

Brook-Spiegler Syndrome Made of Multiple Trichoblastomas only: Clinical and Dermoscopic Appearance

Case Report

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Abbreviations: BSS: Brooke-Spiegler Syndrome.

Case Report

Brooke-Spiegler Syndrome (SBS) is a rare, autosomal dominant genodermatosis characterized by the association of multiple benign annular neoplasms including eccrine spiradenomas, cylindromes, and trichoblastoma [1]. We report the clinical and dermoscopic aspects of a phenotypic variant made of multiple trichoblastomas.

A 42-years-old man who was presented to our department with the clinical complaint of multiple papulonodular lesions on his face evolved since the age of 14 years. His mother also has similar lesions on her face. The man has no clinical symptoms associated with the lesions. At the clinical examination, the patient has numerous papular and nodular of variable size and normal skin color situated at the level of the nasolabial fold, the nose, and the internal nail of the orbit without resulting in any deformation

(Figure1, below). Through Dermoscopy, we found out a calcification, an hyperpigmentation and a telangiectasia. (Figure 2 below). The histological study confirms trichoblastoma, and the diagnosis of Brooke-Spiegler Syndrome (SBS) was retained in front of the multiple aspect of lesions.

Trichoblastoma is a benign follicular tumor. Its multiple appearance is reminiscent of Brooke-Spiegler syndrome, which is a genodermatous mutation of the CYLD tumor suppressor gene located on chromosome 16q12-q13 [1, 3]. The phenotypic mutation is variable even within the same family. Clinically, they can be solitary or multiple and usually occur in adults with an increase in size while the person grows in age. Trichoblastomas are preferentially located in the nasal folds, nose and internal edge of the orbit bilaterally and symmetrical [4]. This syndrome is important to know because would allow us not to mix it with other diseases such as the basal cell carcinomas [2]. Treatment modalities avail-

Figure 1. multiple trichoblastomas of a nasolabial fold.



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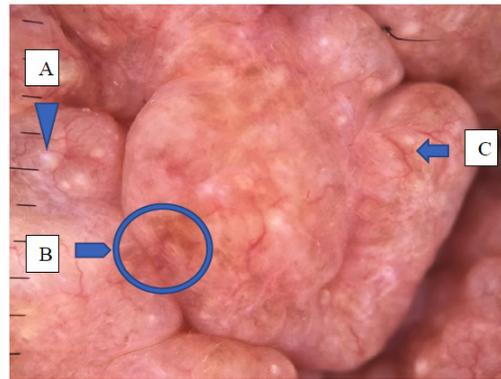
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Figure 2. Dermoscopy (A): Calcification; (B): Hyperpigmentation; (C): Telangiectasia.

able in patients with SSB include tumor excision, dermabrasion, electrodesiccation, cryotherapy and radiotherapy with argon and CO₂ lasers [5].

This article has presented a family with a phenotypic variant of Brooke-Spiegler syndrome made up of multiple trichoblastomas but without Cylindromas or Spiradenoma. Diagnosing this case was based on clinical dermoscopic and histological criteria.

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