Multiple cutaneous and uterine leiomyomas

Madam,

A 39-year-old woman presented to the Dermatology Clinic at Azahra Hospital Center with a 6-month history of intermittently painful, small cluster of subcutaneous nodules and papules of varying sizes ranging from 0.5 cm to 2 cm in diameter on the lateral aspects of the face that were sensitive to cold weather [Figure 1]. The pseudo-Darier sign was positive. On detailed inquiry, she gave a history of menorrhagia 2 years ago that on sonography uterine leiomyomatosis (UL) has been proposed. After myomectomy, on histopathology UL has been confirmed. Otherwise, her medical, occupational, and family history were normal.

Laboratory testing revealed no significant finding. Pathology samples with H and E staining showed a circumscribed, non-encapsulated proliferation of eosinophilic spindle cells with uniform pencil-shaped nuclei in the papillary and upper reticular dermis. Rare normal mitosis has been detected [Figure 2]. The immunohistochemistry staining revealed positive staining with smooth muscle actin antibody, confirming the smooth muscle origin of the lesions. These pathology findings were compatible with cutaneous leiomyomatosis (CL). Therapy with nifedipine markedly results in controlling pain.

Owing to the possibility of occurrence of renal cancer (RC) with the simultaneous multiple CL and UL,[1,6] we considered the evaluation of genitourinary apparatus. However, on abdominopelvic sonography, and urine biochemistry no finding in favor of RC was detected.

CL is a benign neoplasm originating from smooth muscle that presents by painful nodules. Despite the existence of three variants, the most common presentation is the multiple piloleiomyomatosis that the lesions commonly distributed in a grouped or dermatomal pattern. Even though, CL do not progress to malignant transformation into leiomyosarcoma, the main threat is the occurrence of RC especially when there is simultaneous multiple CL and UL.[1,2]

In about 75% of patients with multiple leiomyomas, a mutation in the fumarate hydratase on chromosome 1q42.3-43 has been reported. This mutation is strongly persistent in cases in which RC occur.[3-5]

In our patient, despite the coexistence of CL and UL, work-up for detecting RC, was negative. This finding was predictable due to the fact that RC has association more dominantly in cases in which there is a positive family history of leiomyoma.[6] However, one should always look for the rare internal associations of cutaneous lesions even when the patient does not complain of a significant problem.

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