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Case Report

Congenital infiltrating lipomatosis of the face

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Introduction

Congenital infiltrating lipomatosis (CIL-F) is a very rare entity with less than 50 reported cases[1]. Its etiology is poorly understood. CIL-F is characterized by diffuse infiltration of soft tissue and bones by mature non-encapsulated adipose tissue, causing unilateral facial skeletal hyperplasia. However, the affected individuals show normal psychomotor development[1]. Aesthetics and high rate of recurrence after surgical excision are the main concerns[1-3]. We report a case of CIL-F, in a 28 year old male patient diagnosed based on radiological and pathological findings.

Case history

A 28 year old male patient was referred to the MRI unit for imaging of recurrent right side facial swelling. This had been first noticed at the age of six years. He had undergone surgical excision four times, with recurrence after each surgery.

On physical examination there was right sided facial enlargement with several skin tags over the cheek and mandible (Figure 1). CT scan revealed thickening of the subcutaneous fat and buccal fat pad on the right side of the face with fatty infiltration of the medial pterygoid, platysma and masseter muscles resulting in enlargement of the muscles (Figure 2). Fatty infiltration was also seen in the right parotid gland (Figure 3). There was enlargement of the right ramus of the mandible and filling of the mandibular canal with fatty tissue (Figure 2(A)). No obvious abnormality was seen in the brain parenchyma.

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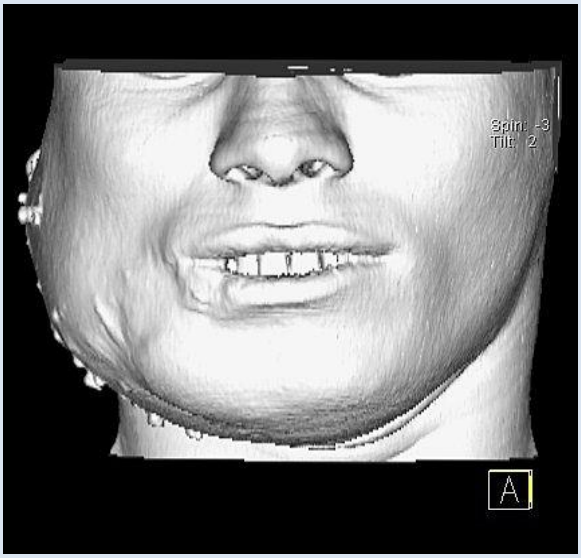

Figure 1 : Three-dimensional volumetric reconstruction frontal image of face showing right side facial swelling with skin tags

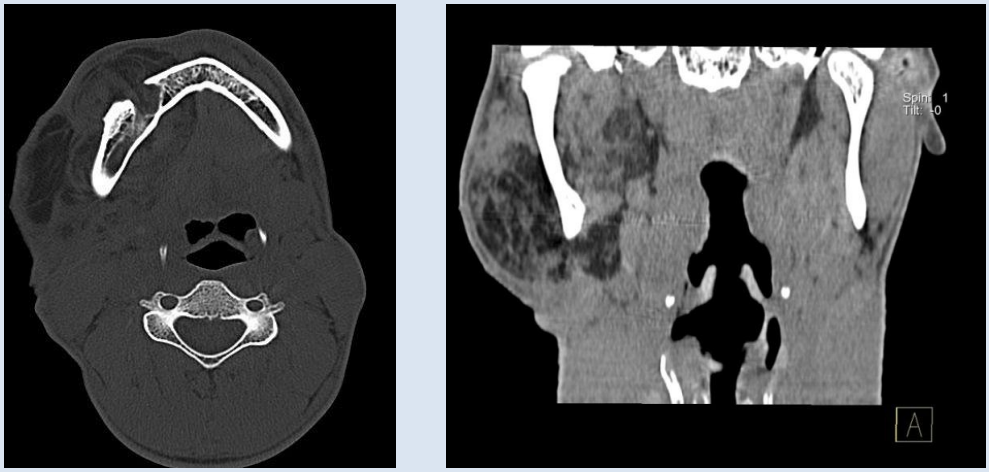

Figure 2 : (A) Axial CT (bone algorithm) showing widened mandibular canal with fatty infiltration (thick arrow) and hyperplastic subcutaneous fat (B) coronal CT images demonstrates fatty infiltration of right masseter and pterygoid (arrow) muscles

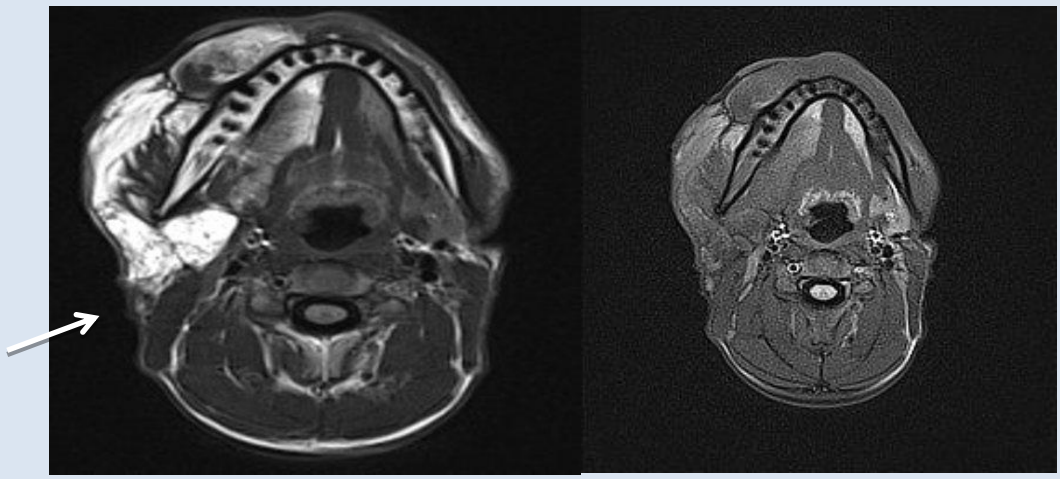

Figure 3 : (A) Axial T2W showing fatty infiltration of right parotid gland (arrows) and buccal fat pad hypertrophy. (B) Fat suppression image showing suppression of fatty infiltration of right side facial tissue

MRI evaluation was done by 1.5-T scanner. It showed fatty infiltration of subcutaneous tissue, muscular and intermuscular planes and parapharyngeal spaces on the right side of the face.

Histopathological examination of growth removed at surgery showed non-capsulate mature adipocyte infiltration. No evidence of malignant cells. Based on the above findings a diagnosis of congenital infiltrating lipomatosis of the face was made.

Discussion

Congenital infiltrating facial lipomatosis is a rare condition resulting in diffuse fatty infiltration of muscle fibers, soft tissue and bones of the face giving rise to facial skeletal hypertrophy[1,2]. Its etiology is unclear and described in literature as related to trauma, chronic irradiation and muscle metaplasia or embryonic under the influence of hormones, congenital cytomegalovirus infection or alteration in chromosome 12[4,5].

This entity was first described in 1983 by Slavin et al[5]. He reported three cases of congenital proliferating and infiltrating lesions of the adipose tissue involving the face. Radiography shows unilateral facial bone hypertrophy and adjacent soft tissue swelling. CT and MRI play a major role in the diagnosis as they can delineate the fatty nature and extent of involvement. In MRI, T1 and T2W images show heterogeneously high signal, with suppression in fat suppression images. Local infiltration and extension within facial planes can be better delineated by MRI due to its better resolution and multiplanar capabilities[1].

Proteus syndrome, lymphangioma, encephalocutaneous lipomatosis, neurofibromatosis of von Recklinghausen and hemangioma are considered in the differential diagnoses of this presentation[1].

CIL-F has an uncertain prognosis and a high recurrence rate (>60%) after surgical intervention. A commonly used treatment option is liposuction. Surgical procedure must be delayed until the

end of facial growth in order to minimize the facial nerve damage and reduce the total number of interventions[4].

Pre-operative diagnosis is usually based on imaging with CT and MRI[1]. Surgery is offered to improve appearance.

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