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22	26
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13	
Simvastatin	, 1
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가,	가	3)	가
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62

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가
17

1 17

(Fig. 1) (Fig. 6, 7)

(Fig. 2), (Fig. 3) (Fig. 4, 5)

26 8

(foamy cell)가

(Fig. 8).

1999 1
6

Simvastatin

, 2 , 1

, 가

451mg/dl, 51mg/dl, 410mg/dl

(Table 1).

2, 3, 4

22

1 ~ 5cm

가

(Fig. 1-5).

(Fig.

6, 7).

469mg/dl,

(atherosclerosis)

111mg/dl,

408mg/dl 가

, Yamamoto ⁶⁾

500 1

Simvastatin

Table 1. Pedigree showed autosomal dominant transmission of the hypercholesterolemia(: patient).

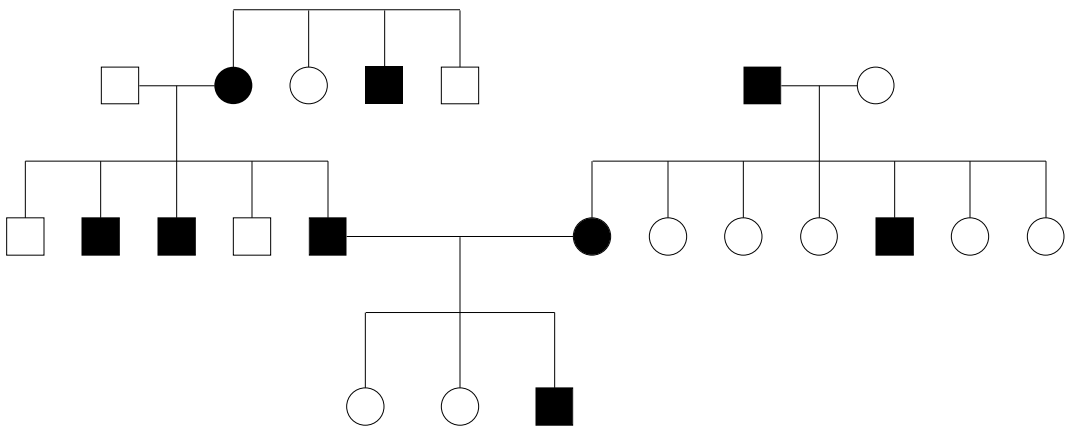




Fig. 1-5. Preoperative and intraoperative photographs showed multiple various sized palpable masses on the extensor tendon sheaths of hand (Fig. 1), triceps tendon of both elbows (Fig. 2), subcutaneous xanthomas on the prepatellar area of both knees (Fig. 3) and both malleoli, Achilles tendon and peroneal tendon sheath of both ankles (Fig. 4, 5)(arrows).

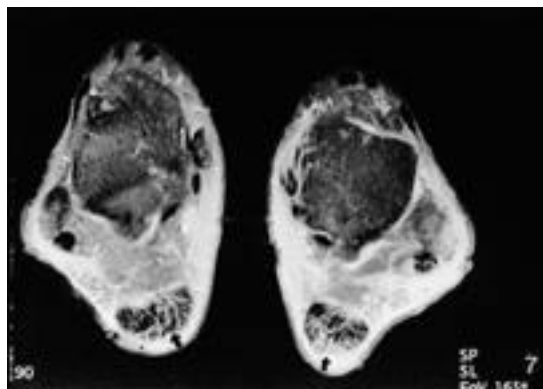


Fig. 6, 7. Sagittal T1-weighted MRI showed a homogeneously enlarged Achilles tendon with increased signal intensity caused by xanthomatous material and parallel striation of low signal intensity caused by the collagen fibers of the tendon (Fig. 6). Axial T1-weighted MRI revealed the characteristic diffuse, stippled pattern of a xanthoma of the Achilles' tendon (Fig. 7)(arrows).

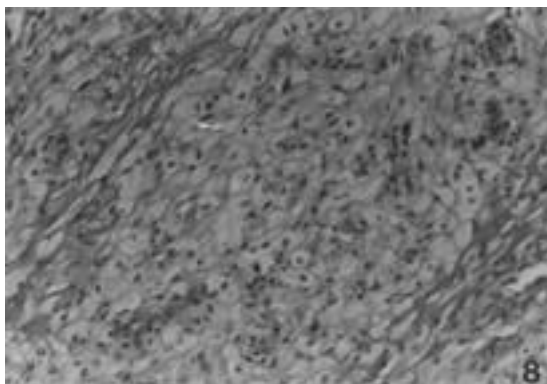


Fig. 8. A biopsy specimen showed foamy xanthoma cells (blank arrow), multinucleated giant cells (filled arrow) and cholesterol crystals, dispersed between the collagen fibers of the Achilles tendon (H-E × 200).

6). 가
가 , 가
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30 ~ 40
가
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(heterozygote)
400mg/dl
(homozygote)
30 ~ 40
50 80%
22 30% 10
4:3
Fahey³⁾ 가

Table 2. Differences between solitary xanthoma of tendon or its sheath and multiple xanthomatosis of tendon with hypercholesterolemia²⁾

	Solitary xanthoma	Multiple xanthomatosis
Number of lesion site	single or a few	multifocal
Frequently involved site at hand	palmar side of finger	extensor surface of metacarpophalangeal joint
Symmetrical involvement	not always	almost always
Involving tissue	tendon and its sheath	tendon and its sheath, subcutis
Skin changes	normal skin color with palpable nodules	scattered pattern of yellowish discoloration of skin with nodules
Associated disease	none	hypercholesterolemia with premature coronary atherosclerosis
Hereditiy	none	autosomal dominant

90%

26

가 8

22

8

20

(Fig. 1),

(Fig. 3)

(Fig. 4)

, 20

17

, 13

173

15

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^{1,3)} Klemm ⁵⁾

62%

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. Andres ¹⁾

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Abstract

**Multiple Xanthomatosis in Familial Hypercholesterolemia Patient
- A case report -**

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We experienced the case of familial hypercholesterolemia with multiple xanthomas which was treated by combined surgical and medical therapy. He was 26-year-old male patient of familial hypercholesterolemia with multiple xanthomas in 22 sites on whole body, and was treated by 17 surgical excisions of the xanthomas and by medical therapy of the hypercholesterolemia.

There was a normal healing process of the surgical wounds. Continual postoperative medical therapy of the hypercholesterolemia was done. There was no recurrence of the symptoms during more than 13 months of follow-up. But the serum level of the cholesterol was not lowered significantly, so we are treating him with drug therapy.

Familial hypercholesterolemia is caused by a specific disorder of lipid metabolism, and is characterized by increased LDL cholesterol, tendon xanthomas, coronary disease associated with autosomal dominant transmission. Xanthomas usually appear in the second decade of life with familial hypercholesterolemia which may have high risk for premature coronary atherosclerosis, which might be prevented with early diagnosis and medical treatment. So, orthopedic surgeons do not only excise the xanthomatosis surgically but also can diagnose the underlying hypercholesterolemia.

Key Words : Xanthomatosis, Familial hypercholesterolemia

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