Facial Infiltrating Lipomatosis: Case report and a review of literature

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ABSTRACT:

Facial infiltration lipomatosis is a rare disorder in which overgrowth of fatty tissue with infiltration of underlying structures leads to craniofacial deformities and asymmetry. We report the case of a 29-year-old female patient, who was consulted for a swelling of the right hemiface progressively evolving since birth. CT scan and MRI image shows a hyperplastic subcutaneous fat on the right hemiface. Also infiltrated the ipsilateral muscles of mastication and extended into the ipsilateral parapharyngeal fat, the side wall of the cavum, the right hemilangue and the submandibular gland.

Keywords: Facial asymmetry, Lipomatosis, Clinique, Imaging.

INTRODUCTION:

Facial Infiltrating Lipomatosis is a very rare lipomatous lesion or congenital infiltrating lipomatosis of the face (CILF). 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 [1]. In this case report, we present an additional case of CILF, with hyperplasia of underlying bone: zygomatic, maxillary and mandible on the homolateral side. 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.

CASE REPORT:

A 29-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.

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, no limited, with no detectable peripheral capsule. also infiltrated the ipsilateral muscles of mastication and extended into the ipsilateral parapharyngeal fat, retromolar trigone, the side wall of the cavum, the right hemilangue and submandibular (Figure 1)

In the bone window, images show hyperplasia of underlying bone: zygomatic, maxillary, mandible and teeth on the homolateral side

MR imaging revealed similar findings, No abnormal enhancement was identified (Figure 2).

A diagnosis of CILF was made based on imaging findings, and clinical presentation.

DISCUSSION:

CILF is a rare disorder of infancy and early childhood and is classified as a subgroup of lipoma. The aetiology is poorly understood. Slavin et al.6 introduced the term ‘congenital infiltrating lipomatosis of the face’ in 1983[2].

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 perivescical regions and diffuse lipomatosis of the limbs or of the trunk [3].

The common clinical presentation of CILF is facial asymmetry at birth. Associated features described include: vascular blush, ptosis, enlargement of the lower lip, ipsilateral hemimacroglossia, increased facial hair on the affected side, early eruption of deciduous and permanent teeth, missing teeth associated with dentigerous cysts and ipsilateral hypertrophy of the underlying facial skeleton [4].

The main role of imaging is to define the extent of lipomatous infiltration in underlying structures and facilitate the planning of surgery.
On CT scans, lipomas appear as homogeneous hypoattenuated masses. They have a CT number ranging from −60 to −120 HU and do not typically show contrast enhancement. In some cases, centrally enhanced areas, corresponding to lipomatous lesions with inflammatory cell infiltration, have been identified. 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 [5].

The differential diagnoses of CILF include vascular malformation, hemangioma, lipoma, liposarcoma, lipoblastomatosis and other overgrowth syndromes. Vascular malformation or hemangioma could be easily differentiated on CT and MRI. Most lipomas are clearly defined and well-encapsulated. Intermuscular lipoma is ill-defined but usually restricted to one or more muscles. Well-differentiated liposarcoma and lipoblastomatosis that involve fat tissue infiltration also need to be ruled out[6].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 [7]

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 anatomical 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 [8,9]

CONCLUSION:

CILF is a rare congenital disorder characterised by overgrowth of fatty tissue of the face which leads to craniofacial deformity and asymmetry. 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.

REFERENCES:
Figure 1:

Axial CT scan (soft tissue): hyperplastic subcutaneous fat on the right hemiface. The fatty infiltration is diffuse in the superficial regions of the face. Also infiltrated the ipsilateral muscles of mastication and the submandibular gland and extended into the ipsilateral parapharyngeal fat, the side wall of the cavum, the right hemilangue

Axial and coronal CT image (bone algorithm): shows thickening of the right hemi facial bone structures (zygomatic, maxillary, mandible)
Figure 2:
Axial T2, T1, T2 IDEAL MRI demonstrates an extensive, non-encapsulated right facial mass with fat signal extending into the soft tissue spaces of the face. Also infiltrated the ipsilateral muscles of mastication and the submandibular gland and extended into the ipsilateral parapharyngeal fat, the side wall of the cavum, the right hemilangue. Axial fat suppressed image demonstrates complete suppression of signal within the lesion consistent with adipose tissue.