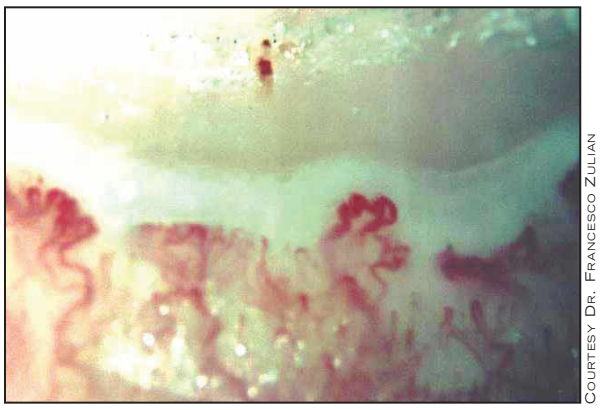

Courtesy Dr. Francesco Zulian

Capillaroscopy changes typical of juvenile systemic sclerosis usually appear after Raynaud's phenomenon and the appearance of serum autoantibodies.

Raynaud's Precedes Autoantibodies' Onset in Juvenile Systemic Sclerosis

BY NANCY WALSH
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ABANO TERME, ITALY — Childhood-onset systemic sclerosis is different from adult-onset scleroderma, typically being characterized by less internal organ involvement and lower mortality rates, Dr. Francesco Zulian reported at a congress on skin, rheumatism, and autoimmunity.

According to data from the Padua international database that includes 1,000 juvenile patients with various types of scleroderma, 153 have systemic sclerosis, said Dr. Zulian of the pediatrics/rheumatology division at the University of Padua (Italy).

The average age of systemic sclerosis onset was 8.1 years, and the disease duration at diagnosis was 1.9 years. The female to male ratio was 3.6:1.0, and 139 of the patients had the diffuse subtype of systemic sclerosis, he said. The limited subtype is more common among adults.

The first manifestation of systemic sclerosis in children typically is Raynaud's phenomenon. This is followed by the appearance of autoantibodies, and then capillaroscopy changes.

The pattern of autoantibodies is different from that seen in adults with scleroderma. Antinuclear antibodies were seen in 81% of the pediatric patients, compared with 94% of adults, whereas anti-Scl-70 antibodies were seen in 34% vs. 43%, respectively, Dr. Zulian said.

Skin involvement accounts for 60% of juvenile systemic sclerosis symptoms in children, which is much lower than that found in adults.

Rheumatoid factor was found in 17% of the children, compared with 23% of the adults, he added.

Anticentromere antibodies were found in 7.1% of children in the database, compared with 23% of adults, which reflects the lower prevalence of the limited subtype of systemic sclerosis in children, he said.

The most frequent manifestations were Raynaud's phenomenon, which was seen in 84% of the juvenile patients, and skin induration, which was found in 76%. Abnormal lung function tests were seen in 40% of the patients and pulmonary fibrosis in 25%.

Skin involvement accounts for 60% of symptoms in children, which is much lower than in adults. It is also more difficult to make the diagnosis in young patients, Dr. Zulian said.

Musculoskeletal involvement also was quite common, with arthralgias reported by 36%, arthritis by 27.5%, and muscle weakness by 24.2%. Gastroesophageal reflux was found in 30%, but small-bowel involvement was rare.

"In 127 patients we have enough data to make some conclusions about outcome," Dr. Zulian said. At present, 15 (11.8%) of the patients have died, 10 of cardiac failure, 2 of renal failure, 2 of respiratory failure, and 1 of septicemia. Of these 15, 4 died within the first year after diagnosis, and 11 within 5 years of diagnosis. "This means that there is a group of patients who have a very aggressive course of disease and who did not respond to treatment," he said.

Organ involvement is the major predictor of poor outcome, so those who present with early respiratory, cardiac, or gastrointestinal involvement must be evaluated and treated aggressively, he said. ■

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