Familial multiple angiolipomatosis : A case report and review of the literature.

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ABSTRACT :

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Familial multiple angiolipomatosis is a rare benign condition inherited in an autosomal dominant manner. The disease is clinically characterized by numerous, discrete, well-encapsulated, round-to-oval, subcutaneous, rubbery masses with variable tenderness locate on the forearms, upper arms, thighs and abdominal wall. The histopathology is characteristic, with circumscribed subcutaneous mass composed of mature fat with numerous capillaries-sized blood vessels containing characteristic microthrombi without organization which frequently accentuated in subcapsular area. This study reported a patient with clinical features of familial multiple angilipomatosis.

Key words: Familial multiple angiolipomatosis

บทคัดย่อ:

กนกวรรณ เศรษฐพงศ์วนิช, ไพลิน พวงเพชร. รายงานผู้ป่วย FAMILIAL MULTIPLE ANGIOLIPOMATOSIS ร่วมกับทบทวนวารสาร วารสารโรคผิวหนัง 2554; 27: 193-197.

สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

Familial multiple angiolipomatosis เป็นโรคผิวหนังที่พบน้อยมาก และถ่ายทอดทางพันธุกรรมแบบ autosomal dominant มีลักษณะทางคลินิกที่สำคัญคือ เป็นก้อนกลมรีจำนวนมาก นุ่ม ขอบเขตชัดเจน บางรายอาจมีอาการเจ็บร่วมด้วย ในชั้นไขมันใต้ผิวหนัง บริเวณต้นแขน ต้นขา และหน้าท้อง โรคนี้มีลักษณะทางพยาธิวิทยาที่จำเพาะคือ ก้อนไขมันมีผนังหุ้มชัดเจน ซึ่งประกอบด้วยเซลล์ไขมันที่โตเต็มวัยร่วมกับเส้นเลือดฝอยที่เพิ่มจำนวนขึ้นโดยจะพบลิ่มเลือดขนาดเล็กอยู่ภายใน มักจะพบได้บ่อยบริเวณใต้ผนังหุ้ม ในรายงานนี้เป็นการนำเสนอผู้ป่วยที่มีลักษณะทางคลินิกของโรคนี้ร่วมกับมีประวัติครอบครัวที่มีบุตรชายมีอาการลักษณะเดียวกัน

คำสำคัญ: เนื้องอกไขมันเส้นเลือดที่ถ่ายทอดทางพันธุกรรม

Case report

A 49-year-old man came to our outpatient department for the evaluation of multiple subcutaneous masses on the trunk, arms and legs that had been presented since the age of 19. He had no underlying diseases. These lesions were not painful or pruritic. They grew slowly and gradually increased in number. The subcutaneous masses were not associated with antecedent medications or previous trauma. He noted that these lesions have caused cosmetic concern. His son also had a history of similar subcutaneous masses. Both of them had not sought for any treatment. No other family member has the same problem.

Physical examination :

Numerous, discrete, round-to-oval, 1-5 cm., rubbery, skin-colored subcutaneous masses on the trunk, arms and legs. There was no visible change of the overlying skin. No mucosal abnormality was noted. No lymphadenopathy of axillary, inguinal and cervical lymph node. (Figure 1)

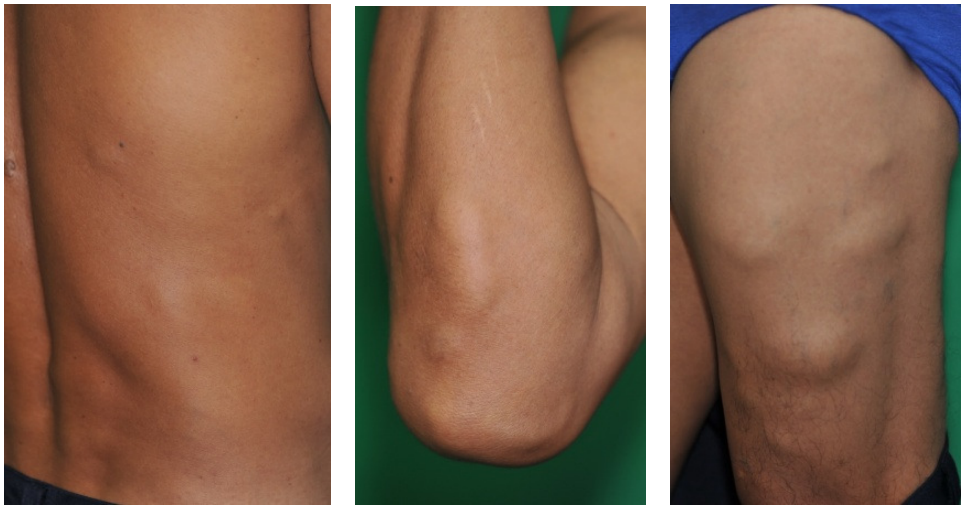

Figure 1

Investigation :

The skin biopsy specimen from the arm showed a well-circumscribed encapsulated subcutaneous mass composed of mature adipocytes and multiple thin-walled blood vessels at the subcapsular area. Microthrombi was noted within many vascular lumens. (Figure 2)

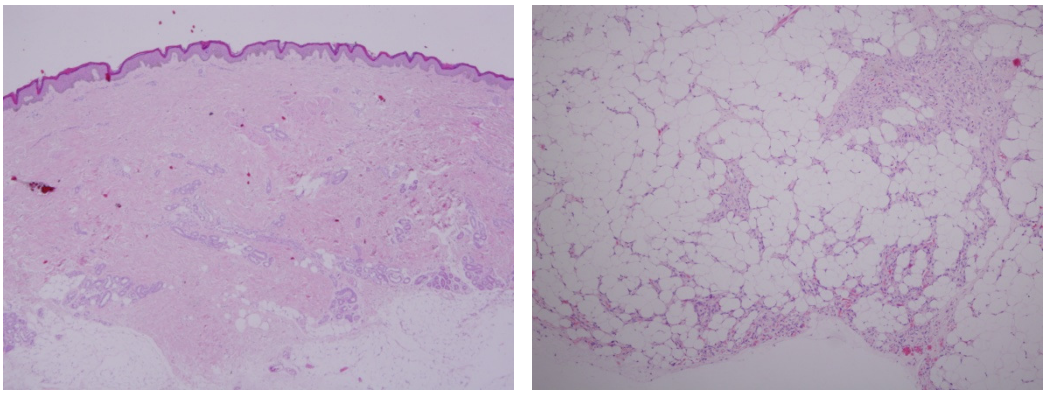

Figure 2

Disscussion :

Familial multiple lipomatosis (FML) is a very rare benign condition with prevalence of 0.002% in general population (1:50,000).[1,4]It is usually transmitted by the autosomal-dominant inheritance.[4] Some authors have found a slightly increased predilection in men with male:female ratio of 2:1.[1,2] In FML, lipomas initially appears in the third to fifth decades of life.[4] FML is

characterized by numerous, discrete, well-encapsulated, round-to-oval, subcutaneous, rubbery lipomas predominate on the forearms, thighs and trunk which usually spares neck and shoulders.[1]

Angiolipoma was first described as a distinct entity in 1912 by Bowen.[7,8]They account for 5-17% of all lipomas.[9,10] Most lesions often develop after puberty.[10,13,14] The size can vary from 1-4 cm. Angiolipomas tend to locate on the forearms, upper arms, thighs and abdominal wall.[7]The pathogenesis of angiolipoma is uncertain. Some authors suggest that it is a hamartoma or mesenchymoma[13], whereas others think angiolipoma results from the alteration within pre-existing lipomas triggered by stimuli such as repeated mild trauma.[10,17]Familial angiolipomatosis is relatively rare[10,14] approximately 5% of all cases are familial[16] and may be regarded as a subtype of FML. Angiolipoma may be clinically differentiated from lipomas by tenderness[10]. There were a few reports that patients with familial angiolipomatosis had no tenderness.[5,14] Previously reported familial multiple angiolipomatosis are listed in Table1.[5,14,15,16] Our patient had asymptomatic subcutaneous masses and histological examination supported the diagnosis of angiolipoma.

Table1 Familial multiple angiolipomatosis included with this patient(case number 8).

Case	Year	Age/Sex	Age of onset	Size(cm)	Location	Tenderness
1	1977	33/Male	19	0.5-3	Trunk, extremities	No
2	1977	37/Male	35	0.5-4	Trunk, upper arms	Slightly tender
3	1980	Adolescent/Male	1	Large	Wrists, knees, ankles	No
4	1987	18/Female(pregnancy)	7 month of GA	0.5-1.5	Forearms, upper arms, midback, thighs	Tender
5	1987	37/Male	32	0.5-1.5	Left arm, abdomen	Tender
6	1999	42/Male	-	1-3	Forearms, wrists, chest wall	Tender
7	2007	80/Male	30	1-10	Abdomen, extremities	No
8	2010	49/Male	19	1-5	Trunk, extremities	No

Histopathologic evaluation of angiolipoma shows circumscribed subcutaneous mass composed of mature fat with numerous capillaries-sized blood vessels containing characteristic microthrombi without organization which frequently accentuated in

subcapsular area.[6] Vascular component varies from 15 to 50%.[5,11]

Several cytogenetic studies of FML demonstrated abnormality in translocation of chromosome 12q13-15 and 6p21 with high-morbidity group A2 (HMGA2) gene and HMGA1 family of genes respectively.[1] Subcutaneous angiolipomas showed normal karyotypes[12] and specific gene rearrangements in familial angiolipomatosis have not been described.[5,11,12]

Familial lipomatosis and angiolipomatosis do not require medical or surgical treatment. Spontaneous regression or malignant degeneration of the tumor is rare.[4,7] Surgical excision and liposuction of disfiguring or large tumors may be benefit to the patients. Local recurrence after excision are common with large lipomas.[13]

We reported a case of familial multiple asymptomatic angiolipomatosis who underwent surgical excision of some disfiguring lesions.

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