

Multiple symmetrical lipomatosis: case report and literature review

K J SIA¹, I P TANG¹, T Y TAN²

¹Department of Otorhinolaryngology, Faculty of Medicine, University of Malaya, and ²Otorhinolaryngology Department, Sarawak General Hospital, Malaysia

Abstract

Objective: To discuss the pathophysiology and various treatment methods of multiple symmetrical lipomatosis.

Case report: We report a case of multiple symmetrical lipomatosis in a middle-aged man. He presented to us with an extensive, disfiguring neck mass. Clinical examination and computed tomography suggested a lipomatous mass without compression of vital neck structures. The proximity of the lipomatous mass to the carotid sheaths attracted our interest. We performed surgical excision in this case, because of the deep infiltrative behaviour of the lipoma. The benefits and drawbacks of liposuction and surgical excision of extensive neck lipomatosis are discussed.

Conclusion: Surgical excision and liposuction are complementary treatments in the management of multiple symmetrical lipomatosis. Patients should be aware of the limitations of both surgical options, and the risk of lipoma recurrence, before surgery. The decision on the mode of surgery relies upon the disease extent, the patient's expectations and the surgeon's experience.

Key words: Multiple Symmetrical Lipomatosis; Madelung Disease; Lipoma; Surgical Procedure, Operative

Introduction

Multiple symmetrical lipomatosis, also known as Madelung disease, is a rare disease caused by abnormal lipid metabolism. Most reports describe its occurrence in middle-age men with a history of alcohol abuse.

The aetiology of multiple symmetrical lipomatosis is unknown. However, mitochondrial DNA mutations have been found on muscle biopsy, suggesting one possible pathophysiological mechanism.¹

Histologically, the lipomatous mass is seen to consist of benign, hyperplastic adipocytes.

The disease is characterised by insidious, progressive growth of the lipomatous mass, which has a high propensity for recurrence after removal. Advanced cases of neck lipomatosis with mediastinal extension can develop airway and vascular compression.

We report a patient with multiple symmetrical lipomatosis and deep neck infiltration. The proposed pathogenesis and treatment options are discussed.

Case report

A 45-year-old man was referred to us with a massive, diffuse neck mass. The mass had been noticed for the previous 10 years, and had gradually increased in size. There was no symptom of upper aerodigestive compression. The patient's head and neck movements were unrestricted. Similar subcutaneous masses were found on both arms and the upper back. The patient was a chronic smoker and social drinker. He was otherwise well, with no neurological symptoms.

On examination, the mass was seen to involve the entire circumference of the neck. It extended from the lower border of the mandible to the supraclavicular region (Figure 1).

There were no significant findings on nasal or laryngeal endoscopy.

Fine needle aspiration cytology showed benign adipocytes, consistent with a diagnosis of lipoma.

Computed tomography of the neck showed diffuse neck lipomatosis distributed between the neck muscles (i.e. the semi-spinalis, splenius capitis and trapezius) and abutting both carotid sheaths (Figure 2). There was no mediastinal extension or significant cervical or mediastinal lymphadenopathy.

The patient was scheduled for surgical excision of the lipomatous mass. Intra-operatively, extensive, unencapsulated fat tissue was found. This involved the parapharyngeal space and was in contact with the carotid sheaths bilaterally. The surgical dissection was difficult due to the ill-defined dissecting plane. Debulking surgery was performed, with removal of 2 kg of fat tissue.

Post-operatively, there was seroma formation, which resolved after repeated needle aspiration.

Histopathological examination of the surgical specimen showed benign lipoma with reactive lymph nodes.

The neck wound healed well, with a good aesthetic outcome seen at follow up (Figure 3).

Discussion

Multiple symmetrical lipomatosis is characterised by extensive fat tissue deposition, especially on the neck, trunk and upper extremities. Its most common presentation is an



FIG. 1

Clinical photograph showing symmetrical fat distribution mainly localised in the neck region.

unsightly neck mass. Advanced cases may have mediastinal extension of fat tissue which leads to aerodigestive compression symptoms.

Although the aetiology of this rare disease remains obscure, mitochondrial dysfunction is believed to play a role in the pathogenesis. Several studies had reported the identification of a deletion mutation of mitochondrial DNA on muscle biopsy in some patients.^{1,2} The identified mutations cause a decrease in oxidative phosphorylation



FIG. 2

Axial computed tomography scan of the neck showing circumferential lipomatosis. The fatty tissue has infiltrated into the muscle plane, abutting the carotid sheaths bilaterally.



FIG. 3

Clinical photograph showing a good cosmetic outcome, one week after surgical excision of the lipoma.

which would result in lipoma formation. Klopstok T *et al.* have also identified a mitochondrial DNA mutation within a sural nerve biopsy.¹ Furthermore, these authors also performed electrophysiological tests which demonstrated axonal neuropathy in 60 per cent of subjects with multiple symmetrical lipomatosis.

Head and neck malignancies have been reported in patients with multiple symmetrical lipomatosis; however, this association remains uncertain.³⁻⁵ It is essential to perform a complete clinical assessment of the head and neck region in patients with a history of alcohol abuse, as the latter is an aetiopathogenetic factor for both head and neck malignancy and multiple symmetrical lipomatosis. Aerodigestive compression symptoms should not be attributed to lipoma compression until regional malignancy has been excluded by endoscopic and imaging investigations; malignant tumour and cervical lymphadenopathy may lurk unnoticed underneath a massive lipoma.

Extremely rare malignant transformation of multiple symmetrical lipomatosis has also been reported.⁶ Tizian *et al.* have described the appearance of intramyxoid sarcoma in a case of benign lipomatosis after six years' follow up, while Durand *et al.* reported a case of liposarcoma transformation.^{7,8}

The treatment for multiple symmetrical lipomatosis ranges from surgery to various modes of medical therapy. Surgical excision and liposuction are still the most effective surgical options.^{9,10} Liposuction has gained popularity recently due to its minimal scar. Liposuction is considered less invasive and technically easier, compared with surgical excision. It is also more suitable for patients who represent a higher surgical and/or anaesthetic risk.¹¹ Based on the high propensity of lipoma for recurrence, some authors suggest that the goal of treatment should be a palliative result with restoration of function. It is unnecessary to subject patients to the risks of radical surgery for this benign condition. Surgical excision

should be limited to cases with airway compression or cosmetic unacceptability.¹² Liposuction is safer and less aggressive, and suffices for debulking purposes.

On the other hand, liposuction in the neck region performed by an inexperienced surgeon always carries the risk of injury to vital neck structures, which could be life-threatening. Faga *et al.* have described ultrasound-assisted liposuction in a case with neck movement restriction.¹¹ This technique allows gentler and more precise suction without major damage to subcutaneous tissue and vascular structures. Nevertheless, in the case in question the procedure was performed for the purpose of functional relief, and the cosmetic outcome was unsatisfactory.¹¹ The limitations of liposuction include inadequate aspiration of lipoma, especially in cases with a dense, fibrous constitution. Furthermore, liposuction is often restricted in the submental region, and in areas where previous surgery has been performed with subsequent scarring.¹² Thus, patients should be made aware of the possibility of residual lipoma due to inadequate liposuction. Despite the above risks, at present there has been no reported case of major complications from liposuction.

The main disadvantage of surgical excision is the long, unsightly scar.

In our patient's case, we opted for surgical excision because we believed this was safer as the lipoma abutted both carotid sheaths. Direct visualisation during surgery may reduce the risk of injury. We anticipated a difficult dissection as the lipomatous mass was unencapsulated and had deep tissue infiltration.¹³ At operation, the pathological hyperplastic lipoma was hard to distinguish from normal subcutaneous fat and the lymphofatty tissue along the carotid sheaths. After the lipoma debulking surgery, redundant skin was apparent. Excess skin was trimmed to restore the normal neck contour, enabling optimal cosmesis.

- **The pathophysiology of multiple symmetrical lipomatosis (Madelung disease) is unclear**
- **Cosmetic deformity is the commonest problem**
- **Airway and vascular compression are rare**
- **Pre-operative computed tomography is crucial to assess extent and proximity to vital structures**
- **Open excision is best for massive, deeply infiltrating lipomatosis**

Lifestyle modification, with alcohol abstinence and control of blood sugar and lipids, may curb fat growth but will not shrink a pre-existing lipoma.¹⁴ Toshio *et al.* have reported the use of phosphatidylcholine as mesotherapy (via intraleisional injection), with satisfactory results.¹⁰ Other reported methods have included the use of an oral $\beta 2$ agonist (i.e. salbutamol) and a peroxisome proliferator-activated α receptor agonist (fibrate). However, at present the consistent efficacy of these treatments has not been demonstrated.¹⁵

Conclusion

Complete clinical assessment of patients with multiple symmetrical lipomatosis is imperative in order to exclude any

associated head and neck malignancy. Surgical excision and liposuction are complementary treatment modalities in the management of this condition. Patients should be aware of the risks and benefits of these surgical options, and of the risk of lipoma recurrence, before surgery. The decision on the mode of surgery depends upon the disease extent, the patient's expectations and the surgeon's experience.

References

- 1 Klopstock T, Naumann M, Seibel P, Shalke B, Reiners K, Reichmann H. Mitochondrial DNA mutations in multiple symmetric lipomatosis. *Mol Cell Biochem* 1997;**174**:271–5
- 2 Silvestri G, Ciafaloni E, Santorelli FM, Shanske S, Servidei S, Graf WD *et al.* Clinical features associated with the A-G transition at nucleotide 8344 of mtDNA ('MERRF mutation'). *Neurology* 1993;**43**:1200–6
- 3 Ujpál M, Németh ZS, Reichwein A, Szabó GY. Long-term results following surgical treatment of benign symmetric lipomatosis (BSL). *Int J Oral Maxillofac Surg* 2001;**30**:479–83
- 4 Chan ESY, Ahuja AT, King AD, Lau WY. Head and neck cancers associated with Madelung's disease. *Ann Surg Oncol* 1999;**6**:395–7
- 5 Ruzicka T, Vieluf D, Landthaler M, Braun-Falco O. Benign symmetric lipomatosis. *J Am Acad Dermatol* 1987;**17**:663–74
- 6 Guastella C, Borsi C, Gibelli S, Berta LG. Madelung's lipomatosis associated with head and neck malignant neoplasia: a study of 2 cases. *Otolaryngol Head Neck Surg* 2002;**126**:191–2
- 7 Tizian C, Berger A, Vykoupil K. Malignant degeneration in Madelung's disease. *Br J Plast Surg* 1983;**36**:187–9
- 8 Durand JP, Thomine JM, Tayot J, Foucault J, Deshayes P. Liposarcoma in a Launois-Bensaude disease. *French* 1973;**40**:287–91
- 9 Alameda YA, Torres L, Perez-Mitchell C, Riera A. Madelung disease: a clinical diagnosis. *Otol Head Neck Surg* 2009;**141**:418–19
- 10 Hasegawa T, Matsukura T, Ikeda S. Mesotherapy for benign symmetric lipomatosis. *Aesth Plast Surg* 2010;**34**:153–6
- 11 Faga A, Valdatta LA, Thione A, Buoro M. Ultrasound assisted liposuction for the palliative treatment of Madelung's disease: a case report. *Aesthetic Plast Surg* 2001;**25**:181–3
- 12 Verhelle NA, Nizet JL, Van den Hof B, Guelinckx P, Heymans O. Liposuction in benign symmetric lipomatosis: sense or senseless. *Aesthetic Plast Surg* 2003;**27**:319–21
- 13 Sreekrishna KD, James CG, Rosemary SS. Lipomatosis of the neck: case report and literature review. *Int J Pediatr Otorhinolaryngol Extra* 2010;**5**:39–41
- 14 González-García R, Rodríguez-Campo FJ, Sastre-Pérez J, Muñoz-Guerra MF. Benign symmetric lipomatosis (Madelung disease): case report and current management. *Aesthetic Plast Surg* 2004;**28**:108–12
- 15 Zeitler H, Ulrich-Merzenich G, Richter DF, Vetter H, Walger P. Multiple benign symmetric lipomatosis – a differential diagnosis of obesity. Is there a rationale for fibrate treatment? *Obes Surg* 2008;**18**:1354–6

Address for correspondence:

Dr Kian Joo Sia,
14A, Lorong 4A, Jalan Pipit,
96000 Sibul, Sarawak, Malaysia

Fax: 6 082419495

E-mail: kj_sia@yahoo.com

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