An Unusual Association of Cutaneous Myxoma with Favre-Racouchot Syndrome

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Received November 20, 2008; Accepted December 31, 2008.

Key words: Myxoma – Dermatoses – Facial – Nodular Elastoidoses – Magnetic resonance imaging

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Abstract: We present a case of bilateral foot multiple cutaneous myxomas, associated with Favre-Racouchot syndrome (FRS) which is a dermatologic condition of multiple large comedones and nodules of the periorbital areas. As the mucocutaneous and cardiac manifestations were absent, Carney complex was excluded. To our knowledge, there has been no report of the association of cutaneous myxoma and FRS. Additionally, the magnetic resonance images of this benign tumor which is rare in the literature are presented.

Introduction

Cutaneous myxomas are relatively uncommon benign skin tumours. More frequently they are solitary; when they are multiple, they may be one of the components of the Carney's complex characterized by lentigines, spotty pigmentation of skin and mucous membranes, cardiac myxoma and elevated endocrine activity [1, 2]. We report a case of patient with the association of multiple cutaneous myxomas and Favre-Racouchot syndrome (FRS). FRS is characterized by large periorbital cornedones and diffusely thickened, yellow skin representing extensive actinic damage and it is found predominantly in Caucasian men who were for long time exposed to the sun and harsh weather [3].

Case report

A 53-years-old man was referred because of the masses on both his feet for ten years. Physical examination revealed soft lobulated masses in both feet. The size of the tumors was 4×3 cm on the left, 3 cm on the right side and they were extending to the plantar side. On his face, diffusely thickened skin with furrows and multiple comedons in both periorbital areas were seen (Figure 1).
There was no family history of any similar lesions. The patient was a winch

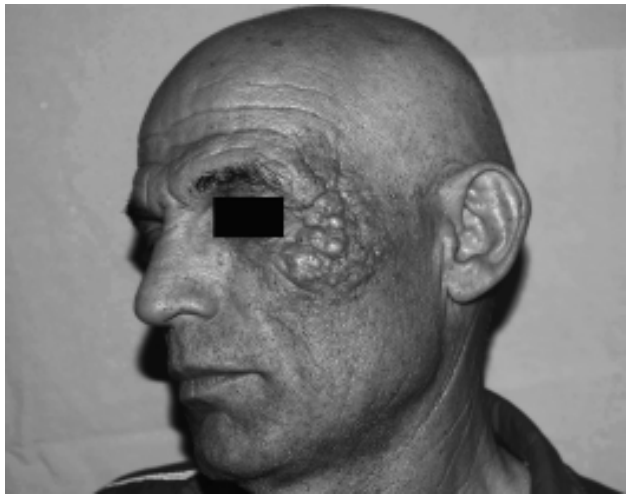

Figure 1 – Multiple large closed comedons, and furrows with diffuse thickened skin.

operator; he was exposed to the sun for long years. He used to smoke for 34 years, 20/day but he quit smoking six months ago.

No myxoma was reported on echocardiogram, and no endocrine pathology was detected clinically. Soft tissue masses were seen, but no bone pathology was detected on roentgenograms. Ultrasound images revealed multiple solid lobulated masses in both plantar aspects of feet (Figure 2A). Magnetic resonance imaging (MRI) demonstrated multiple well-circumscribed lobulated nodular lesions in peritendinous, subcutaneous areas and between fascial planes with hypointense on T1, hyperintense on T2 signal characteristics. Peripheral contrast enhancement was seen after gadolinium administration (Figure 2B, C).

The largest mass which was located on left foot was excised. The tumor was gelatinous, dirty white colour, with 4.5×3×1.5 cm of dimensions (Figure 3). Histology reported a highly vascularised tumor, composed of spindle-shaped

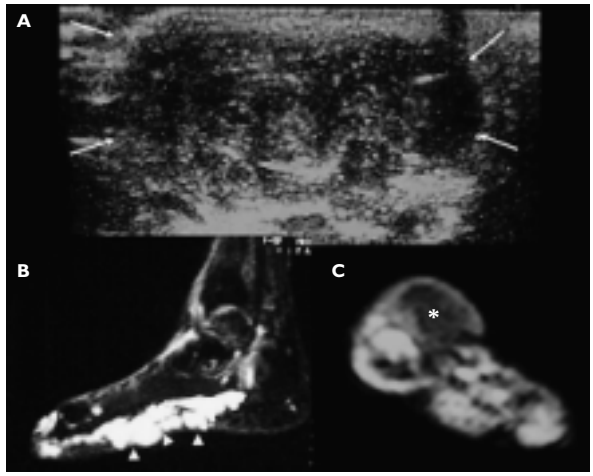

Figure 2 – (A) Sonography depicted solid masses (arrows); (B) T2-W sagittal MRI scan. Multiple, lobulated hyperintense masses in peritendinous, subcutaneous areas and under plantar facsia (arrowheads); (C) After gadolinium injectionT1-W axial scan. Peripheral rim enhancement was seen on masses (asterix).

Figure 3 – Gelatinous, dirty, white colour solid mass was excised surgically.

cytoplasmic cells within a myxoid stroma. Immunocytochemistry for S-100 protein in the stromal cells was negative. These findings were consistent with a myxoma.

Discussion

The large size and infiltrative growth pattern of the lesion makes it difficult to determine whether it originated in the subcutaneous tissue, the joint or the aponeurosis. Juxta-articular and cutaneous myxoma are discussed in the same entity but in different locations. Juxta-articular myxomas involve periarticular tendons, ligaments, joint capsule, muscles, and the adjacent subcutis. As they are generally solitary and associated with degenerative arthritis of the adjacent joint they were excluded in our patient. Knee is the most frequently involved site followed by shoulder, elbow, ankle and hip [4]. In our case, no evidence of degenerative arthritis in the first MTP joint, no connection of the mass with the joint and multiple lesions in different locations proved to be subcutaneous myxoma. The foot location and multilobular, multicentric growth pattern of cutaneous myxomas is extremely rare [3, 5]. The classical MRI features of myxomas are well described; however the MRI scans of myxomas in the literature are rare. Myxomas manifest by cutaneous, subcutaneous masses with homogeneous fluid-like signal intensity and variable enhancement after contrast-injection [6].

When the myxomas are multiple, the association with other types of myxomas and mucocutaneous pathologies are suspicious [4]. Although the rate of this association is low it is recommended to check the patients for Carney complex [5]. The association of FRS with multiple myxomas is not reported in the literature; however it is difficult to accept as an incidental coincidence the FRS occurrence in 6% of persons over 50 years of age [3].

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