

Congenital Infiltrating Lipomatosis of the Face: Case Report and Literature Review

Chien-Ming Chen, MD; Lun-Jou Lo, MD; Ho-Fai Wong¹, MD

Congenital infiltrating lipomatosis of the face comprises a subgroup of lipomatous tumors. While rare, it remains a definite clinical entity. Its etiology is unknown. The tumor is congenital in origin and occurs in infancy or early childhood. It is poorly enveloped and characterized by diffuse infiltration of mature adipose tissue over normal muscle fibers, rapid growth, associated osseous hyperplasia, and a high recurrence rate after surgical intervention. Due to its diffuse infiltration and involvement of important facial structures, complete surgical excision is often impossible. A total of 14 cases of congenital infiltrating lipomatosis of the face was found in a literature review. Herein we present an additional case who had both diffuse infiltrating lipomatosis and a well-encapsulated lipoma on her left cheek. (*Chang Gung Med J* 2002;25:194-200)

Key words: infiltrating lipomatosis, facial tumor, infant.

Congenital infiltrating lipomatosis of the face is rare and difficult to treat. It is characterized by diffuse infiltration of mature adipose cells over facial soft tissue, rapid growth, osseous deformity, and a high risk of recurrence after surgery. A case is presented together with a literature review.

Lipomas are the most-common benign tumors of mesenchymal origin. According to clinical and histological presentations, benign lipomatous tumors are classified into 5 categories: 1) simple or multiple well-encapsulated lipoma; 2) lipoma variants, including angiolipoma and pleomorphic lipoma; 3) heterotopic lipoma, which is intimately associated with a specific tissue, like intramuscular lipomas or fibrolipoma; 4) infiltrating or diffuse lipomatosis; and 5) the benign tumor of brown fat, i.e., hibernoma.⁽¹⁾ Infiltrating or diffuse lipomatosis is characterized by a diffuse overgrowth of mature adipose tissue that usually involves subcutaneous tissue and muscle. Lesions may be associated with bony hyper-

trophy or distortion. Infiltrating lipomatosis is usually located in the shoulder, thigh, and limbs.^(2,3) In most reported series of infiltrating lipomatosis, the lesions were initially observed after the first 3 decades of life, with a peak at ages between 40 and 60 years.⁽¹⁻⁴⁾

Congenital infiltrating lipomatosis is a distinct clinicopathological entity. It seems to represent a distinguishable type of lipomatosis in that it is usually found at birth or early after birth, there is encasement of the facial nerve resulting in poor aesthetic results after operation, and it has a high recurrence rate. The incidence of congenital infiltrating lipomatosis on the face is rare, and only a few cases have been reported. Herein we present another case and review the literature.

CASE REPORT

This female patient was found to have a left cheek mass since birth. There were no relevant

From the Department of Plastic and Reconstructive Surgery, ¹Department of Radiology, Chang Gung Memorial Hospital, Taipei.

Received: May 4, 2001; Accepted: Aug. 3, 2001

Address for reprints: Dr. Lun-Jou Lo, Department of Plastic and Reconstructive Surgery, Chang Gung Memorial Hospital, 5, Fushing Street, Kweishan 333, Taoyuan, Taiwan, R.O.C. Tel.: 886-3-3281200 ext. 2855; Fax: 886-3-3285818; E-mail: lunjoulo@ms1.hinet.net;

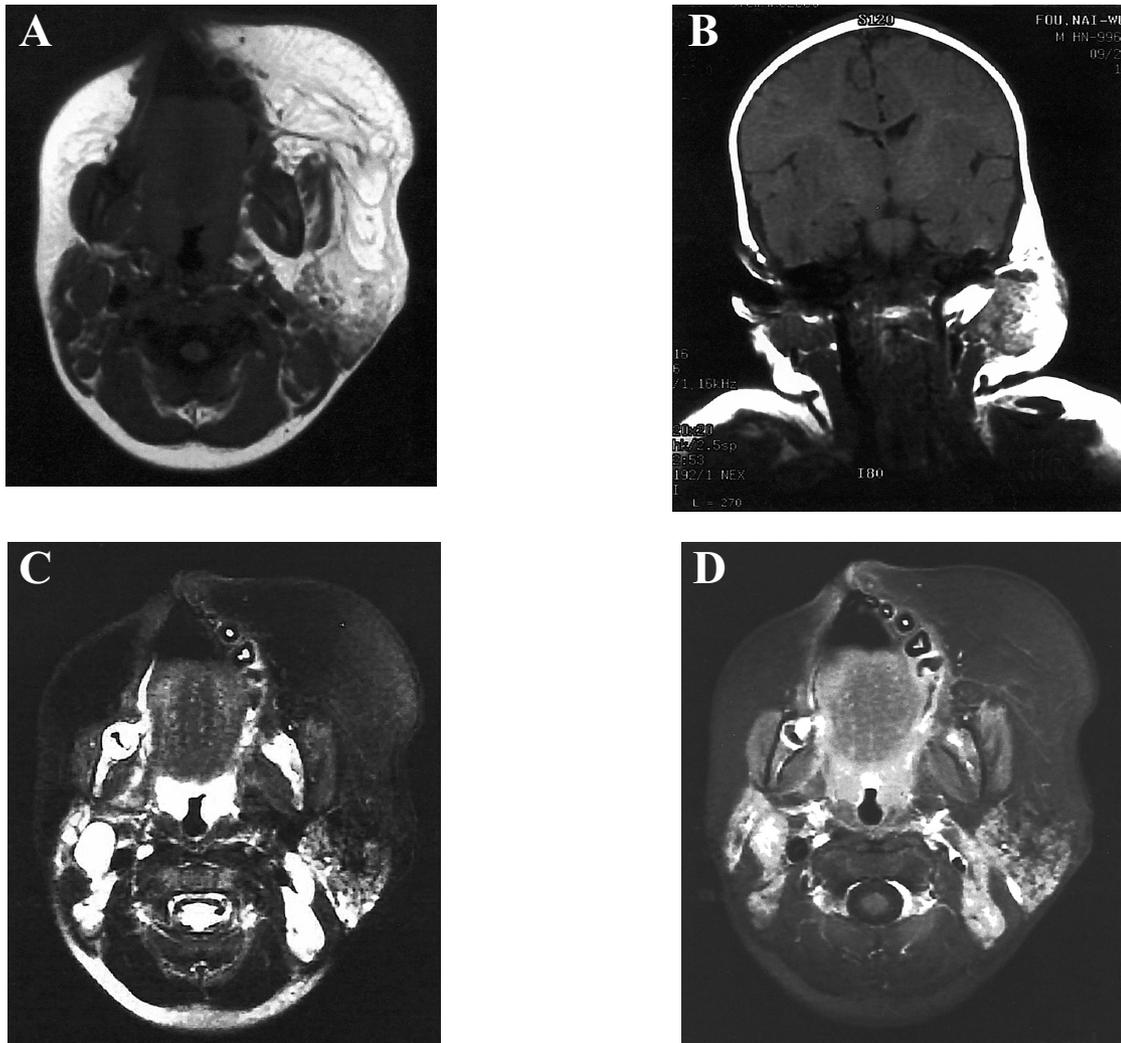


Fig. 1 Magnetic resonance imaging study. (A) T1WI (TR/TE 540/14) axial and (B) coronal sections reveal a poorly demarcated mass with hyperintensity in the left cheek, showing diffuse infiltration to the buccinator space, buccal space, masticator space, parapharyngeal space, and infratemporal fossa. The lesion also involved the left parotid gland and extended to the deep parotid lobe compromising the facial nerve. (C) The lesion was found to have hypointensity on T2WI (TR/TE 4000/90) with the fat-suppression technique. (D) No abnormal enhancement of the lesion after Gd-DTPA.

episodes during the pregnancy, such as infectious disease, drug use, exposure to teratogenic agents, or difficult labor. The cheek tumor grew in her early infancy with distortion of the facial structures such as the eye, nose, and lip on the lesion side. The overlying skin was normal. The tumor was elastic, non-tender, and with ill-defined borders. Differential diagnosis between lymphangioma and lipomatous tumor was difficult clinically. She received an inci-

sional biopsy at 2 months of age, which revealed lipomatous tissues. It was decided to observe the mass. It continued to grow at a slower rate. At 16 months of age, magnetic resonance imaging (MRI) with and without contrast was performed. The results showed diffuse infiltrating tissue of fatty component occupying the left cheek without a clear boundary (Fig. 1). The fatty infiltration was also found to involve the left buccinator space, buccal

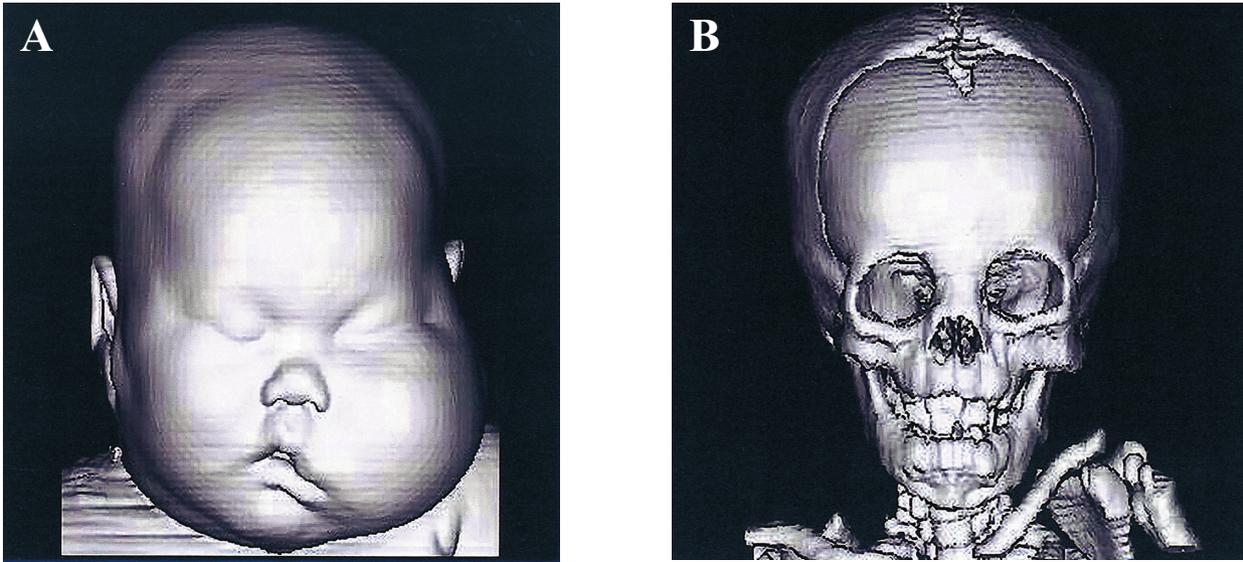


Fig. 2 Three-dimensional computed tomography study. (A) Soft-tissue imaging showing a large mass over the left cheek distorting the ipsilateral eye, nose, and mouth. (B) The soft tissue was removed to display the osseous structures. There was hypertrophy of the left zygoma and maxilla.



Fig. 3 Intraoperative view. The cheek flap was retracted with a double hook, showing diffuse infiltration of fat tissue over the left cheek structures. A piece of well-encapsulated lipomatous tissue was dissected and is ready to be excised.

space, masticator space, parapharyngeal space, infratemporal fossa, and left parotid gland. The parotid portion of the facial nerves was encased. Three-dimensional computed tomography (CT) revealed mild hypertrophy and asymmetry of the left

zygoma and maxilla (Fig. 2).

Because of concern for facial asymmetry and osseous distortion, debulking surgery was performed at 18 months of age. Through a preauricular-submandibular incision, sub-fascia dissection was carefully performed. Lipomatous tissue was found to be infiltrating between the muscles, nerves, and parotid gland. It was difficult to separate the lipomatous tissue from the adjacent structures. One piece of well-encapsulated lipomatous tissue, measuring 5.5 ; 4 ; 1.5 cm, was found anterior to the parotid gland with an extension deep into the buccal area (Fig. 3). This piece of lipomatous tissue was completely removed and used for pathological examination. The facial nerve was encased by infiltrating lipomatous tissue. It was identified with the help of an electrostimulator and preserved. As much of the infiltrating part of the tumor as possible was excised, but complete removal was impossible. Careful hemostasis was achieved, and the wound was closed. Microscopic examination of the specimens showed mature adipose tissue without polymorphic atypical cells. The postoperative course was uneventful. There was no facial nerve injury. She was regularly followed up with a residual tumor on her cheek.

Table 1. Reported Cases of Congenital Infiltrating Lipomatosis of the Face

No.	Gender	Age at report*	Location	Size	Outcome †	Reference
1	M	5 y	left cheek	3 ; 3 cm	recurrence	Scherl, et al. ⁽²⁾
2	M	6 y	right cheek	-	no recurrence	Bataineh, et al. ⁽³⁾
3	F	15 y	left cheek	-	-	Bataineh, et al. ⁽³⁾
4	F	2 y 6 mo	right parotid area	-	-	Johansen, et al. ⁽⁶⁾
5	-	2 mo	right parotid area	14 ; 16 cm	loss of follow-up	Adams, et al. ⁽⁷⁾
6	M	2 y	right parotid region	14 ; 5 cm	recurrence	Slavin, et al. ⁽⁸⁾
7	F	9 y	right temporal area	7 ; 2 cm	recurrence	Slavin, et al. ⁽⁸⁾
8	F	2 y	right buccal area	-	residual tumor	Slavin, et al. ⁽⁸⁾
9	F	9 y	left cheek	7 ; 5 cm	recurrence	deRosa, et al. ⁽⁹⁾
10	F	23 y	right submental area	20 ; 15 cm	recurrence	deRosa, et al. ⁽⁹⁾
11	M	53 y	left cheek	-	recurrence	deRosa, et al. ⁽⁹⁾
12	M	5 mo	left cheek	-	death ‡	Donati, et al. ⁽¹²⁾
13	M	4 mo	left parotid region	5 cm in the larger diameter	no recurrence	Esposito, et al. ⁽¹⁵⁾
14	F	6 y	right cheek	-	residual tumor	Cenk Gorken, et al. ⁽¹⁶⁾
15	F	1 y 6 mo	left cheek	7 ; 5 cm	residual tumor	this report

Abbreviations: M: male; F: female; y: year; mo: month.

† : Recurrence means regrowth of the residual tumor. Residual tumor means incomplete resection mentioned in this article.

‡ : Death at the age of 20 months due to pulmonary infection.

DISCUSSION

Lipomas are the most-common benign mesenchymal tumor in humans. Typically, these tumors are well encapsulated, easy to remove, and rarely recurrent after removal. These tumors are further categorized into several subgroups.⁽¹⁾ They can occur at any age but are more common after the fourth decade of life.⁽²⁻⁴⁾ Infiltrating or diffuse lipomatosis is characterized by diffuse infiltration of mature adipose tissue over normal muscle fibers, a rapid growth rate, associated osseous hyperplasia, and a high recurrence rate after surgical intervention. The lipomatosis lesions found in childhood include congenital infiltrating lipomatosis of the face, diffuse lipomatosis, nevus lipomatosis cutaneous superficialis, Michelin tire baby syndrome, and macrocephalia (Bannayan-Zonana syndrome).⁽⁵⁾

Johansen reported a case of congenital lipomatosis of the parotid gland in 1970.⁽⁶⁾ The tumor was found to have infiltrated the skin and glandular tissue without invasion of the facial nerve. Neither muscular infiltration nor bony deformity was described. Adam et al. reported a congenital parotid lipomatosis in which the tumor was well enveloped by parotid gland tissue.⁽⁷⁾ The term "congenital infil-

trating lipomatosis of the face" was first used in 1983, when Slavin et al. reported 3 cases of congenital proliferating and infiltrating lesions of adipose tissue involving the face. Rosa et al. reported another 3 cases and described the following characteristics of congenital infiltrating lipomatosis: 1) nonencapsulated proliferation of mature adipose tissue; 2) diffuse infiltration of muscle and adjacent soft tissue; 3) the presence of fibrous tissue with various nerve bundles and thickened wall vessels; 4) the absence of lipoblasts and signs of malignancy, in spite of a rapid growth rate; 5) hypertrophy of subjacent bone; and 6) being congenital in origin with a tendency to recur after surgical excision.^(8,9)

Vellios et al., in examining fetal tissue, observed that fetal fat tissue develops at a relatively later stage of development after the third and fourth months of intrauterine life.^(1,8) Small lobules formed by aggregated adipocytes first appear in areas of the subcutaneous region, buccal fat pad, axilla, and breast. Lipomatous degeneration of the face may occur at any site. Infiltrating lipomatosis of the face has been found on the cheek, buccal sulcus, tongue, lip, floor of the mouth, mental area, and parotid gland (Table 1). The etiology of this unusual tumor remains unknown. Trauma, chronic irradiation, muscular

metaplasia, degenerative processes with fatty transformation, multipotential cells of embryogenic origin under the influence of hormones, congenital cytomegalovirus infection, and alteration in chromosome 12 have been proposed as possible mechanisms for the lipomatous change.^(1,2,5,8-12)

Diagnosis of the tumor is made by clinical presentation and imaging study. CT and MRI remain the most-helpful tools for preoperative radiographic evaluation.^(2,13,14) The presence of a low attenuation, grossly infiltrating lesion with sometimes feathery appearance to the density on CT differs from other ordinary well-encapsulated lesions. Three-dimensional CT helps to reveal asymmetry of the facial bone due to osseous hypertrophy. MRI is useful in diagnosis by its signal density and accurately depicting the anatomic extent of lipomatous tumors. A biopsy is sometimes indicated to obtain an accurate histological identification before the definite operation. It should be noted that an inaccurate preoperative impression could be made due to the rarity of the disease and the feature of local facial swelling. The differential diagnosis of infiltrating lipomatosis of the face includes lymphangiomas, hemangiomas, pleomorphic adenomas, mucoepidermoid carcinomas, intermuscular or intramuscular lipomas, angiolipomas, fibrolipomas, liposarcomas, and lipoblastomas. Lipoblastomas occur exclusively in infancy and early childhood.⁽²⁾ Microscopic characteristics of the malignancy include immature adipose tissue with multivacuolated lipoblasts and signet ring cells, abundant plexiform blood vessels, and rich myxoid stromata. Most lipoblastomas occur on the extremities, trunk, and neck. Infiltrating lipomatosis differs from liposarcomas by the absence of lipoblastic activity and pleomorphism. Liposarcomas are usually located on the trunk and extremities, and rarely occur in infantile or congenital forms.⁽¹⁵⁾

The recurrence rate of infiltrating lipomatosis was estimated to be between 27.3% and 62.5%.^(1,2,11) It is not possible for complete removal of the facial lesions involving indispensable structures, as shown in our case. The main purpose of surgery is to improve the esthetic appearance rather than to eradicate the tumor. In order to preserve the important structures and to obtain satisfactory aesthetic results, some surgeons have managed these diseases with superficial parotidectomy and extensive facial nerve

dissection with the help of an electrostimulator. Of the 15 cases reviewed (Table 1), only 2 were reported to have no evidence of recurrence. In 1 case, the tumor was located within the parotid gland rendering radical extirpation of the tumor possible.⁽¹⁵⁾ In another case, no detailed information about the tumor location or size was given.^(2,3,6-9,12,13,15) Aggressive surgical resection or liposuction might not be an ideal treatment for congenital infiltrating lipomatosis of the face. There has been no documented case of infiltrating lipomatosis degenerating into a frank malignancy.⁽¹⁶⁾ We believe that individual surgical planning is required for each patient based on the extent of the tumor and the clinical findings.

In conclusion, congenital infiltrating lipomatosis of the face is a rare disease in infancy or childhood. It is characterized by its congenital origin, diffuse infiltration of mature adipose tissue over normal muscle fibers, rapid growth, associated osseous hyperplasia, and high recurrence rate after surgical intervention. The clinical presentation, biopsy, CT, and MRI help in making an accurate diagnosis before surgical treatment planning. The main purpose of surgery is to improve the cosmetic facial appearance rather than to eradicate the tumor.

REFERENCES

1. Enzinger FM, Weiss SW. Benign lipomatous tumors. *Soft Tissue Tumors*. 3rd ed. Baltimore: Williams & Wilkins Co, 1995:381.
2. Scherl MP, Som PM, Biller HF, Shah K. Recurrent infiltrating lipoma of the head and neck: Case report and literature reviews. *Arch Otolaryngol Head Neck Surg* 1986; 112:1210-2.
3. Bataineh AB, Mansour MJ, Abalkhail A. Oral infiltrating lipomas. *Br J Oral Maxillofac Surg* 1996;34:520-3.
4. Bennhoff DF, Wood JW. Infiltrating lipomata of the Head and Neck. *J Laryngoscope* 1978;88:839-48.
5. Coffin CM. Adipose and myxoid tumors. *Pediatric Soft Tissue Tumors: A Clinical, Pathological, and Therapeutic Approach*. Baltimore: Williams & Wilkins Co., 1997:254.
6. Johansen J, Berdal P. Lipomatosis of the parotid gland. *Acta Otolaryngol* 1970;263:167-9.
7. Adams G, Goycoolea MV, Foster C. *Otolaryngol Head Neck Surg* 1981;9:402-5.
8. Slavin SA, Baker DC, McCarthy JG, Mufarrij A. Congenital infiltrating lipomatosis of the face: clinicopathologic evaluation and treatment. *Plast Reconstr Surg* 1983;72:158-64.
9. De Rosa G, Cozzolino A, Guarino M, Giardino C.

- Congenital infiltrating lipomatosis of the face: report of cases and review of the literature. *J Oral Maxillofac Surg* 1987;45:879-83.
10. Pélissier A, Sawaf MH, Shabana AM. Infiltrating (intramuscular) benign lipoma of the head and neck. *J Oral Maxillofac Surg* 1991;49:1231-6.
 11. Lasso, J.M, España A, Alava D, Bazán A. Congenital infiltrating lipoma of the upper limb in a patient with von Willebrand disease. *Br J Dermatol* 2000;143:180-2.
 12. Donati L, Candiani P, Grappolini S, Klinger M, Signorini M. Congenital infiltrating lipomatosis of the face related to cytomegalovirus infection. *Br J Plast Surg* 1990;43:124-6.
 13. Cottrell DA, Norris LH, Doku HC. Orofacial lipomas diagnosed by CT and MRI. *JADA* 1993;124:110-5.
 14. Fulcinizi F, Califanre L, Zupi A, Veterini A. Accuracy of fine needle aspiration biopsy in head and neck tumor. *J Oral Maxillofac Surg* 1997;55:1094-7.
 15. Esposito C, Califano L, D'Armiento M, Longo F. Lipomatosis of the parotid gland in a child. *Br J Plast Surg* 2000;53:699-701.
 16. Görken C, Alper M, Bilkay U, Çelik N, Songür E. Congenital infiltrating lipomatosis of the face. *J Craniofac Surg* 1999;10:365-8.

顏面先天浸潤性脂肪瘤：病例報告和文獻回顧

陳建銘 羅綸洲 黃浩輝¹

顏面先天性浸潤性脂肪瘤是屬於脂肪腫瘤中罕見但獨具特色的一種，真正引起的原因目前仍然不明，由於是屬於先天性疾病，此腫瘤常於嬰幼兒或孩童時期被發現。臨床上顏面先天性浸潤性脂肪瘤具有下列特性：缺乏良好的包膜覆蓋，由成熟的脂肪組織廣泛浸潤正常的肌肉組織，生長快速，經常伴隨有異常骨質增生，和高手術復發率。由於腫瘤的廣泛浸潤與影響重要的顏面組織，往往無法以手術完全切除腫瘤。回顧過去文獻僅發現有十四個案例，在此我們報告另一個個案，此個案除了在左頰有浸潤性脂肪瘤外，同時也並存有以包膜覆蓋良好的脂肪瘤。(長庚醫誌 2002;25:194-200)

關鍵字：浸潤性脂肪瘤，顏面腫瘤，嬰兒。