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Large Congenital Juvenile Xanthogranuloma in a Two-Month-Old Girl, a Case of Non-Langerhans Cell Histiocytosis

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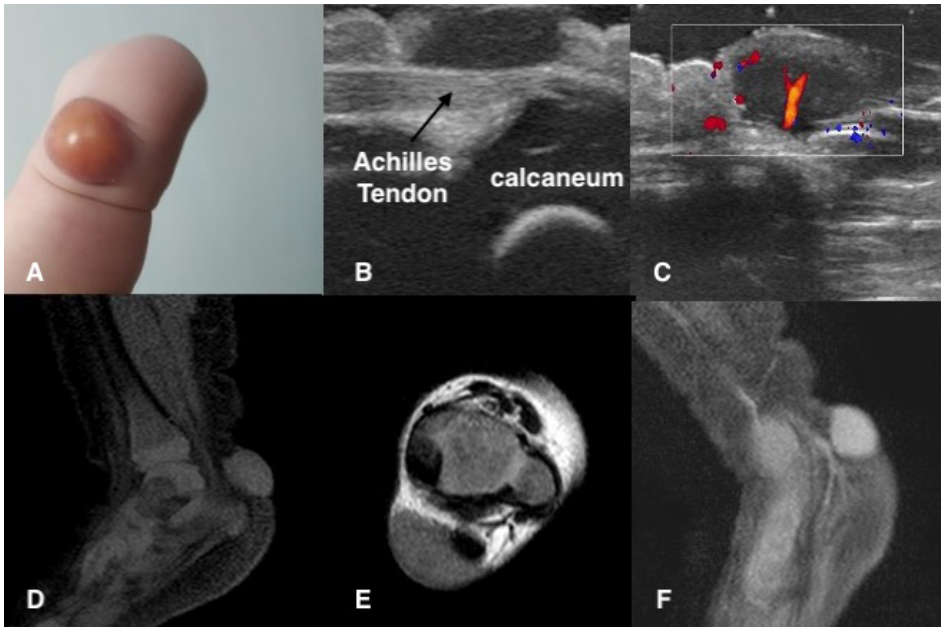
A two-month-old girl presented to the pediatric surgery department with an asymptomatic, solitary, supra-calcaneal mass, which had almost doubled in size since her birth. Upon clinical examination it appeared large, shiny, and orange-yellow, with well-defined erythematous borders (Panel A). Ultrasound (US) examination revealed a large (20 mm) hypoechoic well-circumscribed oval mass within the subcutaneous tissue, without disruption of the dermis or infiltration of the adjacent Achilles tendon. Minor vascular flow was detected within the mass by Color doppler evaluation (Panels B and C). MRI revealed a mass with an intermediate signal on T1-weighted images (Panel D), low signal on T2-weighted images (Panel E), and mild enchancement (Panel F).

Imaging features were atypical and thus the differential diagnosis was broad, including fibrous hamartoma of infancy, myofibroma, tendon-sheath fibroma, juvenile xanthogrannuloma and neurofibromatosis. Juvenile xanthogranuloma was diagnosed by histopathology. Juvenile xanthogranuloma (JXG) is a form of non-Langerhans cell histiocytosis (1) affecting children within the first two years of life. It can also be congenital. Its incidence is unknown, but it has a higher frequency in males (ratio: 1.4:1) (1, 2). Its exact pathophysiological mechanism



is poorly understood. JXG lesions may be cutaneous or extracutaneous with systemic involvement (1, 2). Involvement of the central nervous system with fatal outcome has also been reported (3). In our case, abdominal-US excluded systemic manifestations. Cutaneous lesions most frequently involve the head and neck, the trunk and, less frequently, the limbs (2). They are generally self-involuting and usually treatment is not necessary (1). The majority of patients undergo a diagnostic surgical excision, and no recurrent case is known in the literature (2). The common US appearance includes a well-defined hypoechoic solitary mass, without vascular flow. On MRI, JXG typically appears isointense to hyperintense on T1, and isointense to hypointense on T2-weighted images (4). Clinical and radiological features may vary, and biopsy may be needed (1). Pediatricians, pediatric surgeons and radiologists should be aware of this pathological entity, and be able to differentiate it from other soft-tissue tumors. Abdominal-US can detect possible systemic involvement, and a biopsy will confirm the diagnosis.

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