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Original Paper

Congenital infiltrating lipomatosis of the face

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Abstract Congenital lipomatosis of the face is a very rare syndrome defined as a collection of non-encapsulated mature lipocytes infiltrating local tissues: resulting in cranio-facial deformities. Due to the normal psychomotor development of the children, aesthetic appearance often remains the main concern. After a precise description of a case, a complete overview of the literature data was made. New clinicopathologic data concerning bone involvement will be presented and analyzed in the light of embryonic growth.

Keywords Congenital - Lipomatosis - Vibroliposuction

Introduction

Congenital lipomatosis of the face is a very rare syndrome, first described by Slavin et al. [14] followed by other reports, the total number of cases being 28. As with many other birth defects, the etiopathogeny is poorly understood. However, due to the normal psychomotor development of the children, aesthetic appearance often remains the main concern.

After a precise description of a case, a complete overview of the literature is presented. New clinicopathologic data concerning bone involvement will be discussed and analyzed in the light of embryonic growth.

Case report

A 43-month-old girl was referred to the plastic surgery department with a right diffuse hemi facial deformity. This had been noticed at birth, but was still increasing in size. Clinical

examination demonstrated an infiltration of the subcutaneous tissue extending from the cheek and the mandible to the right fronto-temporal region (Fig. 1). The lesion was soft and painless. We noticed a hyperaemia on the right cheek but no sign of facial nerve compression or bony overgrowth. Although dental occlusion was normal three masses were located on the right side of the lower lip (Fig. 1).

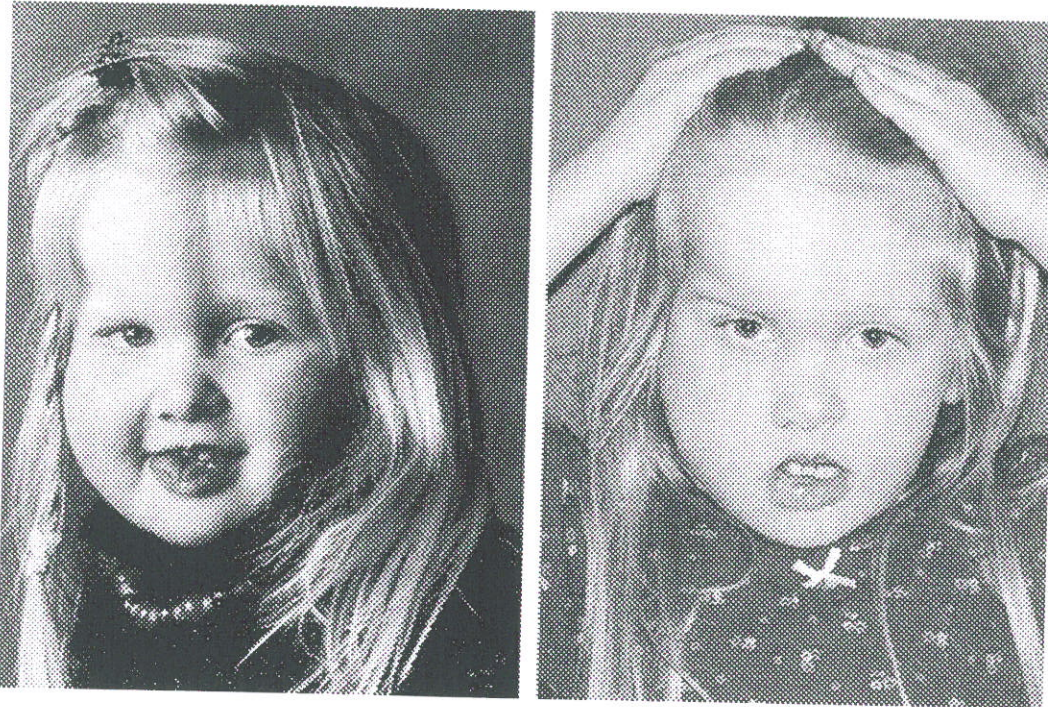


Fig. 1 Clinical pictures. *Left* preop view, *Right* postop view

The CT imaging and MRI did not show central nervous deformity nor ocular, bony, arterial or venous anomaly. A non-encapsulated fatty tissue was infiltrating the cheek, the masseteric and pterygoid region. Fatty tissue was also noticed in the right fronto-temporal subcutaneous layer and diploe (Fig. 2).