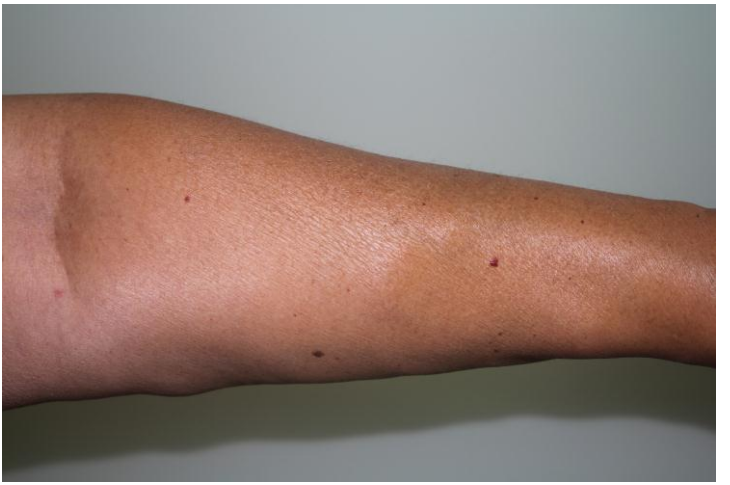
Case 8
A 62-year-old Thai woman from Bangkok.
Chief complaint: Multiple non-painful nodules on both forearms and thighs.



Present illness: This patient came to our clinic with history of multiple bumps and lumps of her scalp, forearms and thighs. The lesions have been present since age of 29. She noticed the first lesion on her scalp. Over the years, similar lesions continued to appear on both forearms and thighs. These lesions gradually enlarged, but some were stable in size. All of the lesions are asymptomatic, but bother the patient cosmetically. She does not drink alcohol and does not have thyroid diseases, psychiatric problems, or cancers.

Past history
Her medical history is significant for diabetes mellitus, non-alcoholic fatty liver disease, and gastroesophageal reflux disease.

Family history
Her father (individual I.1, figure 1) who deceased several years ago also had similar lesions like hers. Her youngest brother(individual II.7) has multiple lumps like our patient. Two of her sisters (individual II.1, 5) have similar but less extensive involvement.

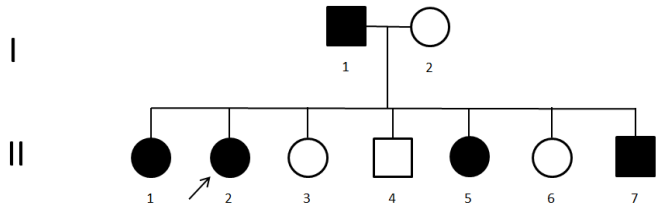
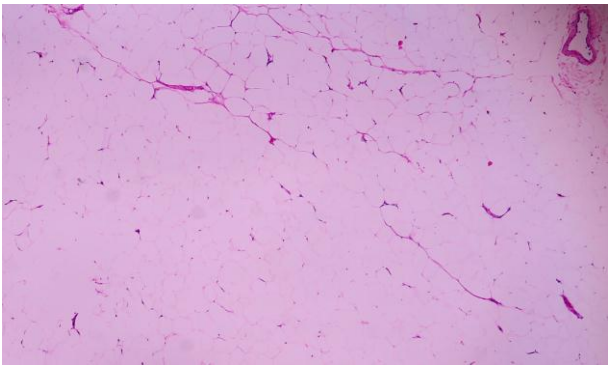


Figure 1. Family pedigree of our patient

Skin examination
Multiple, symmetrical, discrete, non-tender, soft, subcutaneous nodules of sizes varying from 1-7 cm on scalp, both thighs and forearms, sparing trunk.

Histopathology: Large tumor mass composed of mature adipocytes surrounded by thin connective tissue capsule.



Diagnosis: Familial multiple lipomatosis
Treatment: Patient education and excision of selected lipomas

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Discussion

Familial multiple lipomatosis (FML) is a rare inherited disease, characterized by multiple discrete, encapsulated lipomas involving in several members of family. The predilection sites are on the trunk and extremities, especially forearms and thighs, with relative sparing of the head and shoulders.[1-3] The lesions usually appear around the third decade of life, increasing in number with no tendency to involute. The number and size of lesions can vary considerably in each individual. Malignant transformation has never been reported.[3] The disease is not related to abnormal lipid metabolism, and weight loss does not appear to alter the disease.[1,3]

The disease is an autosomal dominant inheritance. Although the exact pathogenesis is yet to be revealed, Mrózek K et al, found in their series that major abnormal karyotypes of lipomas were translocations between chromosome 12 and other autosomes.[4] It is now believed that lipoma results from rearrangement of chromosome 12q13-15.[5-7] However, such genetic research has been conducted mostly from normal lipomas which may not represent the same defect as lipomas in familial multiple lipomatosis.

Other diseases associated with multiple lipomas include benign symmetric lipomatosis (Madelung's disease), adiposis dolorosa (Dercum's disease), infiltrating or diffuse lipomatosis, congenital lipomatosis, and syndromes that can exhibited feature of multiple lipomas such as Proteus syndrome, Gardner syndrome, Bannayan–Riley–Ruvalcaba syndrome, and Cowden disease.[1,8] Madelung's disease has predilection on head, neck, shoulder, and proximal upper extremities.(Figure 2) These lesions are more

diffuse and infiltrative than in FML. Madelung's disease often occurs in middle-age men, and is associated with excessive alcohol consumption. Dercum's disease is characterized by multiple painful tumors on trunk, forearms, and thighs. (Figure 2) The disease is more common in postmenopausal women, and is associated with weakness and psychiatric disorders. In infiltrating or diffuse lipomatosis, the tumor is non-encapsulated mature fat that can infiltrate deep to muscle, fascia, and bone. It commonly affects lower extremities, and usually occurs before age of 30. Congenital lipomatosis is different in onset which occurs in early life. It is characterized by large subcutaneous lipomas primarily on trunk and chest wall, and the tumors are composed of immature fat cells.

Even though the FML lesions are asymptomatic, many patients seek treatment mainly for cosmetic reason. Treatment is mainly removal of tumors. This can be done by simple excision, liposuction, or endoscopic tumor removal.

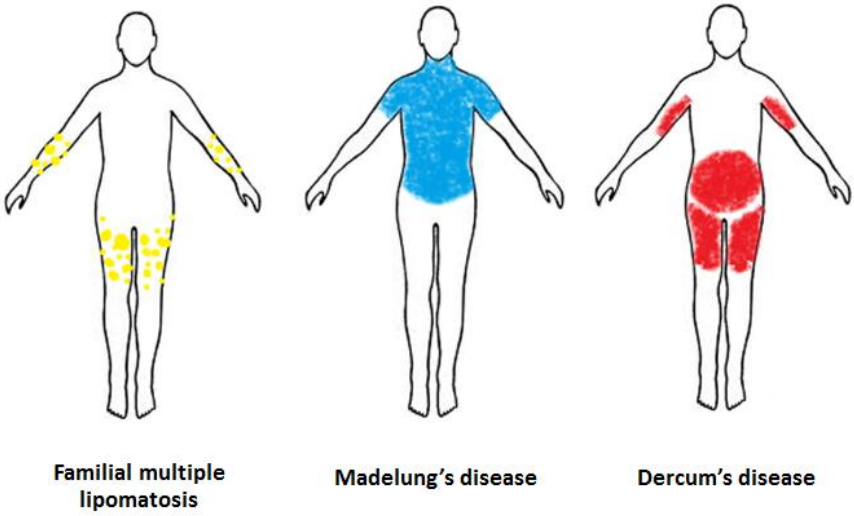

Figure 2 Comparison of distribution of lipomas in different diseases

References

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