

Favre-Racouchot Syndrome

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A 70-year-old white man presented with a 10-year history of slowly expanding lesions with dark spots on his cheeks and gradual coarseness of facial features.

History. The patient had been a farmer until his retirement 3 years ago. He also spent a great deal of time outdoors doing recreational activities such as fishing with significant sun exposure and with minimal protection. He had been smoking more than 25 cigarettes a day for the past 5 decades. He had no history of radiation therapy, exposure to unusual chemicals or toxins, or application of any cosmetics or medical creams to the face. The patient was unhappy and emotionally disturbed with the “dirty” appearance of his face. The dark spots could not be removed, even with frequent washing of the face.

Physical examination. The patient had yellowish, diffusely thickened skin, with deep wrinkles and furrows on the face, especially the forehead and cheeks. Numerous large, open,

black comedones (blackheads) and a few closed comedones (whiteheads) were symmetrically distributed in the malar areas. He also had deep furrowing of the skin at the nape of the neck, with intersecting skin folds resulting in quadrangular and rhomboidal configurations that were characteristic of *cutis rhomboidalis nuchae*. The rest of the physical examination findings were unremarkable.

Diagnosis. Based on the appearance and location of the lesions, the patient received a clinical diagnosis of Favre-Racouchot syndrome.

Discussion. Favre-Racouchot syndrome is a nodular cutaneous elastosis that is characterized by cysts and comedones on actinically damaged skin, typically on the face and less commonly on the neck.¹ The condition was first described in 1932 by the French dermatologist Maurice Favre,² reviewed in detail by Favre and his student Jean Racouchot in 1951,³ and henceforth termed Favre-Racouchot syndrome.

It is estimated that the condition occurs in 1.4% of the adult general population⁴ and in up to 6% of white men older than 50 years.⁵ Although the prevalence is high in white men, the condition has been observed in women and dark-skinned individuals, as well.^{6,7} Middle-aged to elderly individuals are most commonly affected, but cases occurring as early as the second decade of life have also rarely been reported.^{1,5}

Although the exact etiopathogenesis is unknown, chronic exposure to UV light and heavy cigarette smoking are important

triggers.⁶⁻⁸ Chronic exposure to UV light damages the elastic network of the mid and upper dermis with subsequent reduction of its tensile strength.^{6,7} This may cause retention of the sebum, leading to the formation of comedones, cysts, and nodules.^{6,7} Chronic smoking may potentiate the effect of solar damage to the affected skin, and the effect is dose-related.^{1,8,9} Rarely, the condition may follow exposure to radiation therapy.^{1,7}

Histologic findings include marked solar elastosis, epidermal atrophy, basophilic degeneration of the upper dermis, dilated pilosebaceous infundibulum filled with lamellar keratin, and atrophic sebaceous glands.^{1,5} Inflammation is conspicuously absent in the comedones.^{1,6}

Typically, patients with Favre-Racouchot syndrome present with multiple large comedones on actinically damaged skin.^{1,6} Comedones may be closed (whiteheads) or, more commonly, open (blackheads). The comedones differ from those seen in acne by the absence of inflammation.⁶ The underlying skin is solar-damaged and often appears dull, yellowish, thickened, wrinkled, and furrowed.¹⁰ Patients who are severely affected often develop confluent, multiple papules, cysts, and nodules.^{1,5}

The lesions usually symmetrically affect the cheeks, forehead, periorbital area, and/or temples, and sometimes the neck.^{5,6,10} However, unilateral and circumscribed forms have been reported, presumably as a result of asymmetric solar exposure or radiation.⁸

Other conditions reported to be associated with Favre-Racouchot syndrome that also appear on actinically damaged skin include actinic keratosis, cutis rhomboidalis nuchae, trichostasis spinulosa, keratoacanthoma, basal cell carcinoma, and squamous cell carcinoma.⁵

The diagnosis is usually clinical, based on the typical appearance of the lesions (comedones, cysts, and nodules on solar-damaged skin) and typical locations (sun-exposed areas). A skin biopsy is seldom necessary. The differential diagnosis



includes acne vulgaris, sebaceous hyperplasia, milia, epidermoid cysts, chloracne, syringomas, and trichoepitheliomas.

The condition can be cosmetically unsightly and socially embarrassing. The prognosis is good if the patient is properly treated.¹

Avoidance of sun exposure, especially during hours of peak solar UV radiation intensity (11 AM to 4 PM), regular use of broad-spectrum sunscreens, and wearing of protective hats and clothes when outdoors should be emphasized. If the patient is a smoker, cessation of smoking is strongly advised.

Topical retinoids such as tretinoin, adapalene, and tazarotene are the mainstay of medical treatment.^{1,11} The medication works by repairing collagen and remodeling elastin in the dermis.⁷ In addition, the medication may decrease the activity of matrix metalloproteinases within the skin that may further decrease collagen breakdown.⁷ Other treatment options include curettage, excision, comedo extraction, dermabrasion, deep chemical peels, and laser ablation.^{6,9,10} It has been shown that combining medical and surgical treatments yields optimal outcomes.^{1,7,11} ■

REFERENCES:

1. Sonthalia S, Arora R, Chhabra N, Khopkar U. Favre-Racouchot syndrome. *Indian Dermatol Online J.* 2014;5(suppl 2):S128-S129.
2. Favre M. Sur une affection kystique des appareils pilo-sébacés localisée à certaines régions de la face. *Bull Soc Fr Dermatol Syphiligr.* 1932; 39:93-96.
3. Favre M, Racouchot J. L'élastoïdose cutanée nodulaire à kystes et à comédons. *Ann Dermatol Syphiligr (Paris).* 1951;78(6):681-702.
4. Schäfer T, Merkl J, Klemm E, Wichmann HE, Ring J; KORA Study Group. The epidemiology of nevi and signs of skin aging in the adult general population: results of the KORA-Survey 2000. *J Invest Dermatol.* 2006;126(7):1490-1496.
5. Patterson WM, Fox MD, Schwartz RA. Favre-Racouchot disease. *Int J Dermatol.* 2004;43(3): 167-169.
6. Sawicki J, Barankin B. Dermcase. Can you identify this condition? Favre-Racouchot syndrome. *Can Fam Physician.* 2010;56(3): 247-248.
7. Sutherland AE, Green PJ. Favre-Racouchot syndrome in a 39-year old female following radiation therapy. *J Cutan Med Surg.* 2014; 18(1):72-74.
8. Vogel S, Mühlstädt M, Molin S, Ruzicka T, Schneider J, Herzinger T. Unilateral Favre-Racouchot disease: evidence for the etiological role of chronic solar damage. *Dermatology.* 2013;226(1):32-34.
9. Mavilia L, Campolmi P, Santoro G, Lotti T. Combined treatment of Favre-Racouchot syndrome with a superpulsed carbon dioxide laser: report of 50 cases. *Dermatol Ther.* 2010;23(suppl 1):S4-S6.
10. Rai S, Madan V, August PJ, Ferguson JE. Favre-Racouchot syndrome: a novel two-step treatment approach using the carbon dioxide laser. *Br J Dermatol.* 2014;170(3):657-660.
11. Zhang R, Zhu W. Favre-Racouchot syndrome associated with eyelid papilloma: a case report. *J Biomed Res.* 2012;26(6):474-477.