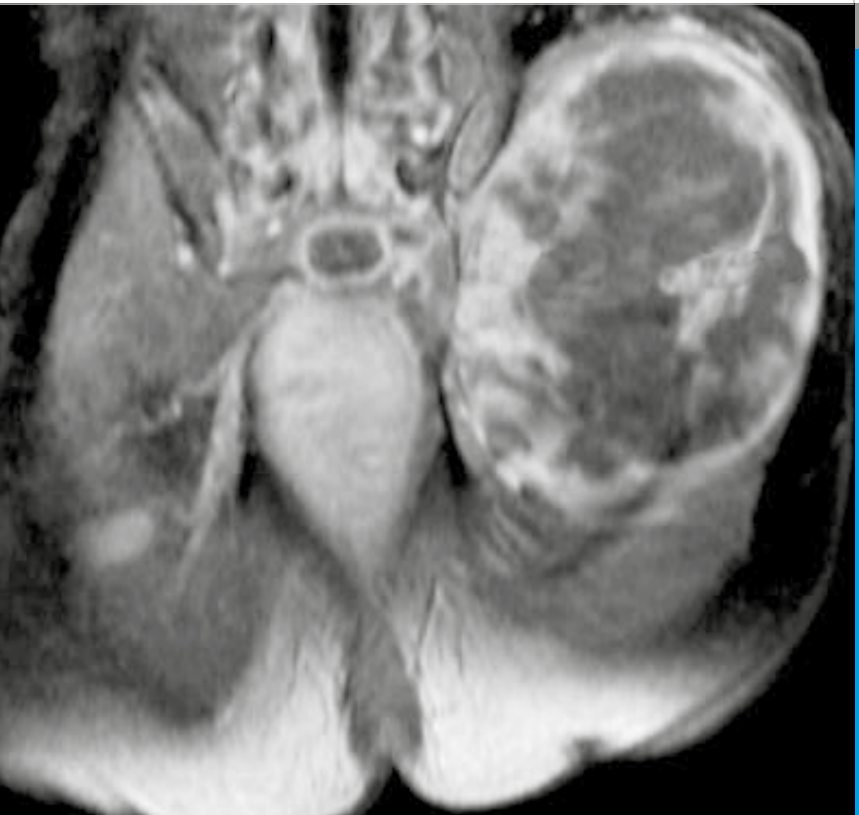
SWISS SOCIETY OF NEONATOLOGY

Congenital infantile myofibromatosis



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INTRODUCTION

Infantile myofibromatosis is a mesenchymal disorder of skin, muscle, bone, and/or viscera. We present a case of congenital multicentric myofibromatosis.

CASE REPORT

This full term female infant was born at 40 weeks of gestation by spontaneous vaginal delivery following an uncomplicated pregnancy. At birth, a firm, painless, purplish colored, subcutaneous tumor measuring 5x3x4 cm was noted over the left gluteal region. The remainder of her physical exam was unremarkable. Ultrasound examination revealed a soft tissue mass with liquid and solid parts (Fig. 1). There were no lytic bone lesions on conventional X-rays. MRI showed many tumors of different sizes in the muscles of the hip region on both sides. The largest mass was noted between the left Mm. gluteus maximus and medius (Fig. 2).

A biopsy of this lesion was obtained. Histopathology was consistent with infantile myofibromatosis of the subcutaneous tissue (central cellular hemangiopericytoma-like zone, surrounded by a nodular and fascicular peripheral zone with fewer fibroblastic and myofibroblastic cells) (Fig. 3). This was confirmed immunohistochemically (Fig. 4) with myofibroblasts being positive for alpha-SMA (smooth muscle actin), but negative for S-100 and cytokeratin.

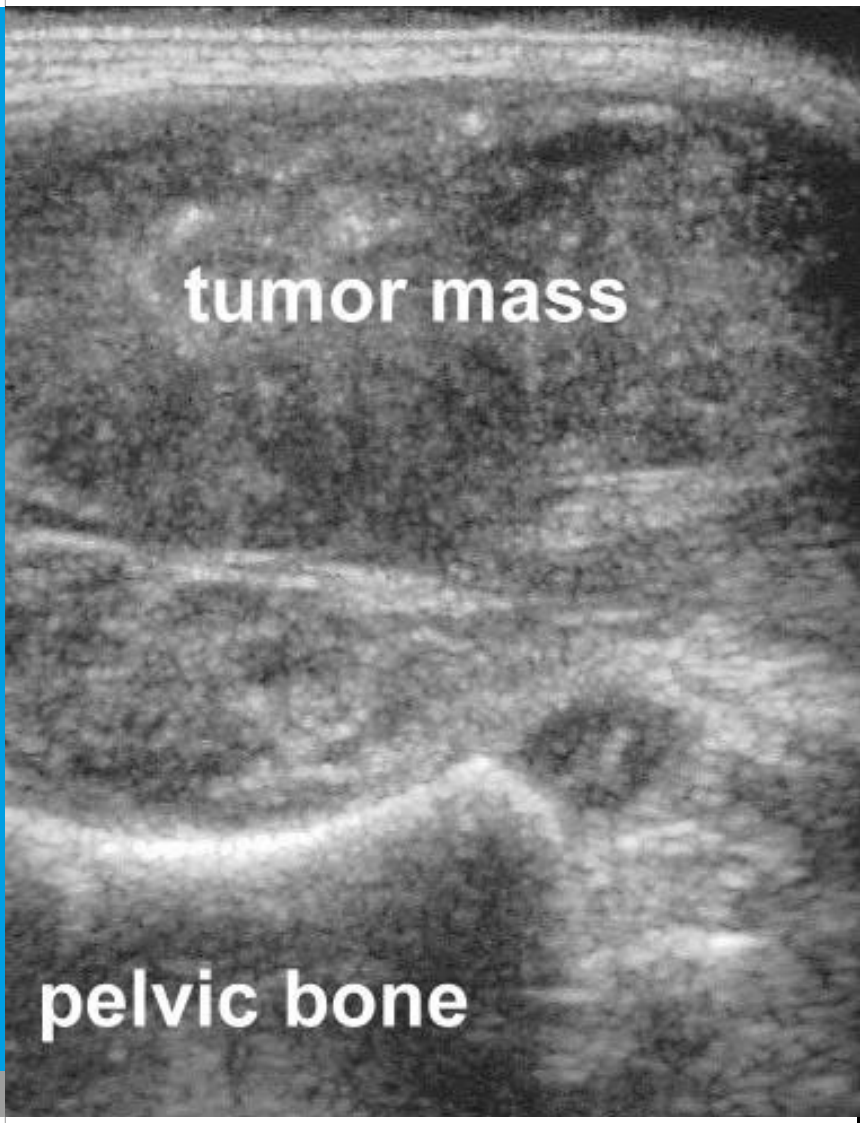

Fig. 1

Ultrasound: tumor mass with liquid and solid parts.

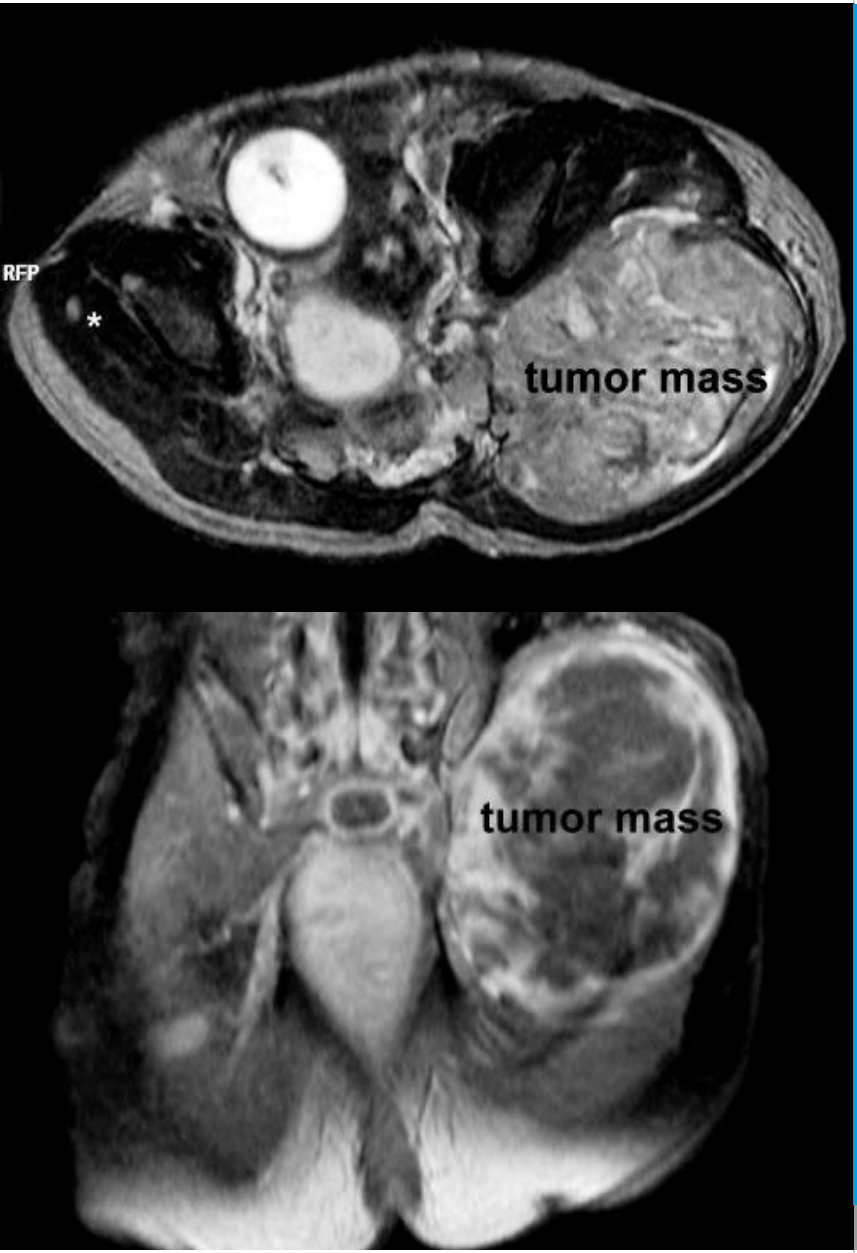

Fig. 2

MRI of the main tumor mass.

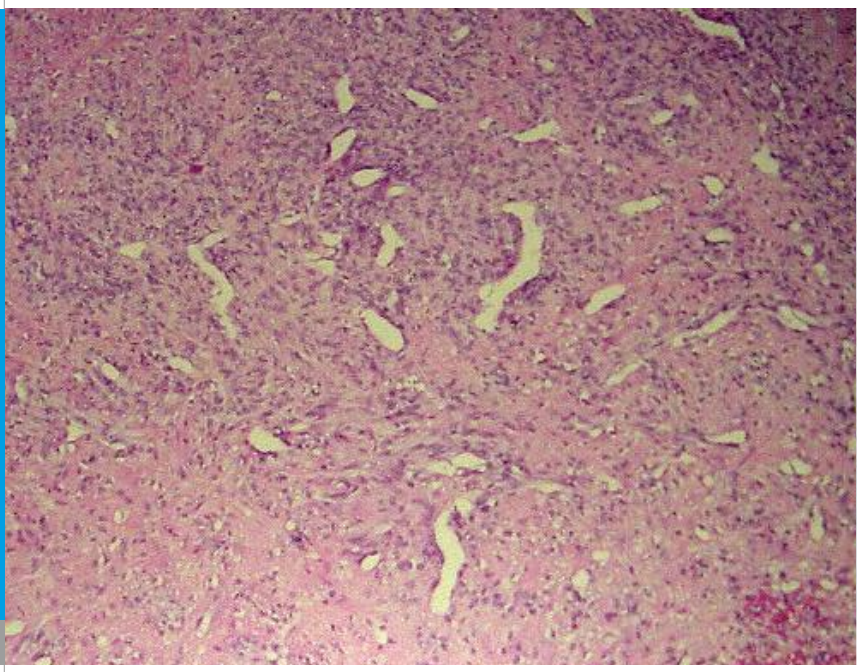

Fig. 3

Histopathology of the tumor (HE stain).

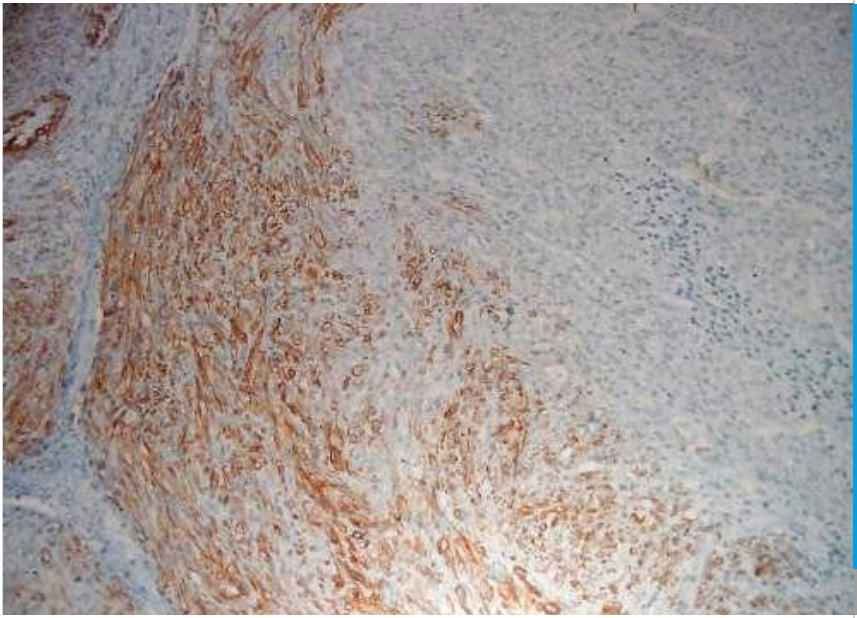

Fig. 4

Histopathology of the tumor (immunohistochemistry: alpha-SMA positive).

DISCUSSION

Infantile myofibromatosis (IM) was first described by Stout in 1954 (1). It represents the most common fibrous tumor in infancy (2). Fifty-four percent are present at birth and thus congenital, 89% are diagnosed during the first 2 years of life. IM is characterized by the appearance of firm, flesh-colored to purple nodules in skin, muscle, bone, and/or visceral tissue. There are three distinct types based on tumor location and tissue involvement (3):

A) solitary myofibromatosis
B) congenital multiple myofibromatosis with multicentric lesions but no visceral involvement
C) generalized myofibromatosis with visceral involvement.

Biopsy is necessary for final diagnosis. The prognosis is excellent in patients with a solitary lesion and multicentric lesions without visceral involvement with frequent spontaneous regression. Surgical excision is sometimes required for locally destructive or obstructive tumors. On the other hand, prognosis is poor in patients with involvement of visceral organs due to organ dysfunction, failure to thrive or infection (4).

REFERENCES

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