Background.Tuberous sclerosis complex (TSC) is a genetically determined hamertomatous neurocutaneus disease with high phenotypic variability. The classical triad of manifestations, which include facial adenoma sebaceum, epilepsy and mental retardation occurs in only 29 % of patients. Many organs may be involved, but lesions on the skin are cardinal for diagnosis suspection. The most common skin lesions are hypomelanotic macules (ash-leaf spots), forehead plaques, shagreen patch, multiple facial angiofibromas and Koenen tumors. Appropriate diagnosis requires a multidisciplinary approach to properly treat affected individuals.

Objective.A 56-year-old caucasian woman presented with multiple periungual skin-colored to reddish tumors,_which vary in size, on each legs' toe. Some of them were bleeding and ulcerate. These periungal lesions with gradual onset in puberty were asymptomatic at the beginning, but covered all toe nails and became more enlarge and painful with the time. All the nails were dystrophic. Since her childhood, she also had multiple dome shaped reddish to brown papular lesion less than 0.5 cm over nasolabial fold and cheeks suggestive of facial angiofibromas (adenoma sebaceum) in a butterfly-shaped pattern on her cheeks and nose. This lesions were treated with electrocoagulation, but they appeared again rapidly after treatment. On the trunk we observed few hypomelanotic macules and shagreen patches. The patient reported a single epileptic seizure at 15 years of age that was not recorded, confirmed, or treated by a physician. There was no other clinical evidence of tuberous sclerosis complex (TSC).

Material and methods. Cortical tubers and periventricular subependymal nodules were excluded by magnetic resonance imaging (MRI). The electroencephalogram (EEG) and the electrocardiography (ECG) were normal, and no pathologic findings were detected on radiographs of the chest, hands, and feet or by abdominal ultrasound imaging, and she had normal cognitive function. Her son also has multiple papulonodular lesion on his face, but he refused any medical examinations and treatments. Biopsy was performed from the periungual lesion and from papular face skin lesion.

Results. Histological examination from periungual lesion, established Koenen tumor, by fibrosis with stellate shaped strands of fibroblasts and dense collagen with numerous ectatic blood vessels. Histological examination from face papule, shows the picture of typical adenoma sebaceum.

Conclusion Periungual fibromas were treat with Cryotherapy, electrocoagulation and trichloroacetic acid with good cosmetic result.

Key words: Tuberous sclerosis complex, Koenen tumors, adenoma sebaceum, shagreen patch, ash-leaf spots