Familial Multiple Lipomatosis

AİLEVİ MULTİPL LİPOMATOZİS

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Summary

Familial multiple lipomatosis (FML) is a very rare disease. Usually daylife of the patient is not affected. But if the lipomas are multiple and big, the patient suffers from particular difficulties in carrying out everyday physical tasks. We have operated a man with multiple lipomas in his extremities and the trunk because of the difficulty in wearing pants. The family of the patient was then investigated over three generations related to FML. We found out that the disease is transmitted by the autosomal dominant route of inheritance.

Key Words: Multiple lipomatosis, Hereditary disease

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Özet

Ailevi multiple lipomatosis (AML) çok nadir görülen bir hastalıktır. Genellikle günlük hayatı etkilemez. Ancak lipomalar ekstremitede çok sayıda ve büyük oldukları zaman bazen sorun yaratabilirler. Gövde ve ekstremitelerinde yaygın olarak lipoma bulunan, sağ uyluk çapında ileri derecede artma nedeniyle pantolon giymede zorluğu olan ve bu nedenle buradaki lipomaların bir kısmını ameliyatla çıkardığımız bir hastanın oldukça geniş olan ailesi 3 jenerasyon boyunca lipomatozis yönünden araştırıldı. Bu araştırma sonunda AML'in otozomal dominant geçiş gösterdiği belirlendi.

Anahtar Kelimeler: Multiple lipomatosis, Herediter hastalık

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Lipoma is one of the most common benign soft tissue tumors. But familial multiple lipomatosis (FML) is rare. In the majority, it has been observed that the disease occurs more frequently with autosomal dominant inheritance (1-5). Case with recessive inheritance have only rarely been reported (2,6).

In FML, lipomas are painless and patient is no longer troubled by the disease (4,5). Usually FML is mentioned in dermatology textbooks. But generally patients with lipomas refer to the orthopaedics outpatient clinics and sometimes tumors have been excised. Although in FML lipomas do not create any problem, but sometimes just like being in this case multiple lipomas on the right thigh cause enlargment so that the patient can not wear pants. We tried to solve this problem by excising some of the lipomas. Our patient has got 4 brothers and 8 sisters and a lot of (60) nephews. We made the pedigree of the our patient's family and we tried to find out the inheritance of the FML.

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Case Report

N.A, 39 years old, male, security personal in a factory. When he was 14 years old, he noticed a tumor on his right thigh first. It became larger slowly and became multiple. Then it appeared on his left thigh, both arms-forearms and body. Recently a tumor arised on the proximal part of the right leg. Lipomas on the right side were more and bigger than the lipomas on the left side. The tumors were movable and painless. Because of difficulties in wearing pants our patient wanted us to excise some of the tumors on the right thigh. Our patient does not use alcohol, but smokes. He was married and has 3 daughters and 1 son. His eldest child was 13 years old. None of his children has tumor on their body.

Clinical features ; on his both thighs, on the dorsal and the lateral parts of the trunk, on the arms there were multiple, painless, mobile, subcutaneous tumors that were 2-10 cm in diameter. Especially right thigh diameter was extremely large because of the multiple big lipomas (Figure 1). In addition there were two tumors on the xyphoid and on the proximal medial part of the right leg that were painless, mobile and 3x4 cm in diamater.

Laboratory investigations; triglycerides, cholesterole, glucose, white blood cell count, hemoglobine, sedimantation rate, renal and hepatic function tests are in the normal range.

In operation under spinal anesthesia; we excised multiple tumors that were localized subcutaneously on the anteromedial and anterolateral part of the right thigh, were looking like a bunch of grapes encapsulated and bright yellow in colour (Figure 2).

Histopathologic diagnosis also revealed a lipoma containing mature fat tissue elements surrounded by a thin fibrous capsule (Figure 3).

Family History; The disease had been seen first on the neck of the father of our patient. The parents of our patient were not relatives and they were from the different places. The married couple have 12 children, 2 of them had died when they were a baby. Another 3 of them had died because of other diseases when they were 23, 24, and 45 years old respectively. 4 female and 2 male children have got the disease FML. The siblings have 60 children. One of these children who was 25 years old, female has FML whose mother has the disease too (Figure 4). Except our patient, patients in the family have a little lipomas on them arms, forearms and thighs. And in these patients the lipomas appeared in the 3rd and 4th decade of life.

Discussion

FML is a very rare benign condition. Usually it is transmitted by the autosomal dominant route of inheritance (1-5). Cases with recessive inheritance have only rarely been reported (2,6). Some authors (7) accept that FML is seen especially in males. But female to male ratio is usually close to each other (1,2,4).

Our patient has a large family that has four brothers, 8 sisters and 60 nephews. The disease had been seen in our patient's father first. Between spouses in the family there was no relationship. The disease was established in 4 sisters and 2 brothers of the family. So that approximately half of siblings was affected. Four persons of the family died before 3rd decade, because of other diseases. So that if they have been living now, they may be affected too. One of the female nephews of our patient whose mother was affected has the disease. We believe that when the children grow older, the number of affected individuals will increase. So that we believe that FML is an autosomal dominant inherited disease. In addition we believe that there is no sex prevelance.

There are some reports that FML is related to hyperlipidemia (3,8). But in this case plasma lipid levels were in the normal range. Furthermore FML can be seen with peripheral neuropathy together (5,6). But in our patient there was'nt such a condition.

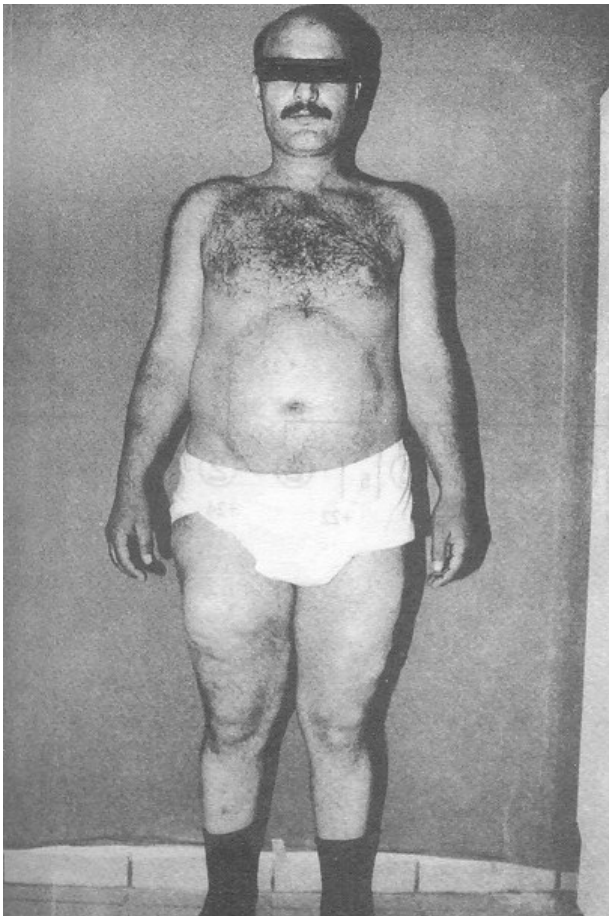

Figure 1. Photograph of the patient.

Figure 2. Lipomas excised in operation.

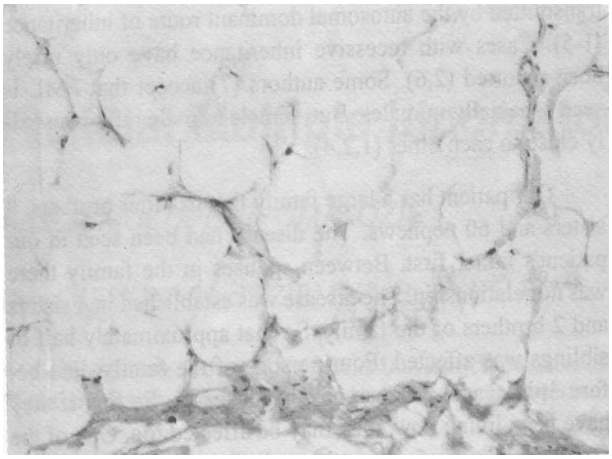

Figure 3. Histopathologic findings (HE, 200X).

Lipomas are generally painless and do not affect the daylife activities (1,2). In this case there was no pain related to lipomas. But because of the multiple lipomas on the right thigh, the diameter of the right thigh enlarged. So the patient had a difficulty in wearing pants. On this occasion according to patient's desire, some of the lipomas on the right thigh were excised.

According to the literature we know that lipomas of the FML appear usually in 3rd decade, rarely in 4th or 5th decade (1,2). Leffell and Braverman (9) reported that lipomas in the FML are seen on the neck, are movable and solitary. But there are some reports that lipomas are seen on the trunk, arm, forearm and the thigh (1,2,4).

According to the family history the disease had been seen only on the neck of the father of our patient. For the other patient members of the family lipomas were localized on the trunk, on the upper extremities and on the thighs.They were a little in number, had not caused any complaint. In additionaly these lipomas had been arised in the 3rd and 4th decade. Contrary to the other patient members of the family, in our patient first lipoma was noticed when he was 14 years old, namely in the 2nd decade. We thought that because of the early set up of the disease, lipomas were larger in number, bigger and diffuse. Because of the first set up of the lipomas on the right side , lipomas on the right side of the body were larger in number and bigger than lipomas on the left side. Reverse to the Leffell and Braverman's (9) reports in FML lipomas can be diffuse, not only localized on the neck.

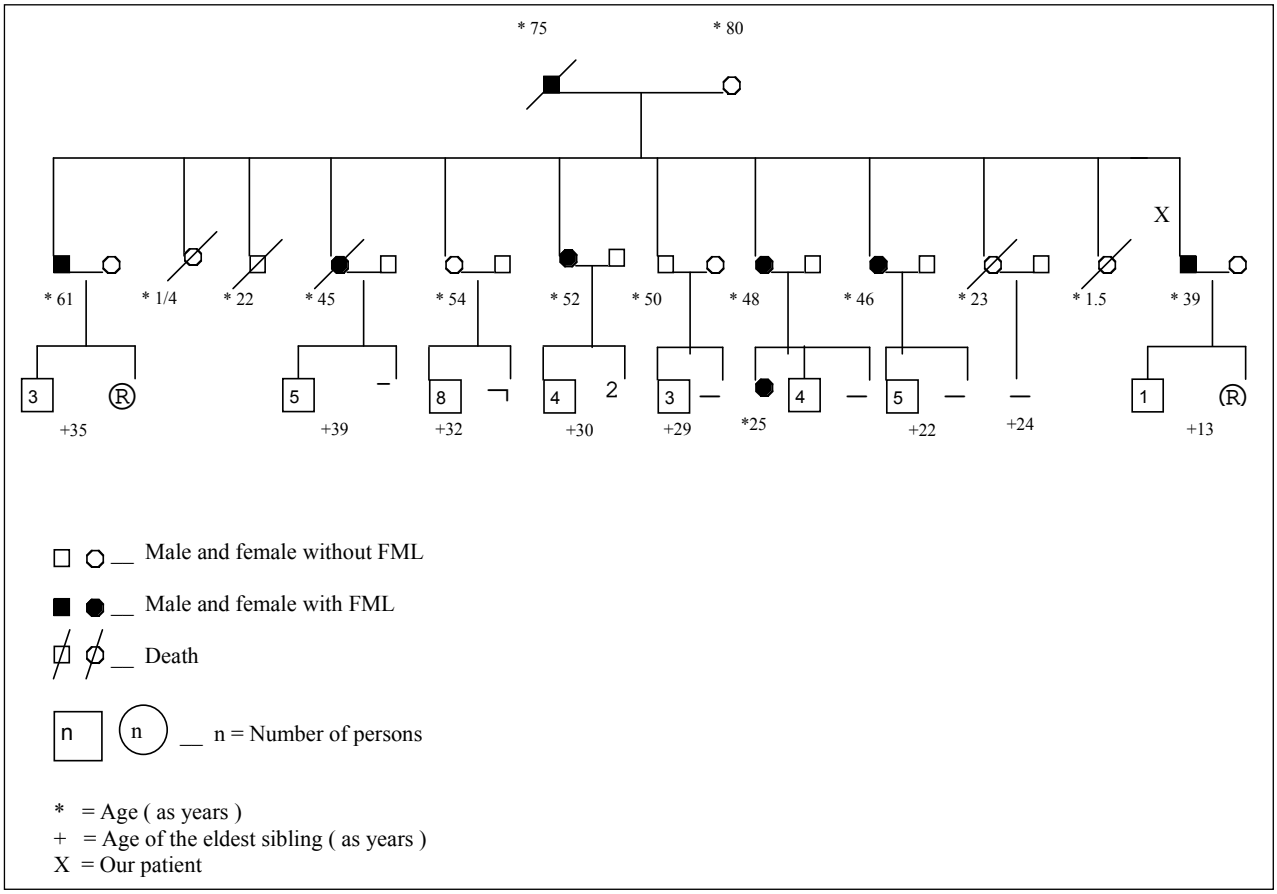

Figure 4. Pedigree of the family.

In this study, we found that FML is an autosomal dominant inherited disease. And if the lipomas are seen at the early age, they can be larger in number, bigger and can be diffuse. And sometimes they have to be excised surgically.

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