



## Case Report

## Facial infiltrating lipomatosis, a rare cause of facial asymmetry to be known: Case report and literature review

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## ABSTRACT

Facial infiltrating lipomatosis is a rare lipomatous lesion, first described by Slavin in 1983. It is a benign pseudotumor pathology. It corresponds to a non-encapsulated collection of mature adipocytes infiltrating the local tissue and hyperplasia of underlying bone leading to a craniofacial deformity. Very few cases have been reported in the literature.

We report the case of a 19-year-old female patient, who was consulted for a swelling of the right hemiface progressively evolving since birth. Physical examination revealed facial asymmetry. On palpation, the mass was soft, painless, not compressible, not pulsatile, not fluctuating. In view of the asymptomatic nature and slow progression of the lesion, a lipomatous tumour, namely lipoma, was suggested. CT scan image shows a hyperplastic subcutaneous fat on the right hemiface. On the right jugal and temporal areas, there is a subcutaneous formation of fatty density, poorly limited, with no detectable peripheral capsule. It merges with the adjacent fat. In the bone window, there was a hyperplasia of underlying bone.

Facial lipomatosis infiltration of the face is a benign pseudotumor pathology. As a result, it can be confused with other disorders, in particular, hemifacial hyperplasia. Combination of physical and radiological findings can establish the diagnosis. Surgical treatment is done for cosmetic purposes.

### 1. Introduction

Facial infiltrating lipomatosis or congenital infiltrating lipomatosis of the face (CILF) is a very rare disorder in which mature lipocytes invade into adjacent tissues in the facial region. The manifestation is always unilateral, with hypertrophy of both soft and hard structures on the affected side of the face. Congenital infiltrating lipomatosis of the face is a nonhereditary disease whose etiopathogenesis is not well understood. Mutation of the PIK3CA and FGFR3 gene is identified as the cause. The gradual enlargement of the affected side of the face and the resulting facial asymmetry are crucial features [1–3].

It was first described by Slavin et al., in 1983. Congenital infiltrating lipomatosis represents a distinct clinicopathologic entity characterized by collections of nonencapsulated, mature lipocytes that infiltrate local tissues. The lipomatous tissue can infiltrate facial muscles, soft tissues and bones to cause ipsilateral hyperplasia of the underlying facial skeleton, asymmetry of the mandible and even dental abnormalities [4,

5]. These children are asymptomatic and have normal psychomotor development. The main concern is the esthetic appearance, which is the reason why it is often neglected.

Because CILF is very rare, few cases have been reported in the literature. We hope that our report and review of literatures will give more suggestions for the diagnosis and treatment of this disease.

### 2. Case presentation

Our work consists of a single case report and has been reported in line with the SCARE 2020 criteria [6].

We report the case of female patient who presented with a tumefaction of the right hemiface progressively evolving since birth. She decided to consult at the age of 19 years, because of esthetic damage. There was no other personal or family pathological history.

Physical examination revealed facial asymmetry. The right side of the face was swollen without any skin abnormality. A deviation of the

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chin to the left. Hypertrophy of the cheek, the lower lip and the auricle on the right side. Oral examination revealed a discreet macroglossia and hypertrophy of the teeth on the right side.

On palpation, the mass was soft, painless, not compressible, not pulsatile, not fluctuating. The oral mucosa appeared normal. There was no sensitivity or motor disturbance. There was no cervical adenopathy. In view of the asymptomatic nature and slow progression of the lesion, a lipomatous tumour, namely lipoma, was suggested.

A facial CT scan was performed. CT image (soft tissue algorithm) shows a hyperplastic subcutaneous fat on the right hemiface. The fatty infiltration is diffuse in the superficial regions of the face. This infiltration realize on the right jugal and temporal areas, a subcutaneous formation of fatty density, poorly limited, with no detectable peripheral capsule. It merges with the adjacent fat. There are a few septa, enhanced after injection of PDC, corresponding to vessels.

In the bone window, images show hyperplasia of underlying bone: zygomatic, maxillary, mandible and teeth on the homolateral side. **Figs. 1 and 2.**

The diagnosis of Congenital Infiltrating Lipomatosis of the Face was retained in view of the examination data.

The patient received surgical treatment. Resection of the bony outgrowth of the zygomatic bone and reduction of the fat mass. Follow-up CT scan was performed 3 months after surgery. **Fig. 3:**

### 3. Discussion

Lipomatoses are infiltrating and/or diffuse non-neoplastic proliferations of mature fat tissue that resemble and may be confused with well-differentiated liposarcoma. This group of lesions according to the classification proposed by Enzinger and Weiss' in 1983, comprises three different entities: cervical symmetrical lipomatosis of the neck, pelvic lipomatosis involving the perirectal and perivesical regions and diffuse lipomatosis of the limbs or of the trunk [7,8].

Facial lipomatosis was not included. Recently, Slavin et al. described CIL-F for the first time in 1983, with histopathologic characteristics of non-encapsulated mass containing mature adipocytes, fat infiltration in muscles and adjacent soft tissue, no sign of malignant characteristics, absence of lipoblasts, detection of fibrous elements, and increased numbers of vessels and nerves [2].

There is infiltration of normal tissue by mature adipocytes, which help distinguish FIL from lipoblastomatosis and liposarcoma that have undifferentiated or immature adipocytes. Areas of normal tissue involved include submucosa, skeletal muscle, and parotid, submandibular, and minor salivary glands [2].

Physical findings: Commonly, infiltrating lipomatosis presents with a diffuse large swelling on the side of the face that is soft, fluctuant, and non-tender. The clinical features of facial lipomatosis include unilateral

hypertrophy of soft tissues of the face, most commonly the cheek, with underlying fat infiltration and skeletal overgrowth, cutaneous capillary blush (usually after resection), macrodontia on the affected side, abnormal root formation, early eruption of deciduous and permanent teeth on the affected side, macroglossia, and protuberances on the tongue and buccal mucosa, which are representative of underlying mucosal neuromas [2–10].

#### Fig. 4:

Radiological findings: show hypertrophy of facial bones and soft tissue swelling. Magnetic resonance imaging is probably the most helpful study because it shows diffuse fatty infiltration and increased thickness of subcutaneous fat on the affected side. Specifically, a bright signal on both T1-and T2-weighted spin-echo sequences with fatty extension into adjacent soft tissue is found. Computed tomography can also be helpful because it typically shows a non-encapsulated diffusely infiltrating low-attenuation mass that usually measures between  $-65$  UH and  $-125$  UH. In addition to bony changes, thickening of the right frontal, zygomatic, maxillary, and mandible bones [1, 2, 3, 4 5,5,7,8, 9,10]. **Fig. 5:**

The differential diagnoses include the Proteus syndrome, encephalocutaneous lipomatosis, and vascular malformations (hemangioma). Proteus syndrome is characterized by overgrowth of tissues from all three germ layers. The spectrum consists of hemihypertrophy, facial hamartomas, macrodactyly, and hyperkeratotic rugae of the soles. Encephalocutaneous lipomatosis is characterized by lipomas of the scalp and central nervous system as well as focal alopecia [5].

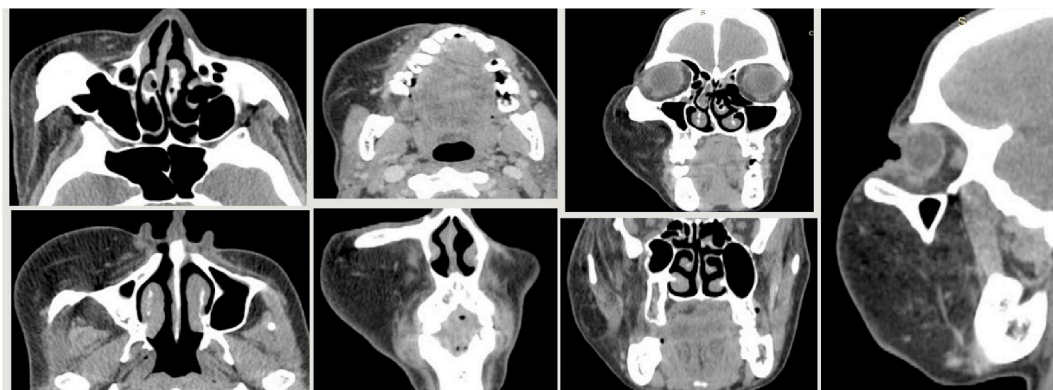
Treatment: While no definitive treatment exists for FIL, various forms of surgical treatment have been attempted with limited success owing to high recurrence rates and surgical risk to important anatomic structures. Earlier literature favored aggressive surgical treatment, but recent articles have recommended a more conservative approach. We also recommend a conservative surgical approach with treatment only for symptomatic cases such speech dysfunction, and gross cosmetic deformity. We also emphasize the importance of a multidisciplinary approach. Also liposuction and excision is done for cosmetic reasons [2, 11].

### 4. Conclusion

Congenital infiltrating lipomatosis of the face is a rare benign condition occurring in childhood leading to craniofacial deformity. It is corresponded to a non-encapsulated collection of mature adipocyte infiltrating the subcutaneous and muscle planes associate with bony hypertrophy.

Combination of clinical examination and imaging (CT scan and MRI) can establish the diagnosis. Surgery is done for cosmetic purpose.

The recurrence rate is very high after surgical excision, so adjuvant



**Fig. 1.** Axial, coronal and sagittal CT scan (soft tissue): image show a fat-density lesion in the subcutaneous, non-encapsulated. Hyperplasia and fatty infiltration in muscular, and intermuscular planes of the right side of the face. Septa, enhanced after injection of PDC corresponding to vascular structures. Narrowing of the right maxillary sinus cavity.

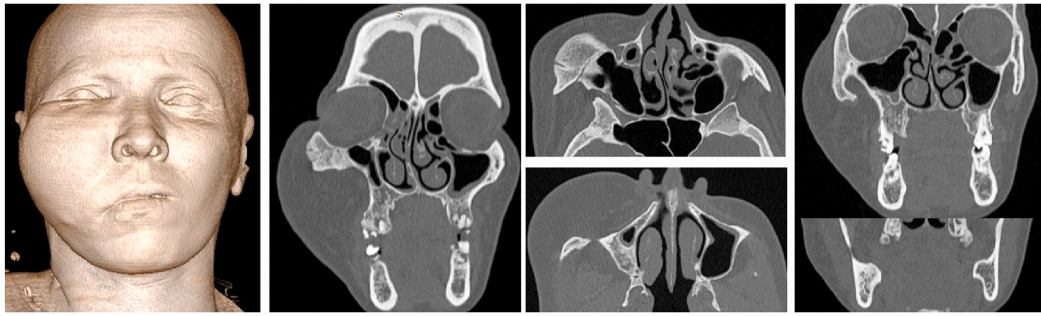


Fig. 2. Axial and coronal CT image (bone algorithm) shows thickening of the right hemifacial bone structures (zygomatic, maxillary, mandible and teeth). Deformation and new bone formation on right zygomatic bone.

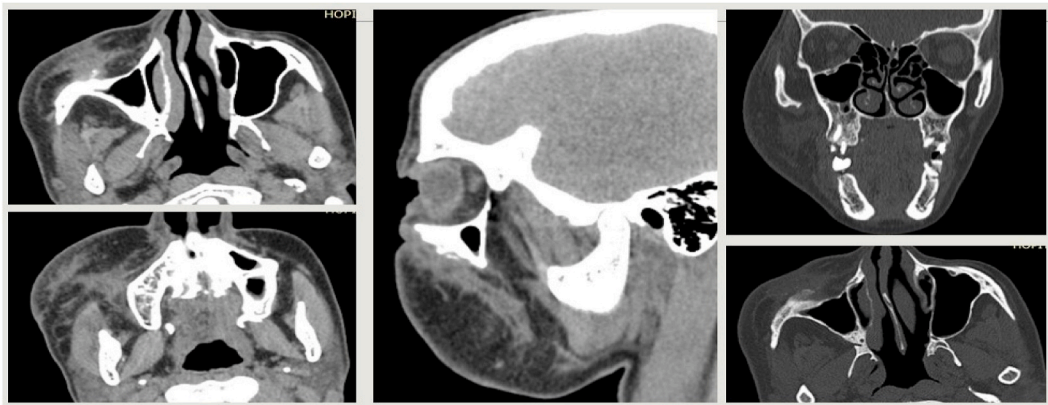


Fig. 3. Control CT scan after 3 months; resection of the bony outgrowth of the zygomatic bone and reduction of the fat mass. Local infiltration is noted in relation to posttherapeutic remodelling.

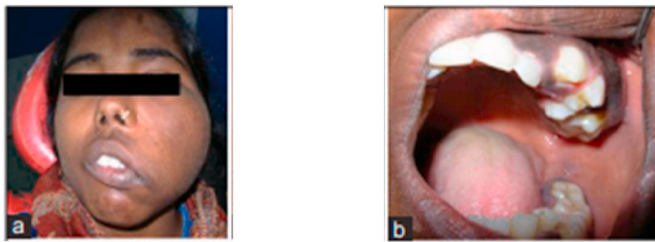


Fig. 4. (a) Large swelling on the left side of the face; (b) Intraoral expansion of maxillary alveolar process [10].

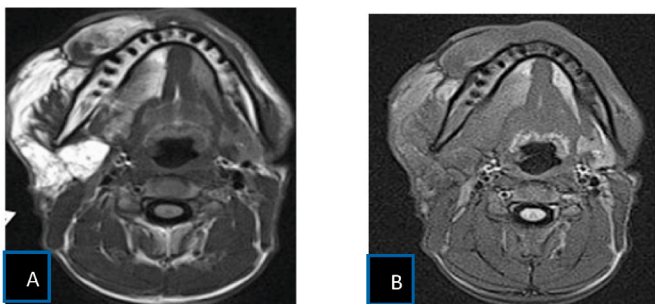


Fig. 5. (A) Axial T2W showing fatty infiltration of right parotid gland (arrows) and buccal fat pad hypertrophy. (B) Fat suppression image showing suppression of fatty infiltration of right side facial tissue [7].

therapy and psychological intervention are important for patients.

**Provenance and peer review**

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**Ethical approval**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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**Author contribution**

Harouna Maman Siradji: Corresponding author, writing the paper. Belgadir Hasna: writing and correction of the paper. Fadoul Achta: writing the paper. Aghrib Fatiha: writing the paper. Merzem Aicha: Correction of the paper. Amris Omar: Correction of the paper. Moussali Nadia: Correction of the paper. Elbenna Naima: Correction of the paper.

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#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Declaration of interest statement

No interest statement.

#### Declaration of competing interest

Authors of this article have no conflict or competing interests. All of the authors approved the final version of the manuscript.

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