DERMATOPATHOLOGY

FAMILIAL MULTIPLE ANGIOLIPOMATOSIS: A SERIES OF FIVE CASES

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Introduction: Familial multiple angiolipomatosis (FMA) is a rare heredity disorder. This benign tumoral affection originates from adipose tissue and blood vessels.

Objective: To study clinical and histopathological patterns of FMA.
Patients and methods: We conducted a retrospective study over a period of 6 years (2013-2018) at the department of Dermatology of Habib Thameur Hospital of Tunis.

Results: We collected five files. Males were affected more than females (sex ratio H/F=4). The average age was 46.2. A family history of similar lesions was found in all cases. Consanguinity was found in only one patient. Descendants were affected in one patient. Abnormality of lipid test was noted in only one patient. The trunk and the limbs were electively involved. Physical examination showed multiple, well-encapsulated, skin colored, mobile, soft, painless oval-to-round subcutaneous nodules. Histopathology reveals a circumscribed neoplasm composed of mature adipocytes and multiple thin walled blood vessels at the subcapsular area. Microthrombi were noted within many vascular lumens.

Discussion: FMA is a rare heredity condition. It is characterized by a progressive appearance of AL during the adulthood. Several authors have found an increased predilection in men. Familial angiolipomatosis has demonstrated autosomal dominant inheritance in our patients with a probable variable penetrance. Autosomal recessive transmission is rarely reported. Angiolipoma is a variety of lipoma in which the vascular component varies from 15 to 50 percent. FMA is usually a benign affection, however, liposarcomatous degeneration has previously been described. The association with visceral AL is extremely rare. On a genetic level, several studies of FML demonstrated abnormality in translocation of the high-mobility group protein isoform I-C (HMGIC) on chromosome 12 (12q15) and the lipoma preferred partner gene on chromosome 3. Surgical excision and liposuction of disfiguring or unusually large tumors offer cosmetic and practical benefit to the patients.