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INFILTRATIVE LIPOMATOSIS A RARE CASE REPORT IN LEBANON

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ABSTRACT
Congenital infiltrative lipomatosis of the face (CIL-F) is rare and difficult to treat. It compromises a subgroup of Lipomatous tumors and characterized by collection of nonencapsulated mature lipocytes which infiltrates local tissue resulting in craniofacial deformities. The tumor-like lesion has rapid growth, associated with osseous hyperplasia and high recurrence rate after surgical intervention. Complete surgical excision is often impossible due to its diffuse infiltration and involvement of important facial structures. A few cases CIL-F was found in a literature review.

KEYWORDS: Congenital Infiltrative Lipomatous of face (CIL-F), Facial Tumor Infiltrates.

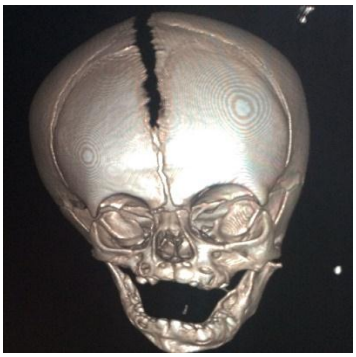
INTRODUCTION
Congenital infiltrative lipomatosis of the face (CIL-F) is rare and difficult to treat. It compromises a subgroup of Lipomatous tumors and characterized by collection of nonencapsulated mature lipocytes which infiltrates local tissue resulting in craniofacial deformities. We present additional case of one month old girl who had diffuse infiltrating lipomatosis that presented with a swelling of the face since birth. The three dimensions CT and MRI showed infiltrated soft tissue mass of fatty density.[1]

Slavin et Al was the first that described CIL-F in 1983 as a distinct clinical pathology entity, as a congenital disorder in which mature adipocytes invade adjacent tissue in the facial region.[2]

CASE REPORT
This one month old baby girl was found to have left mass since birth. The antenatal history showed no relevant episodes during pregnancy such as drug use, exposure to teratogenic agents, infectious disease or difficult labor. Local physical examination revealed diffuse swelling on the left side of the face with distortion of facial structures such as eye, nose and lip on the lesion side. No breathing, no swallowing, and no sucking problems. The lesion was extending superiorly from the infraorbital area up to lower border of the mandible, inferiorly and medially from the midline of the face up to the preauricular area laterally measuring approximately 5, 5 cm in size. On palpation, the tumor was elastic, nontender with ill-defined border, non-pulsatile, non-compressing and no pus discharge. The overlying skin was intact.

Upon admission 3D CT scan of the face and the brain showed hypertrophy and asymmetry of the left mandible and zygoma.

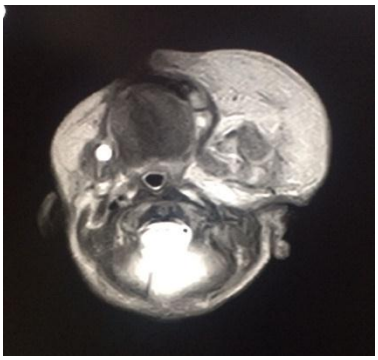
A



B

Three dimensional computed tomography study. (A) Soft tissue imaging showing a large mass. (B) There is hypertrophy of the left maxillary and zygoma.

MRI with and without contrast was performed and showed diffuse infiltrating tissue of fatty component occupying the left cheek without a clear boundary with fatty infiltration to the left cerebellar hemisphere of 15 mm of diameter.



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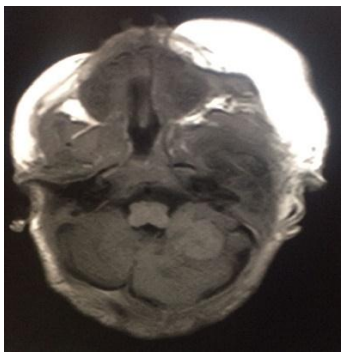

B

Magnetic Resonance Imaging (MRI) revealed: (A) T2W1 MRI demonstrates an extensive non-encapsulated left facial fatty mass extending into the soft tissue of three cm thickening. The subcutaneous and intermuscular planes were involved (masticator, temporal and lateral pterygoidien muscles). (B) T1W1 MRI showed non enhancing ill-defined infiltrative inhomogeneous (isointense to fat) in the subcutaneous muscular and intermuscular planes of the face on the left side and the presence of nodular lesion on the left cerebellar hemisphere of 15 mm OD diameter. Because of high rate of recurrence and small age, it was decided to observe the mass and postpone the surgery until three months of age.

Reported cases of congenital infiltrating lipomatosis of the face

No	Gender	Age of Report	Location	*Outcome*	Reference
1	M	5 y	Left cheek	Recurrence	Scherl, et al
2	M	6 y	Right cheek	No Recurrence	Bataineh, et al
3	F	15 y	Left cheek	-	Bataineh, et al
4	F	2 y 6 m	Right parotid area	-	Johansen, et al
5	-	2 m	Right parotid area	-	Adams, et al
6	M	2 y	Right parotid region	Loss of follow up	Slavin, et al
7	F	9 y	Right temporal area	recurrence	Slavin, et al
8	F	2 y	Right buccal area	recurrence	Slavin, et al
9	F	9 y	Left cheek	Residual tumor	DeRosa, et al
10	F	23 y	Right sub mental area	recurrence	DeRosa, et al
11	M	53 y	Left cheek	recurrence	DeRosa, et al
12	M	5 m	Left cheek	death	Donati, et al
13	M	4 m	Left parotid region	No recurrence	Esposito, et al
14	F	6 y	Right cheek	Residual tumor	Cenk Gorken, et al
15	F	1 y 6 m	Left cheek	Residual tumor	Chien-Ming, et al
16	F	16 y	Left cheek	-	Kamal, et al
17	F	18 y	Left cheek	-	Kamal, et al
18	M	2 y	Right cheek	Not operated	Kamal, et al
19	F	3 Y 6 m	Left cheek	recurrence	Kim, et al
20	M	11 y	Right cheek	-	Rajeswaran, et al
21	F	1 m	Left cheek	Not operated	Bassem, et al (This Report)

DISCUSSION

Lipomas are benign neoplasms compromised of fat and are typically asymptomatic. They present as soft and ill-defined, slowly enlarging masses, easy to remove and rarely recurrent after removal. These tumors are further categorized into several subgroups:

1- Simple encapsulated lipoma
2- Lipoma variants like angiomyolipoma
3- Hamartomatous lesion
4- Infiltrating or diffuse lipomatosis
5- Benign tumor of brown fat hibernoma.[3,4]

Infiltrating lipomatosis is characterized by painless diffuse deposits of mature fat cells over normal muscle fibers, a rapid growth rate, associated osseous hyperplasia and a high recurrence rate after surgical

intervention. Pathologically, this lesion show the following characteristics as by Rosa et Al: 1) - Non-encapsulated congenital fatty tumors, 2) - Infiltrate adjacent muscles and soft tissues, 3) - Presence of fibrous tissue with various nerve bundles and thickened wall vessels, 4) - Absence of lipoblasts and signs of malignancy, 5) - Hypertrophy of adjacent bone, 6) - Being congenital in origin with a tendency to recur after surgical incision. Infiltrative lipomatosis of the face has been found on the cheek, buccal sulcus, tongue and lip, floor of the mouth, mental area and parotid gland. The etiology of this rare tumor remains unknown. Trauma, chronic irradiation, muscular metaplasia, degenerative processes with fatty transformation, multi potential cells of embryogenic origin under the influence of hormones, congenital CMV and alteration in chromosome 12 have been proposed as possible etiologies for the lipomatous change. Diagnosis is made by clinical presentation and imaging study. CT and MRI remain the most helpful tools for preoperative radiographic evaluation. 3 D CT helps to reveal asymmetry of the facial bone due to osseous hypertrophy. MRI helps in depicting the anatomic extent of the lipomatous tumor. Histological identification is obtained by a biopsy before the definite operation. Differential diagnoses are: Lymphangiomas, Hemangiomas, Pleomorphic Adenomas, Liposarcomas, Lipoblastomas and Fibrolipomas. The recurrence rate of infiltrating lipomatosis is very high between 27% and 63%. We believe that individual surgical planning is required for patient based on tumor extent and clinical findings. No surgical procedure must be performed until the end of the facial growth period and the advantages of delaying surgery were: minimizing the chances of damage to the facial nerve, decreasing the total number of procedures having a mature contralateral cheek contour to match.

CONCLUSION

CIL-F is a rare benign condition that usually occurs in adulthood. It's characterized by mature adipocytes that infiltrate subcutaneous and muscle plans and causing bony hypertrophy. Accurate diagnosis is made by: clinical presentation, biopsy, CT and MRI. The surgery is performed for cosmetic facial appearance.

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