting the head, neck, and trunk. Though the lesions disappear spontaneously without any treatment, it is at times associated with systemic disorders, such as neurofibromatosis and myeloproliferative disorders[1]. When the lesions are confined to the skin, complete removal is suggested only for cosmetic purpose.

Received March 16, 2016, Revised November 3, 2016, Accepted for publication November 7, 2016

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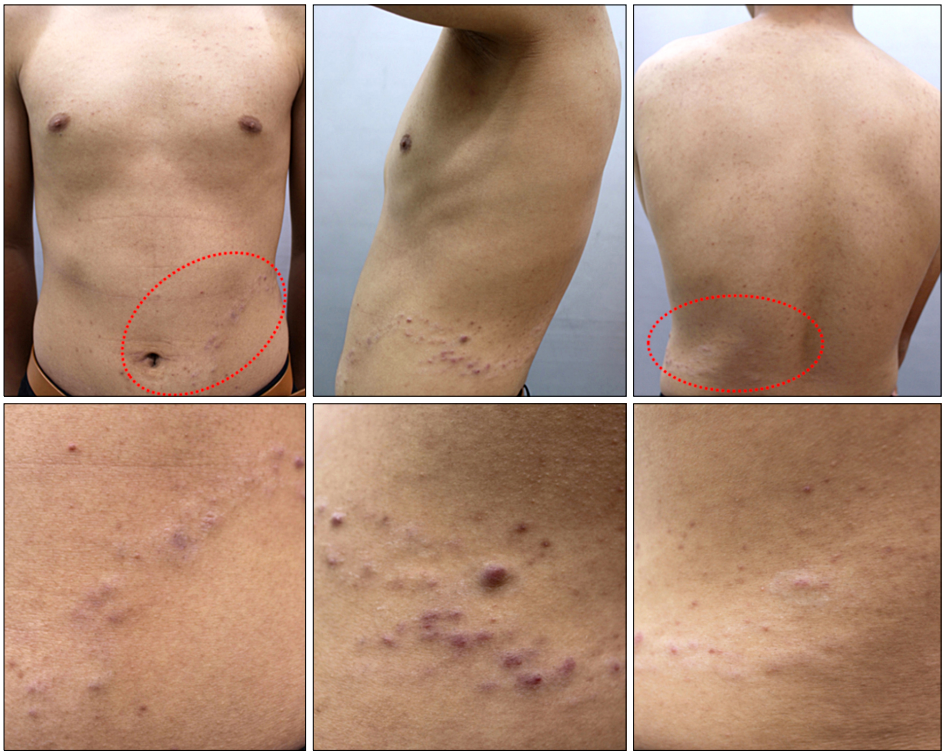

Fig. 1. Multiple, variably-sized, brown to skin-colored nodules along the waist (dotted circles).

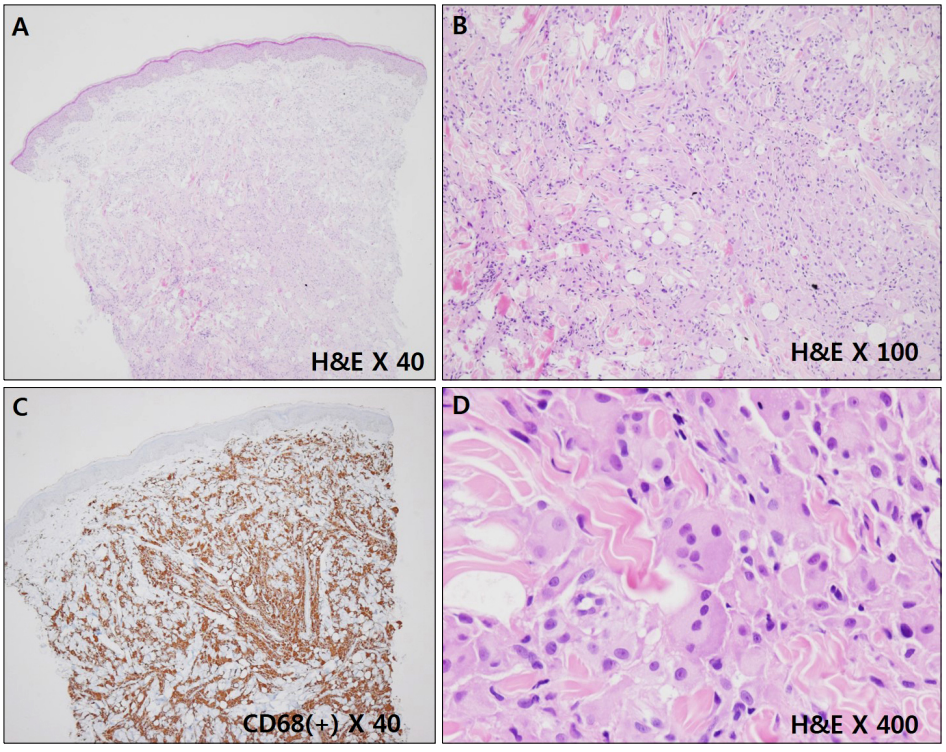

Fig. 2. (A, B, D) Histopathologic examination shows dense lympho-histiocytic infiltration and touton-type giant cells with foamy cytoplasm in the dermis. (C) Histiocytic cells are positive in CD68 stain.

Atypical variants of JXG include generalized, lichenoid, giant, mixed, plaque, subcutaneous, coupled, granuloma annulare-like, muscular and clustered forms. Among the variants, 'segmental' or 'clustered' distribution of JXG is extremely rare[2]. To our knowledge, only two cases of segmental JXG have been reported where the cases presented as asymptomatic papules or nodules on the neck, trunk and right leg. Differential diagnoses include tumors that

arise in a linear pattern such as leiomyoma and primary cutaneous lymphoma[3]. The prognosis of segmental or linear JXG does not seem to differ from solitary JXG where spontaneous remission was reported in a case of segmental JXG[2]. The segmental distribution of JXG raises the question of the mechanism of development of the lesions. The most plausible explanation is somatic skin cell mosaicism. The origin of mutated cells composing the lesion are derived from the bone marrow. Homing and local expansion of the mutated predominant cell species of JXG occur and segmental distribution can evolve[4]. In autosomal dominant diseases, such as Hailey-Hailey disease and neurofibromatosis, segmental manifestation has been reported. Moreover, similar cases of 'agminated' or 'segmental' distribution of dermatofibromas and fibrous histiocytomas have been reported and these reports support our hypothesis of postzygotic somatic mutation in an embryogenic period[5].

This case highlights the unusual manifestation of this relatively common disorder in childhood. JXG can present with a wide range of morphologic characteristics including segmental or linear distribution.

ACKNOWLEDGMENT

This study was supported by a grant of the Korean Healthcare technology R&D project, Ministry of Health & Welfare, Republic of Korea (Grant no. HN15C0105).

CONFLICTS OF INTEREST

The authors have nothing to disclose.

REFERENCES

1. Garay M, Moreno S, Apreéa G, Pizzi-Parra N. Linear juvenile xanthogranuloma. Pediatr Dermatol 2004;21:513-515.
2. Kaur MR, Brundler MA, Stevenson O, Moss C. Disseminated clustered juvenile xanthogranuloma: an unusual morphological variant of a common condition. Clin Exp Dermatol 2008;33:575-577.
3. Ng SY. Segmental juvenile xanthogranuloma. Pediatr Dermatol 2014;31:615-617.
4. Kiorpelidou D, Stergiopoulou C, Zioga A, Bassukas ID. Linear-agminated juvenile xanthogranulomas. Int J Dermatol 2008;47:387-389.
5. Soon SL, Howard AK, Washington CV. Multiple, clustered dermatofibroma: a rare clinical variant of dermatofibroma. J Cutan Med Surg 2003;7:455-457.