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Multiple Symmetric Lipomatosis

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Multiple symmetric lipomatosis (MSL) is an extremely uncommon disorder. In the medical literatures about 200 cases have been reported. MSL is not associated with other generalized lipomatous disorders, nor are these patient to be necessarily obese. The cause of MSL is unknown. The disorder usually occurs in middle-aged males and there is frequently a history of alcoholoism. Some instances of familial occurrence have been reported, but the majority of cases are sporadic. Two cases of MSL are presented.

Key Words: Multiple symmetric lipomatosis

INTRODUCTION

Multiple symmetric lipomatosis (MSL), also called Madelung's disease, Launois-Bensaude syndrome or benign symmetric lipomatosis, is an uncommon disorder of fat distribution. It is characterized by progressive enlargement of subcutaneous symmetric fat masses on the neck, shoulders, chest, axilla, abdomen, and groin, and also in deep structures such as the retroperitoneum and mediastinum. And it predominates in middle aged males.

The earliest description of the disorder was by Brodie in 1846 (Brodie, 1846), but it was Madelung who collected a series of 33 patients, including three of his own, and reported them as a specific disease in 1888 (Madelung, 1888). Launois and Bensaude presented a complete review of the literature included 65 cases in 1898 (Launois et al., 1898).

The cause of MSL remains unknown. In recently published reports, the association of MSL with hyperuricemia, hyperlipidemia, alcoholism, cirrhosis. hypertension, obesity, diabetes, impaired glucose tolerance, renal tubular acidosis, thyroid abnormalities, and peripheral neuropathy has been demonstrated. (Argenta et al., 1981; Green et al., 1983; Enzi et al., 1983 & 1985; Rhoads et al., 1986).

We herein document our recent experience with two patients of multiple symmetric lipomatosis. And these cases are discussed together with the cases reported in the literatures.

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CASE REPORT

Case 1

A 63-year-old man was admitted to the Soon Chun Hyang University Hospital on September 17, 1984, complaining of multiple palpable masses on the surface of the body.

Four years earlier, he had begun to notice a single mass on the back of the neck. Over 3 years before admission, similiar masses developed progressively on the trunk, upper extrimities and inguinal areas, and increased gradually in size. He used to smoke one pack of cigarettes daily for 35 years and drink alcohol heavily. There were no other previous medical or surgical illness. Family history was unremarkable.

On admission, he was found to have bilaterally symmetric soft subcutaneous masses in the neck, upper extremities, trunk, back and inguinal area (Fig. 1). The masses varied in size from 1×1 cm to 3×3 cm Table 1). The results of physical examination were otherwise normal. The blood pressure was 120/60 mmHg. The pulse rate was 86/minute and the respiratory 20/minute. The height was 164 cm. and the weight 57 kg. Values for electrolytes, uric acid, fasting blood glucose, complete blood count and urinalysis were normal. Also a chest roentgenogram was normal. The serum glutamic oxaloacetic transaminase was 17.2 U/L and the alkaline phosphatase 42.9U/L. The cholesterol

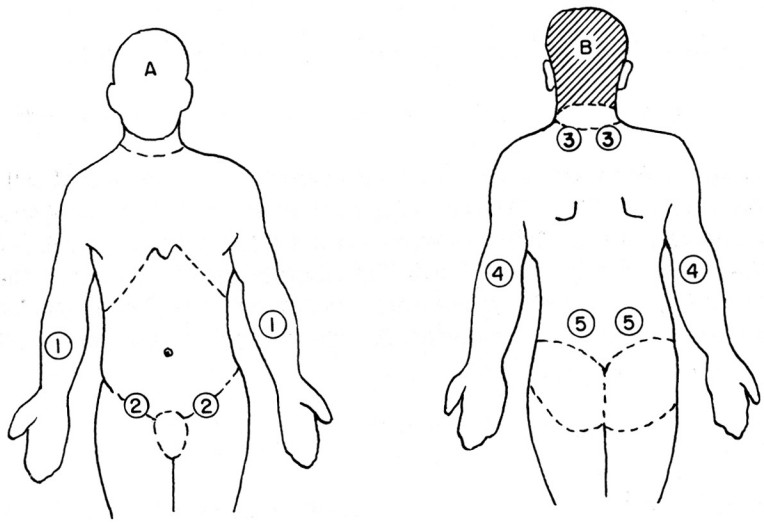

Fig. 1. Localization of lipomatous masses in case I
A: anterior aspect B: posterior aspect

Table 1. Size of lipomatous masses in 2 MSL patients

CASE I		CASE 2			
No.	Size (cm.)	No.	Size (cm.)	No.	Size (cm.)
1	4×5	1	3×4	7	4×5
2	10×7	2	3×4	8	2×2
3	7×5	3	3×4	9	4×5
4	3×4	4	4×5	10	3×3
5	4×5	5	4×5	11	1×2
		6	5×6	12	1×2

*The number of Fig. 1 and Fig. 3 is the same as the number of the Table 1.

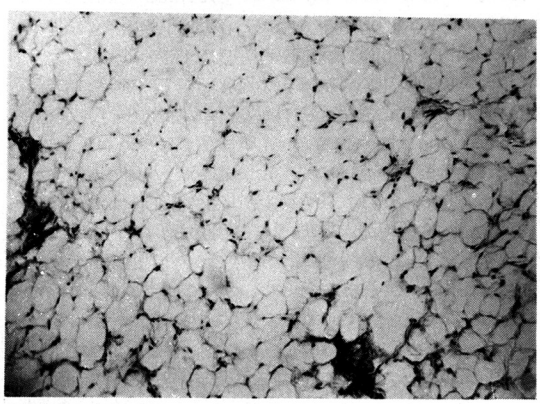

Fig. 2. Histologic section demonstrating mature fat cells showing only slight variation in size and shape in case I (Hematoxylin-eosin stain, ×100)

was 135.0 mg/dl, the total lipid 536.0 mg/dl, the triglyceride 108.6 mg/dl, the ß-lipoprotein 223 mg/dl, the pre ß-lipoprotein 119 mg/dl and the α-lipoprotein 193/dl. An electrocardiogram demonstrated low voltage tendency in limb lead, incomplete right bundle branch block and anterolateral myocardial abnormality.

On the 3rd hospital day, the masses on the neck, both forearms and arms, and both inguinal area were excised.

Specimens were encapsulated tumors which were shelled out easily and grossly resembled typical lipomas. Microscopic examination of the specimens revealed characteristic lipoma (Fig. 2).

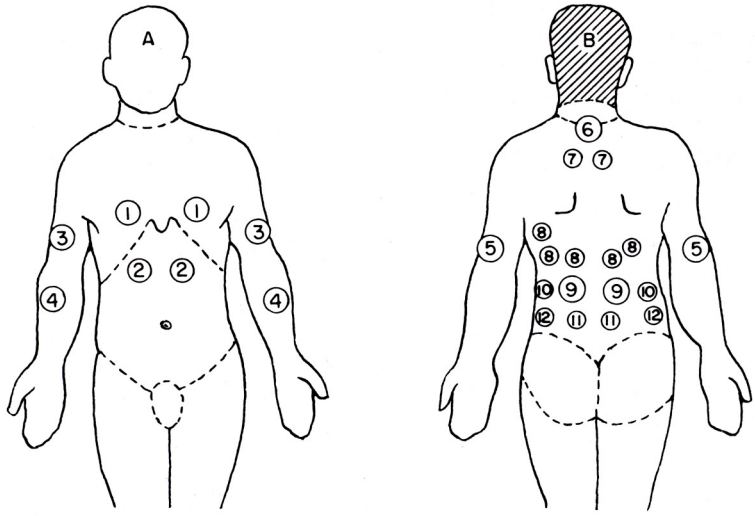

Fig. 3. Localization of lipomatous masses in case 2
A: anterior aspect B: posterior aspect

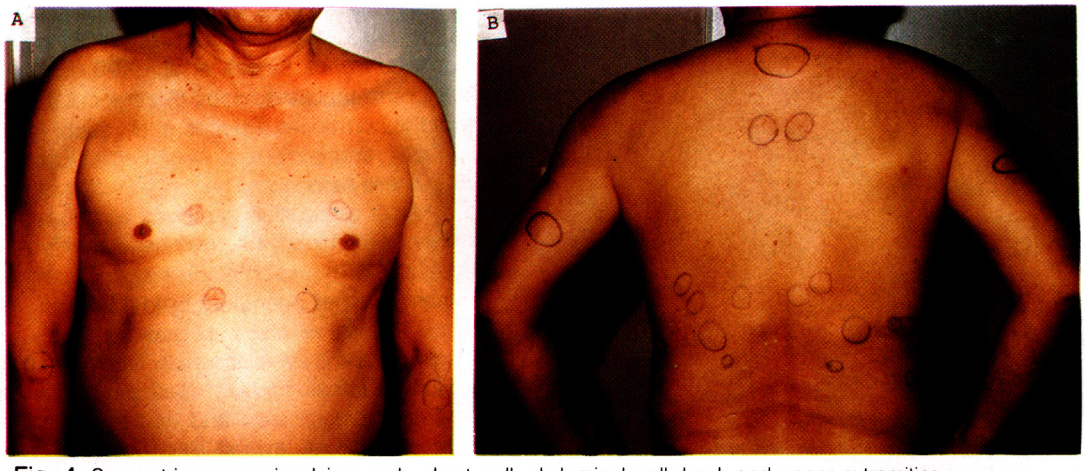

Fig. 4. Symmetric masses involving neck, chest wall, abdominal wall, back and upper extremities.
A: anterior aspect B: posterior aspect

Case 2

A 62-year-old man was admitted to the Department of Internal Medicine of the Soon Chun Hyang University Hospital with hypertension and multiple masses on the surface of the body.

He had been treated for hypertension for 10 years. He had had multiple masses on the neck, chest will, abdominal wall and extrimities for about 9 years. A single mass on the back of the neck was noted 9 years ago. Over 8 years before admission similer masses ceveloped progressively on the neck, chest wall, abdominal wall, upper extrimities. He smoked 1/2 pack of cigarettes daily for 30 years and drank moderately. There was no history of surgical and medical illness. Familial history was unremarkable.

On admission, he complained of headache, motor weakness of the upper extremities. and multiple

palpable masses on the surface of the body. Blood pressure was 160/100 mmHg, pulse rate 95/minute, respitatory rate 20/minute. The height was 173 cm., and the weight 76 Kg. Physical examination revealed bilaterally symmetric soft and nontender subcutaneous masses on the neck, chest wall, abdominal wall, back and upper extremities (Fig. 3.4). The masses varied in size from 2×2 cm. to 3×3 cm. (Table 1). The remainder of his physical examination revealed no abnormalities.

Laboratory examinations on admission revealed the cholesterol of 253 mg/dl, the total lipid 600.0 mg/dl, the triglyceride 123.2 mg/dl, the beta lipoprotein 317.0 mg/dl, the pre ß lipoprotein 138.0 mg/dl and the α lipoprotein 145 mg/dl. Serum electrolyte, serum glutamic pyruvic oxaloacetic transaminase, alkaline phosphatase, blood urea nitrogen, creatinine, calcium, complete blood count and fasting blood sugar were all within normal limits. Urinalysis showed +++ for protein, negative for glucose and a few finely granular cast per high power field. A chest roentgenogram and electrocardiogram were normal. A renal ultrasonographic study showed a round hypoechoic mass in the upper pole of right kidney (2.7cm×3cm) and in the lower pole of left kidney (2.9cm×3cm).

On the 4th hospital day, one mass on the left arm and two on the anterior chest wall were excised under

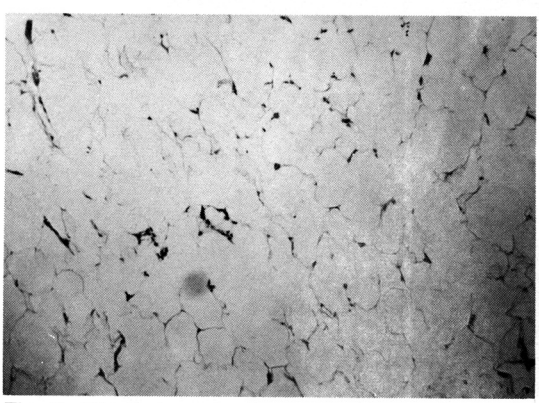

Fig. 5. Histologic section demonstrating mature adipose tissue in case 2 (Hematoxylin-eosin stain, ×100)

the local anesthesia. The specimens were encapsulated tumors and grossly resembled typical lipomas. Microscopic examination of the speciman revealed characteristic lipoma (Fig. 5).

DISCUSSION

Multiple symmetrical lipomatosis is an uncommon disorder manifesting as symmetric abnormal deposits of adipose tissue in the subcutaneous layer or in the deep structures. It is also called by Madelung's disease, Launois-Bensaude adenolipomatosis, and benign symmetric lipomatosis.

MSL occurs more frequently in countries around the Mediterranean area. The disease affects almost exclusively adult males usually with a background of excessive alcohol intake as in case I and liver disease. The onset of disorder usually occurs in middle age groups, but it has been described in other age groups, ranging from 20 to 68 years old (Enzi, 1984; Enzi et al., 1986). The cause of MSL has not been determined. Several attempts have been made to link MSL to systemic metabolic and biochemical defects (Goldwell et al., 1972.), and other endocrine and neurologic disturbances (Rhoads et al., 1986; Enzi et al., 1986). The exact cause of the condition remains obscure. Also laboratory studies are largely inconclusive. MSL has been reported in association with various disorders such as hyperuricemia and Type IV hyperlipidemia (Green et al., 1970; Enzi et al., 1983), alcohclism (Enzi et al. 1983; Gray et al., 1959), cirrhosis. (Gray et al., 1959), hypertension (Strange et al., 1968), obesity (Argenta et al., 19181; Strange et al., 1963; Taylor et al., 1961), diabetes (Argenta et al., 1981; Strange et al., 1968; Taylor et al., 1961), impaired glucose tolerance (Green et al., 1970; Enzi et al., 1983.), renal tubular acidosis (Springer et al., 1972), thyroid abnormalities (Rhoads et al., 1986; Hugo et al., 1966), and peripheral neuropathy (Enzi et al., 1985). These associations have been inconsistent and no constant association has been established. The familial occurrence of MSL has been described (Kurzweg et al., 1951) and an autosomal dominant mode of inheritance has been postulated (Mckusick, 1978), but the majority of cases are sporadic.

The disorder is subdivided into Type I in which the fatty tumors remain circumscribed and protrude from the surrounding tissues and type II in which the tumors are diffuse, Involve extensive areas of subcutaneous tissue, and give the patient the appearance of simple obesity.

MSL can arise in any location in which fat is normally present. The majority occur in the upper half of the body, particularly the neck and trunk. Enzi (Enzi, 1984) reviewed nineteen MSL which were located symmetrically in the subcutaneous fat layers at the neck, shoulders, chest, abdomen, and groin as in our two cases; the neck and the shoulders were involved in almost all subjects. Also they occur in deeper structures such as in the mediastinum and retroperitoneum (Enzi et al., 1982). Deep accumulation of

fatty tissue is independent of involvement of the overlying subcutaneous fat.

The clinical course of MSL is characterized by progressive growth of fatty tumors which are nontender and painless, but varies from patient to patient. In some patients, the size of the masses remains relatively stationary for a long time and then increase in size rapidly within a few weeks.

Clinically, the benign lipoma is usually totally asymptomatic save for its presence as a tissue mass. Its soft, yielding quality frequently permits a correct clinical diagnosis. Deep seated benign lipomas may cause concern by producing symptomatic compression or displacement of deep organs, such as the mediastinum or the trachea (Enzi et al., 1982). It is demonstrable by radiograpy or computed tomography.

Grossly, a liopoma even if apparently circumscribed is characteristically nonencapsulated, soft, multilobular mass of typical adult adipose tissue. Microscopically, the accumulated fat is indistinguishable from mature fat except for varying degrees of interstitial fibrosis. Analysis of the fatty growth by Mueller et al (Mueller et al., 1976) revealed high levels of acid mucopolysaccharides when compared to normal fat.

Surgical removal of lipoma in the subcutaneous layer is cosmetically palliative with preservation of all structures. Deep seated mass causing pressure symptoms on contiguous structures requires surgery late in the course of the disease.

Complications of MSL are generally cosmetic, but infiltration of deep tissues with mediastinal involvement and tracheal compression have occurred (Enzi et al., 1982). One case of malignant degeneration to a myxoid liposarcoma has been reported (Tizian et al., 1983).

REFERENCES

Argenta LC, McClatchey KD, Ferrell WG: *Benigh symmetrical lipomatosis (Madelung's disease). Head Neck Surg 3:240-243, 1981.*

Brodie BC: *Lectures in pathology and surgery. Longman, Brown, Green and Longmans, London. p375, 1846*

Enzi G: *Multiple symmetric lipomatosis: an updated clinical report. Medicine 63:56-64, 1984.*

Enzi G, Angelini C, Negrin P, Armani M, Pierobon S, Fedele D: *Sensory, motor, and autonomic neuropathy in patinets with multiple symmetric lipomatosis. Medicine 64:388-392, 1985.*

Enzi G, Biondetti PR, Fiore D, Mazzoleni F: *Computed tomography of deep fat masses in multiple symmetrical lipomatosis. Radiology 144:121-124, 1982.*

Enzi G, Favaretto L, Martini S: *Metabolic abnormalities in multiple symmetric lipomatosis: Elevated lipoprotein lipase activity in adipose tissue with hyperalphlipoproteinemia. J Lipid Res. 24:566-574, 1983.*

Gray GH, Jones H: *A case of diffuse symmetric lipomatosis. Plast Reconstr Surg. 23:547-549, 1959.*

Green ML, Gloeck CJ, Fujimoto WY: *Benign symmetric lipomatosis (Launois-Bensaude adenolipomatosis) with gout and hyperlipoproteinemia. Am J Med. 48:239-246, 1970.*

Golwell-JA, Crug SR: *Effect of resection of adipose tissue on the diabetes and hyperinsulinism of benign symmetric lipomatosis. Diabetes 21:13-15, 1972.*

Hugo NE, Conway N: *Benign symmetric lipomatosis: A case report. Plast Reconstr Surg 37:69-71: 1966.*

Kurzweg FT, Spencer R: *Familial multiple lipomatosis. Am J Surg 82:762-765, 1951.*

Launois PE, Bensaude R: *De L'adeno-lipomatose symetrique. Bull Mem Soc Med Hop Paris 1:298-317, 1898.*

Madelung OW: *Ueber den Fettals. Arch Klin Chir 37:106-130, 1888.*

McKusick VA: *Mendelian inheritance in man. Catalogs of autosomal dominant, autosomal recessive, and x-linked phenotypes. The Johns-Hopkins University Press, Baltimore. 5th ed: 242-243, 1978.*

Mueller MM, Fuchs M, Schwarzmeier JD: *Zur Biochemie der benignen symmetrischen lipomatose (Adenomatouse Launois-Bensaude, Madelung'sche Krankheit). Wien Klin Wochenschr 88:94, 1976.*

Rhoads DD, Dixon AE, Taylor SA: *Multiple symmetric lipomatosis in a patient with hürthle cell carcinoma of the thyroid. Laryngoscope 96:91-95, 1986.*

Springer HA, Whitehouse JS: *Launois-Bensaude adenolipomatosis. Plast Reconstr Surg 50:291-294, 1972.*

Strange DA, Fessel WJ: *Benign symmetric lipomatosis. JAMA 204:129-130, 1986.*

Taylor LM, Beuhrs OH, Fontana RS: *Benign symmetric lipomatosis. Proc Staff Meet Mayo Clin 36:96-1000, 1961.*

Tizian C, Berger A, Vykoupil KF: *Malignant degeneration in Medelung's disease benign lipomatosis of the neck : case report. Br J Plast Surg 36:187-189, 1983.*