

## Flexural Comedones

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### *To the Editor:*

A new form of comedones, known as childhood flexural comedones, has recently been reported.<sup>1</sup> The authors described the development of comedones in the large skin folds during childhood, in which the comedones had 2 orifices connected via a thin layer of epidermis. No predominance in either sex has been reported. In most cases, the patients consulted for another condition and the comedones were an incidental finding. The lesions were usually solitary, unilateral, and located in the axillae.

Since this entity was first defined, we have seen 3 patients with lesions

consistent with those described. A 5-year-old boy was seen for multiple comedones with double orifice in the axillae that had been present for 9 months (Figure). A soft, white, keratinous material was expressed from 1 of the larger, cystic lesions. A 25-year-old man, who consulted for severe acne, presented numerous comedones with double orifice on the neck and back from childhood. A 40-year-old woman, who had experienced polymorphic acne mainly on the face during adolescence, presented comedones with double orifice on both sides of the neck from childhood. None of the patients were aware of any other cases of flexural comedones in their families.

The appearance of comedones is usually related to acne, hidradenitis suppurativa, chronic sun damage, or other types of cutaneous damage.<sup>2</sup> It can also occur after molluscum contagiosum infection.<sup>3</sup> The site of childhood flexural comedones, which present particularly in the axillae and occasionally also in the groin, indicate that this entity could be related to hidradenitis suppurativa. The 2 adult patients described, who presented

abundant comedones with double orifice on the back and neck, had developed severe acne during adolescence. This suggests that childhood flexural comedones could be related to the development of acne or hidradenitis suppurativa during adolescence or adulthood. These observations also indicate that childhood flexural comedones may persist into adulthood and may be found along with comedones with double orifice at different sites in the skin folds, as well as on the back.



**Figure.** Comedones with double orifice, with a cystic lesion.

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## Unilateral Multiple Facial Angiofibromas: Description of a New Case

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### *To the Editor:*

Tuberous sclerosis, also called tuberous sclerosis complex (TSC),<sup>1</sup> is a rare multisystem genetic disease that causes benign tumors in the brain and other vital organs, such as the kidneys, heart, eyes,

lungs, and skin. The prognosis of the disease is determined by renal manifestations, in which the appearance of renal angioliipomas causes retroperitoneal bleeding and progressive renal failure, the main causes of death in these patients.<sup>1</sup>

Bilateral multiple facial angiofibromas are the most common dermatologic manifestation of TSC and are considered a major criterion in establishing the diagnosis.<sup>1</sup> The unilateral presence of facial

**Table.** Case Reports of Unilateral Multiple Facial Angiofibromas Published to Date

Case No.	Year Published	Author	Sex	Age	Age at Onset of FA	Facial Site	Other Findings
1	1996	McGrae <sup>2</sup>	F	23	5	L	No
2	1997	Ankiller <sup>3</sup>	M	26	8	L	No
3	1998	García Muret <sup>4</sup>	F	16	3	R	Renal angioliopomas
4	1998	García Muret <sup>4</sup>	M	11	7-8	R	No
5	2000	Silvestre <sup>5</sup>	M	5	2	R	No
6	2000	Silvestre <sup>5</sup>	M	12	5	L	Hypopigmented macula
7	2002	Del Pozo <sup>6</sup>	F	59	5	L	No
8	2003	Trauner <sup>7</sup>	M	52	5	L	Occipital poliosis
9	2004	Alonso <sup>8</sup>	F	39	20	L	No
10	2004	Alonso <sup>8</sup>	F	37	9	L	Amaurosis
11	2004	Sharma <sup>9</sup>	F	12	3	L	Hypopigmented macula
12	2004	Mashhood <sup>10</sup>	M		7	R	Periungual fibromas Hypopigmented macula Cranial calcifications Retinal tumors
13	2006	Camprubi <sup>11</sup>	M	13	3	L	No
14	2007	Hall <sup>12</sup>	F	28	14	R	No
15	–	Bordel	F	40	15	L	No

Abbreviations: FA, facial angiofibromas; F, female; L, left; M, male; R, right.

angiofibromas is rare, however, and only 14 cases have been published in the literature (Table).<sup>2-12</sup> Other cutaneous manifestations are hypomelanotic macules, connective tissue hamartomas or nevi, and periungual or subungual fibromas. We present a new case of unilateral multiple facial angiofibromas with no other manifestations of TSC.

Our patient was a 40-year-old woman with a personal history of traumatic cataract in the right eye and appendectomy. She is currently being monitored by the neurology department for multiple sclerosis treated with AM3 (Inmunoferon). She consulted for the progressive appearance of completely asymptomatic, small papular lesions located in the left nasolabial crease that had been slowly, but steadily, growing since she was 15 years old. These lesions had been treated on various occasions by her primary health care physician with keratolytics, but with no apparent

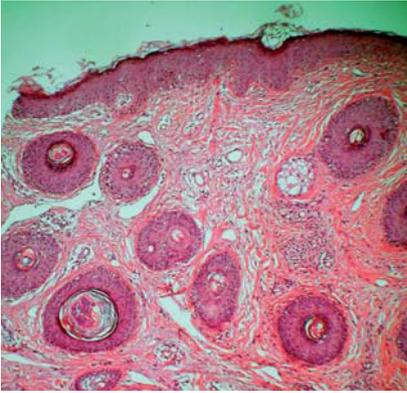
clinical improvement, hence her referral to our department. She had no personal or family history of any dermatologic, neurologic, renal, or cardiac disease, but did mention that a brother occasionally presented epileptic seizures of unclear etiology. Her parents were not consanguineous. The physical examination revealed multiple flesh-colored dome-shaped papules of 2 to 4 mm in diameter, randomly distributed, but primarily located in the left nasolabial creases, without crossing the midline (Figure 1). The remaining physical examination, including a comprehensive neurologic examination, was normal. Examination under Wood light did not show any hypopigmented lesions, and no unguinal lesions were observed.

A histopathologic study of 1 of the cutaneous lesions confirmed the clinical suspicion of facial angiofibroma, revealing perivascular fibrosis associated

with angiomatous hyperplasia (Figure 2). All imaging tests performed by the neurology department were carefully reviewed and, because the physical examination was strictly normal, no other additional tests were requested. We recommended carbon dioxide laser treatment, which was refused by the patient; she is currently attending periodic follow-up visits.



**Figure 1.** Multiple small dome-shaped papules located in the left nasolabial crease, without crossing the midline.



**Figure 2.** Histopathologic study of a papular lesion showing the presence of telangiectases, along with noticeable perivascular and perifollicular concentric fibrosis (hematoxylin-eosin,  $\times 40$ ).

TSC is a genetic disease of highly variable phenotype, characterized by the triad of mental retardation, epilepsy, and facial angiofibromas. It is presently considered a hamartomatous process characterized by a cell proliferation, migration, and differentiation disorder inherited as a dominant autosomal trait with variable penetration, although 60% to 70% of cases are sporadic.<sup>1</sup> These sporadic cases represent new spontaneous mutations and, therefore, lack any prior family history of the disease as in our patient. The condition occurs as a result of mutations in the *Tsc1* and *Tsc2* genes. These determining genes have been identified on chromosomes 9q34 and 16p13.3.<sup>13</sup>

Bilateral multiple facial angiofibromas, formerly and incorrectly known as sebaceous adenomas, are a major criterion for diagnosis of TSC, but have been found in patients with type 1 multiple endocrine neoplasm<sup>14</sup> and type 1 neurofibromatosis.<sup>15</sup> The condition begins to develop in the mid-facial area during childhood (age 4–10 years) and affects 80% to 90% of patients with TSC,<sup>7</sup> manifesting clinically as erythematous dome-shaped papules with a smooth, glossy surface. These lesions are composed of vascular and connective tissue, and although pathognomonic of TSC, are not very useful for early diagnosis because they

appear in late childhood. The unilateral presence of these facial angiofibromas is rare: only 14 cases have been reported in the literature (Table), 8 of them with no other criteria of TSC. Our case represents an addition to this second group. However, the remaining 6 patients with unilateral facial angiofibromas presented other clinical findings that support the TSC diagnosis, such as hypopigmented macules,<sup>4,5,9,10</sup> poliosis,<sup>7</sup> amaurosis,<sup>8</sup> or renal angioliopomas.<sup>4</sup>

The significance of the unilateral development of facial angiofibromas has remained uncertain over the years. The first cases described were attributed to the various forms of expression of TSC, but they were later considered 1 of the first clinical signs of TSC. At present the condition is considered a segmental, genetically well-defined form of TSC caused by germline mosaicism.<sup>4,5,8,10</sup> The presence of late postzygotic somatic mutation during embryonic development may be responsible for this mosaicism and would explain why only 1 segment of the body surface is affected.

Facial angiofibromas are an unsightly blemish on the face, with occasional episodes of bleeding and skin infections (in vegetating lesions, due to difficulty with hygiene). Various therapeutic measures, such as resection, cryosurgery, curettage, dermabrasion, carbon dioxide laser,<sup>16</sup> argon laser, and pulsed diode laser have been recommended.<sup>7,9,12</sup> Carbon dioxide laser has been successfully used to treat these lesions, although one of the most important problems is long-term recurrence because the lesions cannot be permanently removed, most likely due to their nature.

Early diagnosis of these segmental forms of TSC is important and, although the type of diagnostic examination and genetic counseling to be undertaken in patients with unilateral facial angiofibromas is unclear, like other authors, we believe that regular follow-up is essential.<sup>12</sup> Such follow-up will detect any tumor growth or other

complications and permit adequate therapeutic measures to be taken.

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## Pale Orange Perifollicular Halo as a Dermatoscopic Sign in Scurvy

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### *To the Editor:*

The cutaneous lesions of scurvy have traditionally been described as disseminated purpura, follicular keratotic papules, and "corkscrew" hair.<sup>1-3</sup> The usual dermatoscopic findings have been described as follicular hyperkeratosis, bleeding, and corkscrew hair.<sup>4</sup> Our patient with chronic scurvy presented peculiar scurvy-related findings in the dermatoscopic examination.

A 68-year-old man with a history of alcoholism and no teeth, who had been following a diet consisting solely of plain cakes for quite some time, came to the emergency department of our hospital. He presented considerable deterioration in his overall condition from 2 months previously as well as asthenia, anorexia with significant weight loss, and progressive tendency to be bed-ridden, pain and swelling in the right knee, and

violaceous cutaneous lesions on the legs and abdomen (Figure 1). His personal history included several episodes of hemarthrosis and flare-ups of hematuria in the previous 2 years that had not been specifically diagnosed. Dermatoscopy of the cutaneous lesions revealed a pale orange perifollicular halo surrounded by another peripheral hemorrhagic violaceous halo, along with "corkscrew" hair and follicular hyperkeratosis (Figure 2). A skin biopsy showed a ringlet-like hair shaft sectioned at different levels inside the follicle, compact perifollicular fibrosis, and extravasated erythrocytes in the dermis, but not in the perifollicular fibrotic area mentioned above (Figure 3).



**Figure 1.** Note the large violaceous lesions, follicular hyperkeratosis, and coiled hairs observed in our patient.



**Figure 2.** In addition to "corkscrew" hair, the dermatoscopic image showed a pale orange perifollicular halo, surrounded by violaceous lesions.