

Benign Symmetric Lipomatosis: A Patient Report

Yoshiyuki Tanji, Noriko Ikeda, Masami Shibata, Isamu Kodani, Kazuhito Tobashi, Kazunori Kidani, Fumihiro Otake, Kazuhiko Tanio and Kazuo Ryoke

Division of Oral and Maxillofacial Biopathological Surgery, Department of Medicine of Sensory and Motor Organs, School of Medicine, Tottori University Faculty of Medicine, Yonago, 683-8504 Japan

Benign symmetric lipomatosis (BSL) is a disease characterized by symmetrical and diffuse adipose tissue deposits on the neck, shoulder, chest, abdomen and groin. It is comparatively common in the Mediterranean basin, but rare in Japan. We report on a 53-year-old male patient with BSL along with a review of the literature. The lesions were distributed bilaterally in the submandibular, neck and upper back region. This patient had consumed an excessive amount of alcohol with elevated liver enzymes including aspartate aminotransferase (133 IU/L), alanine aminotransferase (159 IU/L) and gamma-glutamyl transpeptidase (355 IU/L). The liver function was normalized after a 1-year restriction of alcohol consumption through life management. Tumors in the posterior and anterior cervical regions were resected. The patient is still in a good condition at 3 years and 5 months after the operation.

Key words: alcoholic hepatopathy disease; benign symmetric lipomatosis

Benign symmetric lipomatosis (BSL) was first reported in 1846 by Brodie et al. (1846) as a comparatively rare disease characterized by diffuse symmetric deposits of fat. Etiology of BSL has not been clear. The neck is affected most often, but the shoulder, chest, abdomen and inguinal regions can also be involved (Enzi, 1984). Although the BSL is comparatively common in the Mediterranean basin, it is rare in Japan. Here, we report a patient of BSL in the cervical region with alcoholic liver disease.

Patient Report

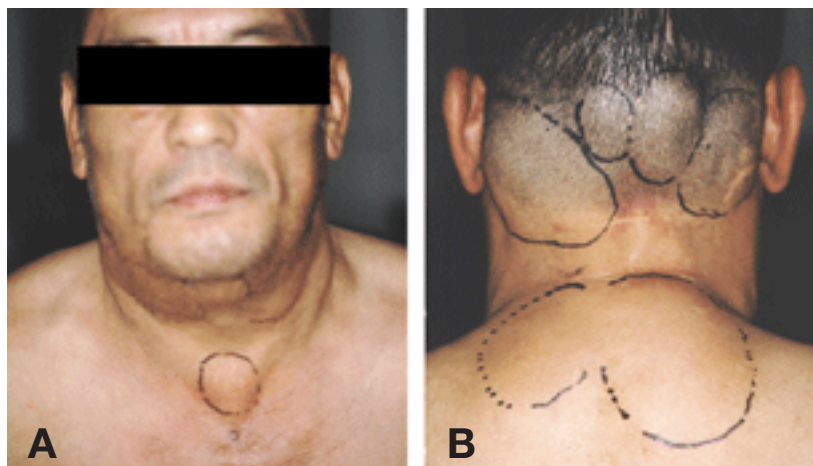
In 1993, a 46-year-old male visited a surgical clinic at a general hospital, complaining of swell-

ings in the right submandibular and right lumbar regions. Only tumors in the right lumbar region (2.5 cm and 1 cm in diameter, respectively) were excised, and diagnosed as lipoma histopathologically, while follow-up was performed on the submandibular swelling. He visited an internist in early 2000 when he was 53 years old, because he started to notice a swelling in the right anterior region of the neck. He was referred to our hospital. He began to drink at the age of 18. His drinking increased gradually from the age of approximately 32, eventually drinking 2L of Japanese sake (ca. 15% alcohol) a day. Though the amount of drinking was gradually reduced thereafter, he still drank about 1L of beer everyday. His sake index was 183 [sake index: the number of units of daily sake consumption (1 unit = 27 g alcohol) multi-

Abbreviations: ALT, alanine aminotransferase; AST, aspartate aminotransferase; BSL, benign symmetric lipomatosis; CT, computed tomography; γ -GTP, gamma-glutamyl transpeptidase; HE, hematoxylin and eosin; LDH, lactate dehydrogenase; TTT, thymol turbidity test; ZTT, zinc sulfate turbidity test

Fig. 1. Appearance of the patient at the 1st visit.

- A:** The front side shows diffuse masses in the cervical and sub-mandibular regions.
- B:** The back side has masses in the neck and back regions, looking like the buffalo hump.



plied by number of years of drinking] (Miyahara et al., 1981).

Physical examination revealed a number of masses: one (60 × 100 mm) in the right sub-mandibular region, one (45 × 45 mm) in the left submandibular region, one (50 × 50 mm) in the submental region, one (30 × 45 mm) in the anterior neck, one (30 × 50 mm) in the left lateral neck, four (57 × 90 mm, 25 × 25 mm, 40 × 40 mm and 50 × 60 mm) in the posterior neck and two (60 × 50 mm and 70 × 100 mm) in the upper back regions (Figs. 1A and B). These masses were moderately hard but without tenderness, and their margins were indiscernible. When he visited our hospital, his height was 161.5 cm with a body weight of 64 kg. His cervical region appeared like the buffalo hump. His platelet level was 75,000/μL. In liver function tests, the levels were: aspartate aminotransferase (AST), 133 IU/L; alanine aminotransferase (ALT), 159 IU/L; gamma-glutamyl transpeptidase (γ-GTP), 355 IU/L; thymol turbidity test (TTT), 19.6 KU; zinc sulfate turbidity test (ZTT), 11.2 KU; lactate dehydrogenase (LDH), 236 IU/L and uric acid, 8.4 mg/dL (Table 1). CT (computed tomography) showed swelling in the bilateral cervical regions, the posterior region of the neck, and the supra-clavicular region. His CT value for the swellings was -100 Hounsfield units, the same as that for fatty tissues. The inner tissues of the masses were comparatively uniform, and no findings suggested

Table 1. Clinical laboratory findings

Examination	Level		
Blood count			
White blood cell	(/μL)	5.5 × 10 ³	
Red blood cell	(/μL)	4.18 × 10 ⁶	
Hb	(g/dL)	15.5	H
Hematocrit	(%)	45.6	H
Mean corpuscular volume	(fL)	109.1	H
Mean corpuscular Hb	(pg)	37.2	H
Mean corpuscular Hb concentration	(%)	34.1	
Platelet	(/μL)	75 × 10 ³	L
Biochemical test			
Na	(mEq/L)	141	
K	(mEq/L)	4.4	
Cl	(mEq/L)	105	
Blood urea nitrogen	(mEq/L)	10	
Cr	(mg/dL)	0.75	
Ca	(mg/dL)	8.8	
Aspartate aminotransferase	(IU/L)	133	H
Alanine aminotransferase	(IU/L)	159	H
Gamma-glutamyl transpeptidase	(IU/L)	355	H
Lactate dehydrogenase	(IU/L)	236	H
Creatine kinase	(IU/L)	84	
Thymol turbidity test	(KU)	19.6	H
Zinc sulphate turbidity test	(KU)	11.2	
Uric acid	(mg/dL)	8.4	H
Urine test			
Urinometry		1.022	
Urine sugar		Negative	
Urine protein		Negative	

H, higher than normal levels; L, lower than normal levels

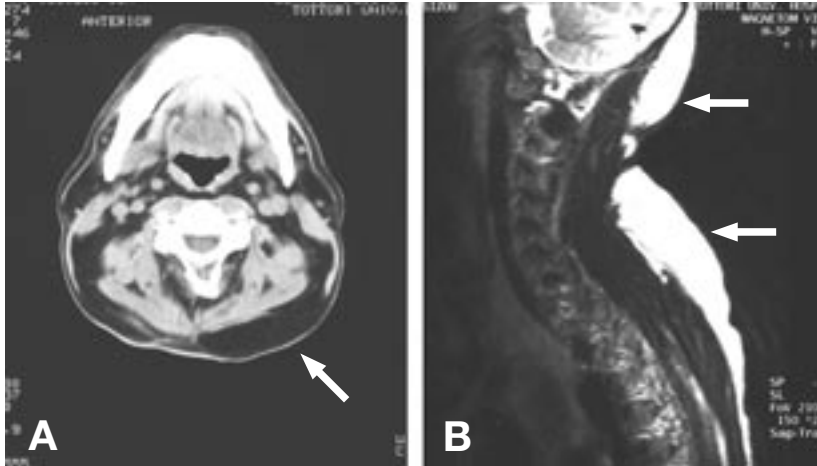


Fig. 2. Radiographic images of masses

A: A CT image visualizes hyperplasia of the fatty tissue in the posterior region of the neck (arrow).

B: A T2-emphasized magnetic resonance imaging shows high signals at the posterior subcutaneous region of the neck and the upper back region (arrows).

malignancy. The borders with normal tissue were not clear (Fig. 2A). Ultrasonic inspection showed thick connective tissues in the submandibular and neck regions. A low net-shaped signal area was visible. T2-emphasized magnetic resonance imaging presented high signals in the neck, submandibular regions and posterior regions of the neck, bilaterally. The boundary of lesions was not clear in each region (Fig. 2B). On July 19, 2000, the right submandibular lesion was subjected to biopsy. Histopathologically, it was diagnosed as lipomatosis due to hyperplasia of mature, non-atypical fat cells (Figs. 3A and B). A diagnosis of BSL was determined from these results.

Moderate levels of hepatopathy were found in the 1st routine blood test (Table 1), and the patient was referred to the Second Clinic of Internal Medicine at our hospital for further evaluation. Being diagnosed with alcoholic liver disease at the clinic, alcohol restriction was begun through life habit guidance for improvement of hepatic function.

On July 19, 2001 when he laid down to rest, he had shortness of breath and bilateral swelling in the upper arms. Vascular phlebography revealed no arctation of the subclavicular vein on either side, nor displacement toward the outside of the brachiocephalic vein. After improvement

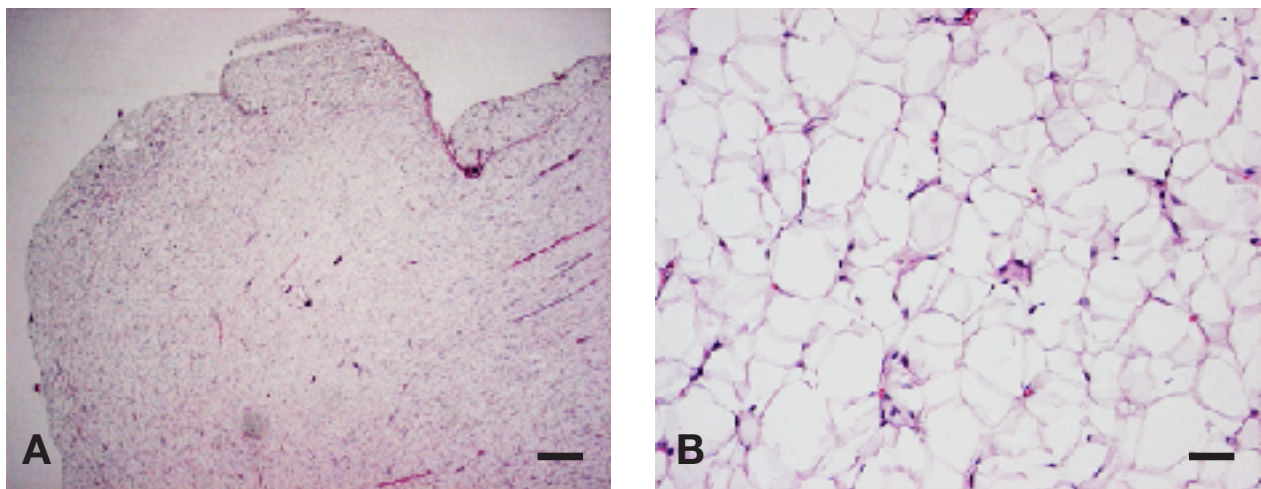


Fig. 3. Histopathological figures obtained at biopsy. Hyperplasia of matured fat cells with no atypical formation is recognized diffusely.

A: HE stain. Bar = 1 mm.

B: HE stain. Bar = 50 μ m.

of hepatic function (AST: 79 IU/L, ALT: 80 IU/L, γ -GTP: 161 IU/L, TTT: 4.1 KU and ZTT: 5.7 KU), breathlessness and swelling of the upper arms, tumors of the posterior and anterior regions of the neck were surgically removed on August 23, 2001. A mass in the right posterior neck was not resected for the change of the position. Gross examination of a fatty mass resected from the left posterior neck region revealed diffuse infiltration into the surrounding tissue with an unclear border (Fig. 4). The excised tumor was histopathologically diagnosed as lipomatosis on the basis of lobulated hyperplasia of mature adipocytes. Since the operation, the patient has refrained from drinking. Up until 3 years and 5 months post-operation, no additional masses have appeared, and no enlargement of the non-resected masses have been detected.

Discussion

Multiple lipomatosis was first reported by Brodie (1846). Madelung described 33 patients with "Madelung's disease" (1888). Since then, this disease had been called Madelung's diffuse symmetric lipomatosis, but most recently, cases have been reported as BSL. Approximately 400 patients have been described in the literature so far (Harsch et al., 2004). BSL predominantly affects white males (male:female ratio, 15:1). The fat deposits first appear after the age of 20 and becomes most common between the ages of 30 and 60 (Ruzicka et al., 1987). The highest prevalence is in the Mediterranean basin (reported incidence in Italy, 1 in 25,000 males) (Parmar et al., 1996). In Japan, after the first report by Okuma (1941), BSL appeared from time to time, showing a tendency towards increase. Although no classification for lipomatosis is established at present, Carlsen's classification which clinically organizes systemic benign lipomatosis into 3 types is generally accepted (Carlsen et al., 1978). Type I is diffuse and congenital lipomatosis. It develops mainly in the body trunk with no inheritable characteristics,

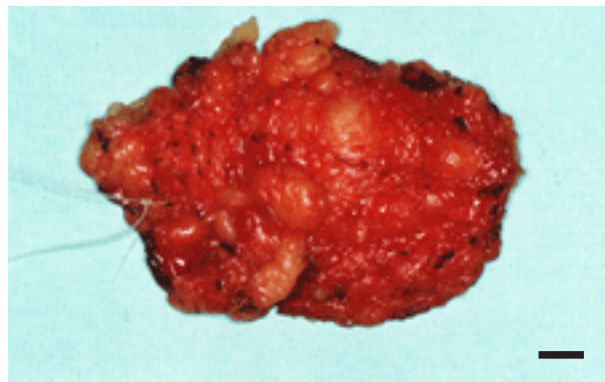


Fig. 4. A tumor specimen excised from the left posterior region of the neck. Bar = 1 cm.

and its boundary with the surrounding tissue is vague; it also infiltrates the muscle and the retroperitoneal region, diffusely. This often leads to recurrence after excision. Type II is symmetrical, diffuse lipomatosis appearing in adult life, primarily localized in the neck region but occasionally accompanied by symmetrical lipomatosis in other parts of the body. This condition is hereditary. Type III is multiple lipomatosis consisting of usually numerous, small, well-defined, subcutaneous lipomas, mainly localized to the limbs, especially the forearms, but occasionally also to the trunk. The tumors which are surrounded by fibrous capsules, are easy to enucleate and never recur in the same localization although new elements will often appear elsewhere. This condition is also considered to be hereditary. According to Carlsen's classification, our patient may be Type II, the symmetrical, diffuse type due to symmetrical bilateral lesions in the neck and upper back and presenting a buffalo hump, though its hereditary character was not recognized. There are some reports that these lesions have been seen in the chest, abdomen and other places. A favorite site of lipomatosis Type II is in the cervix, also seen in the present patient, though the ends of the limbs and the lower half of the body are not usually invaded. There are other reports that lipomatosis occurs in the tongue though oral area lesions are rare (Vargas-Dies et al., 2000;

Sato et al., 2004). There may be various factors that cause lipomatosis, which have not yet been clarified. Alcoholic liver disease (Berkovic et al., 1991; Nobukiyo et al., 1997), diabetes (Springer et al., 1972), hyperlipemia (Greene et al., 1970), gout (Taylor et al., 1961), hypothyroidism (Hugo et al., 1966) and hyperuricemia (Kawamoto et al., 1985) are often accompanying complications. Etiologically, liver dysfunction can relate to a long history of drinking and this, coupled with hyperuricemia, could be related to our patient. Miyahara et al. (1981) reported that a sake index of over 50 tends to cause hepatopathy due to alcohol consumption. Our patient's sake index was 183, and his liver seemed to be damaged.

One report explained that the fatty deposition of this disease is not systemic but localized, and that a decrease of adipose tissue in the lesion by means of lessening excessive weight is apparently low compared to normal regions (Greene et al., 1970), so it is suggested that this disease has no direct relation to obesity. There is another report that these lesions may grow gradually for several years after onset and then stop at some point, becoming a fixed size in many patients. No recurrence has been observed in our patient 3 years and 5 months after operation, and progress can be noted through the improvement of hepatic function. The histopathological findings showed hyperplastic adipose tissue consisting of normal matured fat cells, which are peculiar for this disease. Many patients without capsules have been reported, thus the capsule is not necessarily specific to this lesion. It is also difficult to distinguish with usual adipose tissue (Enzi et al., 1983). In our patient, the lesion was infiltrative lipomatosis without capsules.

In this disease, surgery is the 1st choice of treatment. An operation can be especially helpful for patients with considerable cosmetic deformity or presentation of a compressed airway. We decreased the volume of adipose tissue as much as possible for our patient, and he was satisfied with his appearance. Another choice of treatment has been reported such as liposuction (Samdal et al.,

1991), dietetic treatment, (Ruzicka et al., 1987) including forbidding alcoholic intake, as other modalities of treatment. In this patient, refraining from alcohol has helped keep a healthy hepatic condition. Also, during the 3 year and 5 month follow up, no additional masses were detected. Thus, the etiology of BSL might be associated with alcoholic hepatopathy.

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Corresponding author: Yoshiyuki Tanji