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LIPOBLASTOMA-LIPOBLASTOMATOSIS ASSOCIATED WITH UNILATERAL LIMB HYPERTROPHY: A CASE REPORT IN A NEWBORN

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ABSTRACT

Lipoblastoma (1) is a benign mesenchymal tumor of embryonic adipose tissue, uncommon in infancy. Multicentricity, absence of a capsule and histopathology best defines a lipoblastoma (2,3). Synonyms for this lesion are (4) embryonic lipoma, fetal lipoma, lipoblastic tumor, and congenital lipomatoid tumor.

Lipoblastoma is more common in males (~80%) (5), is usually located in the subcutaneous soft tissue (benign lipoblastoma) or in the deep interstitium (benign lipoblastomatosis), or sometimes in all corporeal segments.

Primary treatment is complete excision. Relapse is between 14-25%, many of which are more "mature," and difficult to differentiate from lipoma. The differential diagnosis includes liposarcoma, which is rare under ten years (6). Radical excision in children is recommended with relapses, especially with lipoblastomatosis (7). Chromosomal markers help discriminate between liposarcoma and lipoblastoma (8).

CASE REPORT

A one-month old female baby presented with an asymptomatic soft tissue tumor on the bottom of the left foot (three times bigger volume than a contralateral unaffected foot) without specific signs, but with otherwise normal growth of toes and similar length of feet and limbs *(Fig. 1a,b)*. There was no other tumor in other parts of the body. Magnetic resonance imaging (MRI) showed a lobulated architecture and a peripheral capsule *(Fig. 1c)*. Plain x-rays disclosed abnormal growth of the 4th metatarsal with continuous periosteum, hypertrophic tarsus and metatarsus in the involved foot *(Fig. 1d)*.

Initial treatment was an extended but not radical excision of the tumor. Postoperative function and sensitivity of the foot was intact.

At operation, the lesion was a lobulated fat tumor, infiltrating the spaces between the bones but not specific tissues on the foot, and was pseudoencapsulated with difficulty in determining the limits between normal and abnormal fat tissue. Hemostasis was excellent, there were no grossly abnormal blood vessels and no damage to what appeared to be a normal blood circulation.

Total volume of the resected specimen was 200 mg. There was proliferation of adipocytes in different stages of maturity, confirming a well-lobulated tumor. Lipoblasts mixed with well-differentiated adipocytes *(Fig. 2)*. The fat tumor involved all other structures without infiltration (9), and the final diagnosis was lipoblastomatosis (10).

The post-operative course was without complications. Over the next several months, there was increased discrepancy of the length

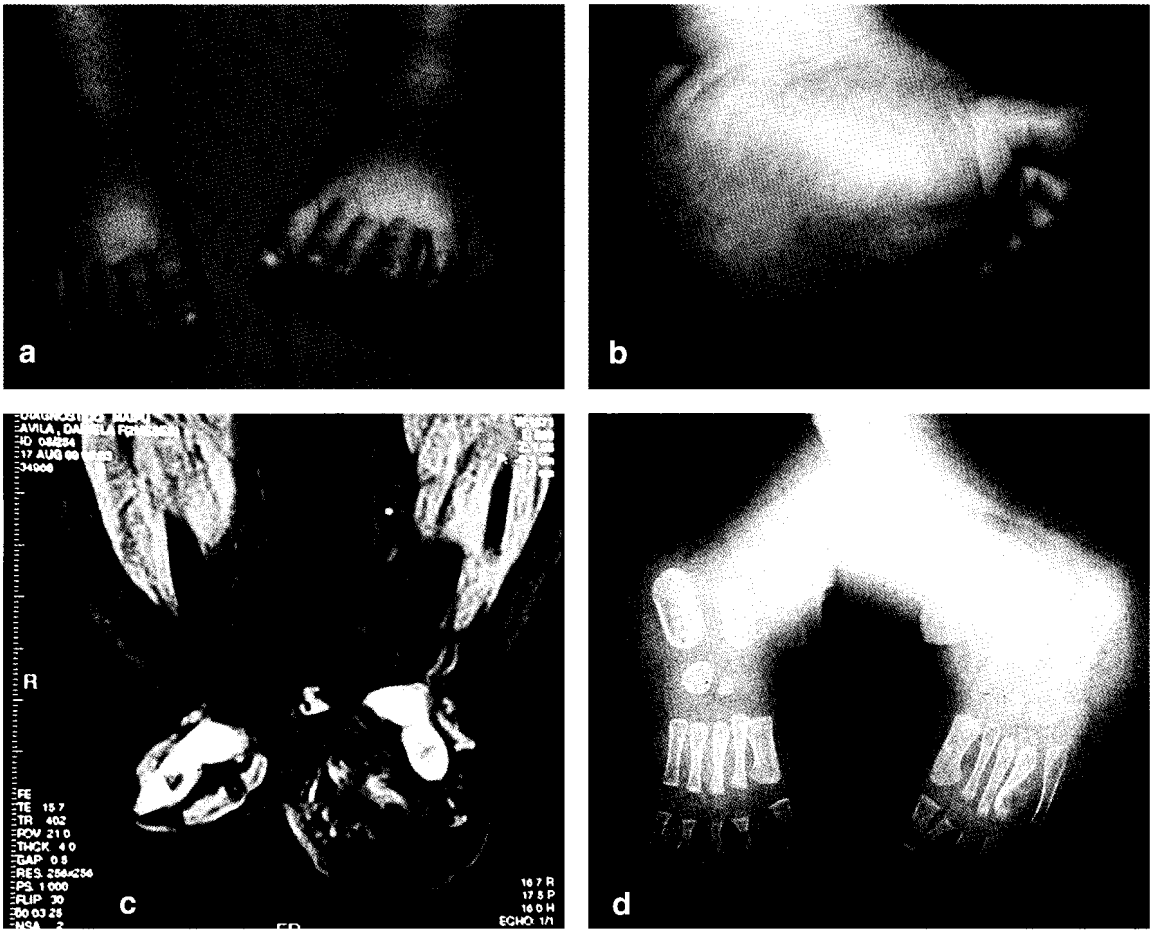

Fig. 1: (a) Pre-operative image of both feet; (b) lateral view of the affected foot; (c) MRI without specific signs of lipoblastoma; (d) comparative plain x-ray of feet. Nonspecific lesion of the 4th metatarsal bone in its metaphysis (arrow).

and volume of the lower limb (hypertrophy) on the affected side, with increased size (recurrence) of the tumor on the sole and both malleolar regions (pseudohypertrophy). At the age of eight months, reoperation was undertaken with attempt at complete resection *(Fig. 3a)*. At age three years, there was discrepancy of limb length of ~2 cm. The abnormal growth of the 4th metatarsal bone is still seen *(Fig. 3b)*.

DISCUSSION

Soft tissue tumors in the newborn and the first year of life are a clinical and surgical challenge. Diagnosis before operation is not always possible. An example is lipoblastoma. This report requires other considerations. First, the peculiar bone lesion which is unclear was not progressive. Second, hypertrophy (increase of the bone growth in proportion to the increase of the soft tissue enlargement) is not easily explained in comparison with the healthy side. This association may be coincidental or the confluence of three derangements namely, lipoblastomatosis, hypertrophy, and dysmorphogenic bone growth. Each aspect can have an established explanation separately. Dysmesenchymal lipogenesis, in

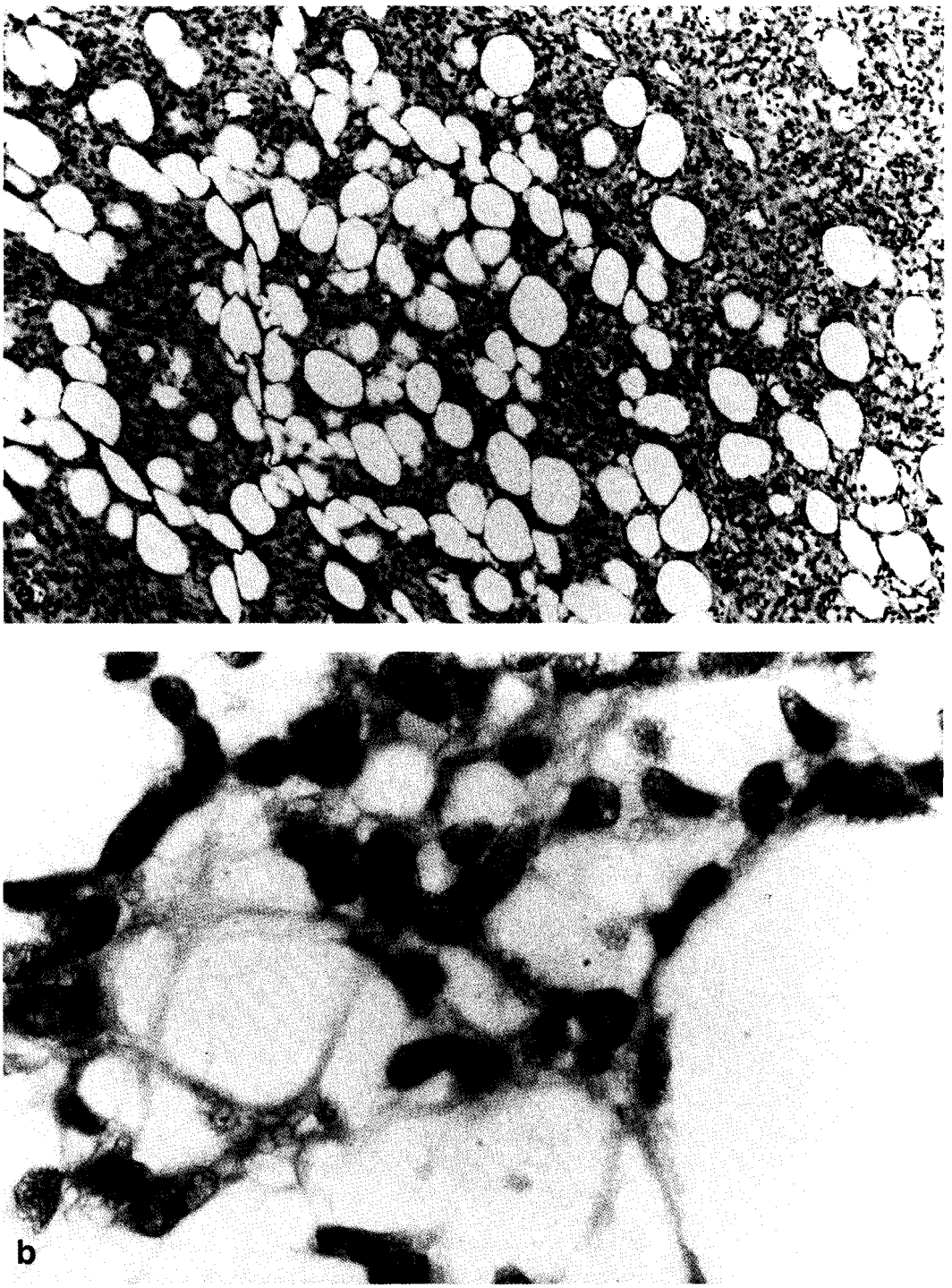

Fig. 2: Histology of resected tissue. Proliferating lobulated adipocytes in different stages of maturation; lipoblasts mixed with well differentiated adipocytes. Mitotic activity not prominent. Low and high magnification. [H&E, (a) 200x and (b) 1000x]

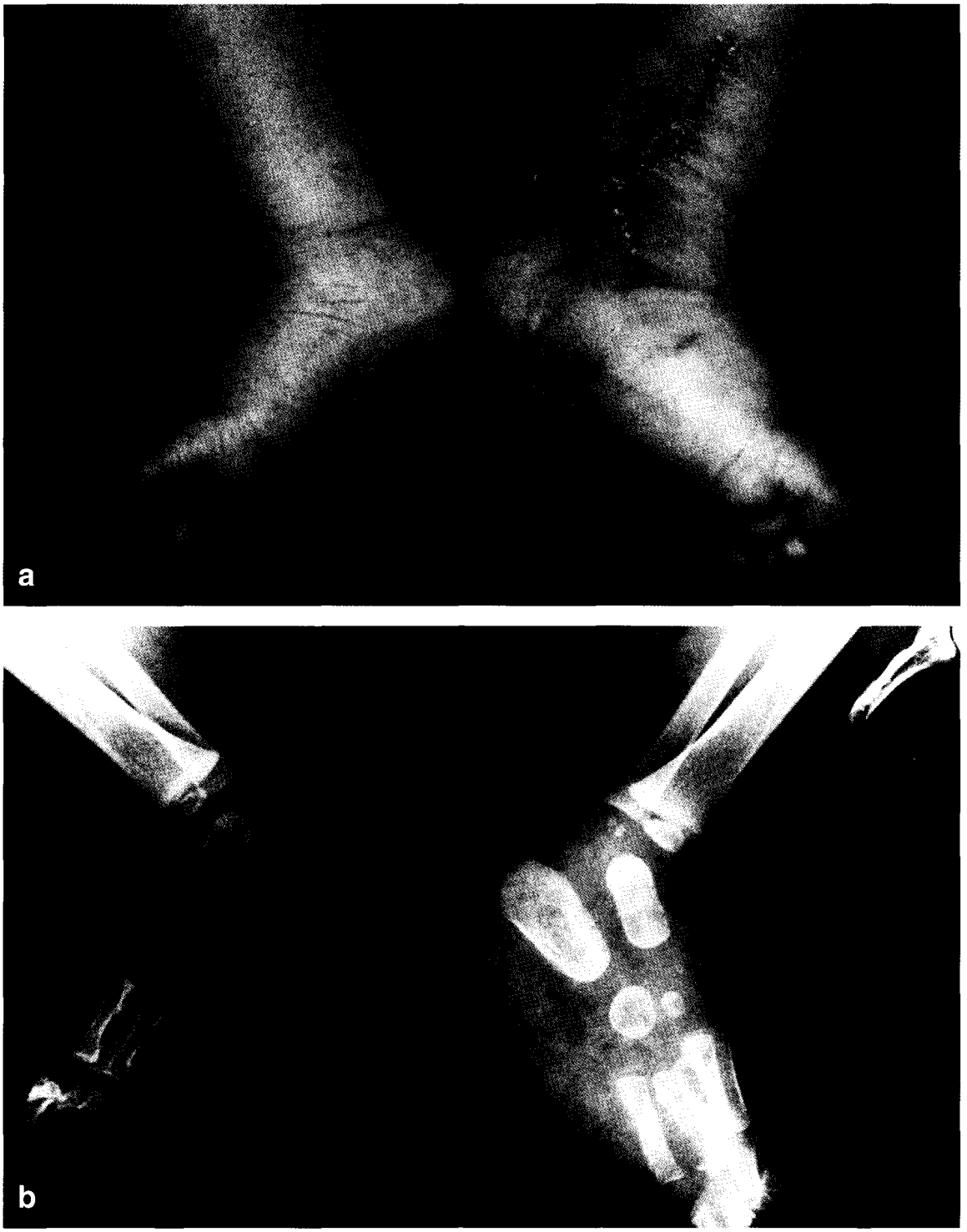

Fig. 3: (a) Lateral view of both feet after resection of tumor recurrence on the sole and maleolar areas. (b) Plain x-ray showing increase of growth of bones and soft tissue (hypertrophy) on the involved left side.

the context of lipodystrophy, in combination with limb hypertrophy, may be related to angiodysplasias (11-12). In this patient after reoperation, no direct or indirect signs of recurrence have thus far appeared.

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