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RESUMEN

bjective. Juvenile localized scleroderma is usually considered a disease that is confined to the skin and subcutaneous tissue. We studied the prevalence and clinical features of extracutaneous manifestations in a large cohort of children with juvenile localized scleroderma. Methods. Data from a multinational study on juvenile scleroderma was used for this in-depth study. Clinical features of patients with extracutaneous manifestations were compared with those of patients who had exclusively skin involvement. Results. Seven hundred fifty patients entered the study. One hundred sixty-eight patients (22.4%) presented with a total of 193 extracutaneous manifestations, as follows: articular (47.2%), neurologic (17.1%), vascular (9.3%), ocular (8.3%), gastrointestinal (6.2%), respiratory (2.6%), cardiac (1%), and renal (1%). Other autoimmune conditions were present in 7.3% of patients. Neurologic involvement consisted of epilepsy, central nervous system vasculitis, peripheral neuropathy, vascular malformations, headache, and neuroimaging abnormalities. Ocular manifestations were episcleritis, uveitis, xerophthalmia, glaucoma, and papilledema. In more than one-fourth of these children, articular, neurologic, and ocular involvements were unrelated to the site of skin lesions. Raynaud's phenomenon was reported in 16 patients. Respiratory involvement consisted essentially of restrictive lung disease. Gastrointestinal involvement was reported in 12 patients and consisted exclusively of gastroesophageal reflex. Thirty patients (4%) had multiple extracutaneous features, but systemic sclerosis (SSc) developed in only 1 patient. In patients with extracutaneous involvement, the prevalence of antinuclear antibodies and rheumatoid factor was significantly higher than that among patients with only skin involvement. However, Scl-70 and anticentromere, markers of SSc, were not significantly increased. Conclusion. Extracutaneous manifestations of juvenile localized sclerodenna developed in almost one-fourth of the children in this study. These extracotaneous manifestations often were unrelated to the site of the skin lesions and sometimes were associated with multiple organ involvement. The risk of developing SSc was

very low. This subgroup of patients with juvenile localized scleroderma should be evaluated extensively, treated more aggressively, and monitored carefully.

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