Acro-osteolysis in a Patient with Scleroderma

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A 32 year old woman was admitted due to diffuse systemic sclerosis. Clinical manifestations included Raynaud’s phenomenon, skin fibrosis, telangiectasia, esophageal involvement and pulmonary fibrosis. Examination revealed diffuse skin changes, reduced oral aperture, mouse-like facial features, flexion contractures of both hands, digital ischemic ulcerations of the fingertips and under the fingernails, appearing as necrotic debris trapped under the nail. There were areas of hypopigmentation (vitiligo-like) on the dorsal surface of the hands. The patient was unable to make a fist. The Rodman score (assessment of skin fibrosis) was 15. There was no evidence of calcinosis.

Radiography of both hands revealed symmetric resorption of soft tissue of the fingertips. There were erosions of the distal tufts due to acro-osteolysis. No evidence of erosive disease of the interphalangeal joints was found. No subcutaneous calcification was detected. Calcinosis is characteristic of diffuse, anti-Scl 70-positive scleroderma in only 20% of cases.

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Procrastination is the art of keeping up with yesterday

Don Marquis (1878-1937), American humorist and journalist

**Capsule**

**Smoking mothers and infant birth weight**

Little is known about genetic susceptibility to cigarette smoke in relation to adverse pregnancy outcomes. Wang et al. tried to investigate whether the association between maternal cigarette smoking and infant birth weight differs by polymorphisms of two maternal metabolic genes: CYP1A1 and GSTT1.

A case-control study was conducted in 1998–2000 among 741 mothers (174 ever smokers and 567 never smokers) who delivered singleton live births at Boston Medical Center. A total of 207 cases were preterm or low-birth-weight infants and 534 were non-low-birth-weight, full-term infants (control). The results showed that without consideration of genotype, continuous maternal smoking during pregnancy was associated with a mean reduction of 377 g (SE, 89 g) in birth weight. When CYP1A1 genotype was considered, the estimated reduction in birth weight was 252 g (SE, 111 g) for the AA genotype group but 520 g (SE, 124 g) for the Aa/aa genotype group. When GSTT1 genotype was considered, the estimated reduction in birth weight was 285 g (SE, 99 g) and 642 g (SE, 154 g) for the present and absent genotype groups, respectively. When both CYP1A1 and GSTT1 genotypes were considered, the greatest reduction in birth weight was found among smoking mothers with the CYP1A1 Aa/aa and GSTT1 absent genotypes (-1,285 g, SE 234 g, P<0.001). Among never smokers, genotype did not independently confer an adverse effect. A similar pattern emerged in analyses stratified by maternal ethnicity and in analyses for gestation.

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