Cutis marmorata telangiectatica congenita and vascular twin naevi are rare vascular anomalies in which focal acantholytic dyskeratosis is usually not observed. We describe a 44-year-old-man who presented for evaluation of skin lesions that had been present since birth. Physical examination revealed anaemic macules adjacent to a naevus telangiectaticus on the chest. Naevus anaemicus was also seen on the shoulders, arms, and left leg. There was bluish-reddish reticulate marking of the skin and cutaneous atrophy. Shortening and hypoplasia of the left leg was observed. Histologic examination of two biopsy specimens revealed focal acantholytic dyskeratosis. In vivo confocal laser scanning microscopy showed dilated capillaries and vessels of the upper dermal plexus in the telangiectatic and decreased capillary blood flow in the anaemic skin sites. The findings were consistent with a diagnosis of cutis marmorata telangiectatica congenita, vascular twin naevi, and incidental focal acantholytic dyskeratosis. The particularities of the present case are the following: firstly, the association of two rare vascular anomalies to which the genetic concept of mosaicism can be applied; secondly, the occurrence of incidental focal acantholytic dyskeratosis in sites of vascular naevi.