PAEDIATRIC DERMATOLOGY

REDEFINING STIFF SKIN SYNDROME AS SPECTRUM DISEASE

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Background: Stiff skin syndrome (SSS) is rare disease characterized by progressive, noninflammatory fibrosis of the skin that often occurs mostly in a bilateral distribution. Recently, segmentally distributed SSS has been reported. However, new SSS cases presented with classic, segmental and generalized distribution have been noted. Whether SSS could be categorized as localized, segmental and generalized subtypes should be discussed.

Observation: Here, we presented sixteen unreported cases of SSS from our clinic and reviewed other reported SSS in China to analyze their clinical features, histopathology and possible prognosis. Of 16 cases, there was 1 case presented with only buttock involved, 4 cases with unilateral segmental involved, and 8 cases with generalized SSS involving buttock, lumber, shoulder and sporadic nodules. Seven cases have hypertrichosis. All skin biopsy revealed thickened collagen bundles, lack of inflammation, and adipocyte entrapment. After reviewing all other 40 cases reported in China, we found that except 1 case presented only on neck and 3 cases on shoulder, 13 cases were distributed segmentally and 23 cases were generalized. Four cases have no hypertrichosis and only 2 reported cases had familiar history.

Key message: Based on analysis of clinical and histopathologic features, most SSS were classic with bilateral buttock distribution, but variable types such segmental and generalized with sporadic nodules should be aware of in clinic. Adipocyte entrapment has been considered as an important histopathological feature of SSS. However, differential diagnosis from connective tissue nevus and morphea should be considered. Genetic factor and prognosis of SSS will be further studied.