Comparative Dermatology* Dermatologia comparativa*

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Abstract: The impressive facial angiofibromas, from a 32 year-old male paciente, with the classical features of tuberous sclerosis, were compared with mulberries. Keywords: Angiofibroma; Facial dermatoses; Tuberous sclerosis.

Resumo: Demonstra-se o quadro exuberante dos angiofibromas faciais em paciente do sexo masculino, de 32 anos, com esclerose tuberosa, os quais podem ser comparados com amoras. Palavras-chave: Angiofibroma; Dermatoses faciais; Esclerose tuberosa.

ANGIOFIBROMAS MORIFORMES

Tuberous sclerosis, a disease that belongs to the group of neurocutaneous disorders, is also called Bourneville Disease, after the physician who first related the cutaneous lesions to the neurologic affection. The term epiloia is also used, albeit with less acceptance in the Anglo-Saxonic literature.¹

The classical cutaneous picture consists of multiple face angiofibromas, mistakenly called sebaceous adenomas, which are preferentially located in the nasogenal sulci, malar regions and mentum. Leaf-shaped hipomellanotic macullae, periungueal fibromas and fibrotic plaquae in the lumbossacral region complete the dermatological picture.¹

The genetic defect was described in genes TSC1 and TSC2, and it is possible to detect the causing mutation in 90% of the cases.² Affection of gene TSC2 leads to more severe clinical pictures. Gene TSC1 is located in chromosome 9 and encodes hamartine; gene TSC2 encodes tuberine and is located in chromosome 16.³

These two proteins interact in a not fully understood manner, being tumoral supressors;⁴ the absence or dysfunction of one or the other leads to the picture of tuberous sclerosis. It has already been shown that an enhanced expression of these proteins inhibits cellular proliferation and that mutant animals that do not express them exhibit reduction of mitosis time.⁴ Likewise, a smaller expression of these two

proteins has been demonstrated in fibroepithelial polypi. Thus, they are likely to be involved in other illnesses involving tissue hypertrophy/hyperplasia.⁵

A 32 year-old white male with a characteristic picture of tuberous sclerosis was examined. His angiofibromas began during childhood, having a typical location on the nasogenal sulcus (Figure 1) and



FIGURE 1: Angiofibromas on the right nasogenal sulcus

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FIGURE 2: Angiofibromas on the mentum



FIGURE 3: Mulberry (Morus nigra)

mentum (Figure 2). However, the lesions were very exuberant, acquiring an unequivocal mulberry-like aspect (Figure 3). This patient also presented convulsions with a normal neuropsychomotor development,

reflecting the known lack of parallelism between the neurologic and cutaneous pictures, such as seen in this patient.

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