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CASE STUDY

FAMILIAL MULTIPLE LIPOMATOSIS REVISITED

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ABSTRACT

Familial multiple lipomatosis (FML) is a rare hereditary syndrome of multiple encapsulated lipomas which are seen over trunk and extremities. We report a case of a gentleman who presented for evaluation of multiple swellings over body that caused cosmetic impairment. Such larger lesions were excised. Diagnosis of familial multiple lipomatosis (FML) was made based on characteristic clinical and family history.

INTRODUCTION

Solitary or multiple lipomas, composed of mature fat, represents by far the most common benign mesenchymal neoplasm occurring throughout the whole body. Most cases with lipomas may be ignored due to their silent nature. It can arise in any location in which fat is normally present and hence known as the "universal tumour." Clinical manifestations depend on the size and location of the growth. In most patients, symptoms are few or absent. Multiple lipomas over the body occur in specific hereditary syndromes. Familial multiple lipomatosis (FML) is one of them.

CASE HISTORY

A 55 year-old Gentleman presented with history of multiple widespread nodules over his body. The lesions began to appear at the age of 20 years and were long standing with more lesions appearing over the period of time. On physical examination, more than 60 painless subcutaneous, soft, mobile nodules were observed in the arms, thighs and abdomen distorting the affected areas. His past medical history was unremarkable. He had history of open appendectomy and bilateral inguinal hernia repair. Similar but less extensive lesions were observed in his two younger brothers. Patient had excision biopsy of some swellings over the abdomen, thigh and arms.

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The histopathological examination report confirmed these swellings as lipoma. Based on the characteristic clinical history, family history and histopathology, the diagnosis of familial multiple lipomatosis (FML) was made.

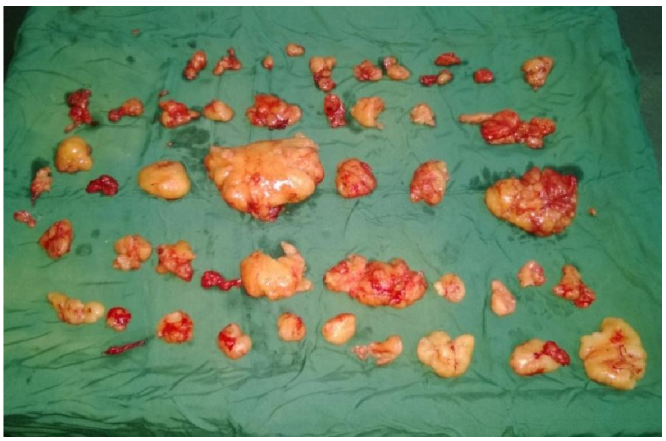
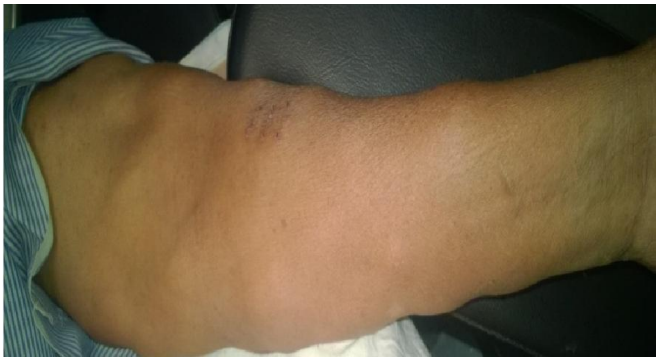


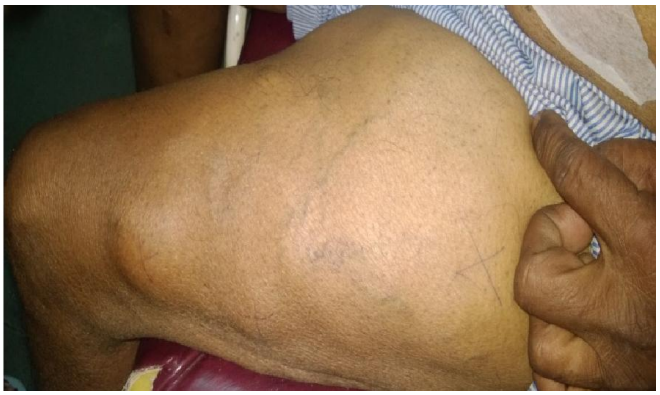
Figure 1. Excised Lipomas

DISCUSSION

Familial multiple lipomatosis is an uncommon benign hereditary syndrome with an incidence of 0.002%. Brodie first described FML in 1846 (Muller, 1951) and Blaschko noted its familial association in 1991. (Toy, 2003) Later, Murchison shed light on this syndrome: Familial multiple lipomatosis (FML). (Murchison) It is seen twice more commonly in men.[4]FML usually appear in third decade of life but can occur at an earlier age. (Stephens and Isaacson, 1959) It is usually

transmitted as an autosomal dominant disorder affectingseveral members in a family (Mohar, 1980).





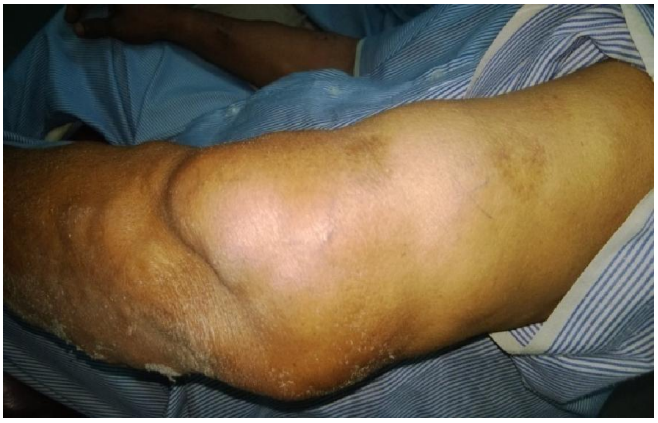


Figure 2, 3 & 4. Multiple lipomas over the extremities

In 1993, the genetic defect was isolated to chromosome 12q15, which encodes the high-mobility-group protein isoform I-C (HMGIC). (Schoenmakers *et al.,* 1995) It is now believed that this disorder results from a translocation involving HMGIC on chromosome 12 and the lipoma preferred partner gene (LLP) on chromosome 3. (Mrozek *et al.,* 1993) These lipomas are painless. The number of tumors in a patient may vary considerably. Although tumors usually range in size from a few millimeters to 6 cm in diameter, lipomas as large as 25 cm have been reported. (Lefell and Braverman, 1986) Spontaneous regression and malignant degeneration are rare. No association with disorders of lipid metabolism. (Rubinstein *et al.,* 1989) Treatment can include simple excision, endoscopic removal, or liposuction (Ronan and Broderick, 2000).

Surgical excision is the treatment of choice for large lesions causing functional and cosmetic impairment.Recurrence is also reported but very few cases.Differential diagnosis of diseases with subcutaneous lipomas include Dercum's disease, Madelung's disease, Cowden's syndrome and Bannayan-Zonana Syndrome.Lipomatosis can be easily confused with neurofibromatosis or Legius syndrome, and all cases of multiple lipomatosis merit taking a detailed family history. Familial Multiple Lipomatosis (FML) is often confused with Dercum's Disease. But the latter has asymmetrical lipomas over the body. Dercum's Disease is classically seen in obese postmenopausal females having painful deposits of adipose tissue. Asthenia and mental disturbances are associated symptoms. Madelung's disease is characterized by symmetrical, painless lipomas and associated with chronic alcoholism in 90% of cases. In Bannayan-Zonana Syndrome there is accumulation fatty tissue with developmental delay, hypotonia and lipid myopathy. These patients present at an earlier age. Cowden's Syndrome has other skin tumours such as facial trichilemmomas, fibromas etc. present.

CONCLUSION

FML though rare should be considered as differential diagnosis in patients with multiple lipomas. In our case since deposition of fat was asymmetrical and asymptomatic, Dercum's disease and Madelung's were immediately excluded from differential. Classical history led to diagnosis of FML.

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