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PILOMATRIXOMA

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Background: A 6-year-old girl presented with a painful hard nodule located at the distal humerus.
The nodule was surrounded by redness of the skin. The mass had been detected 2 months prior to presentation and was growing slowly. There was no relevant clinical history.

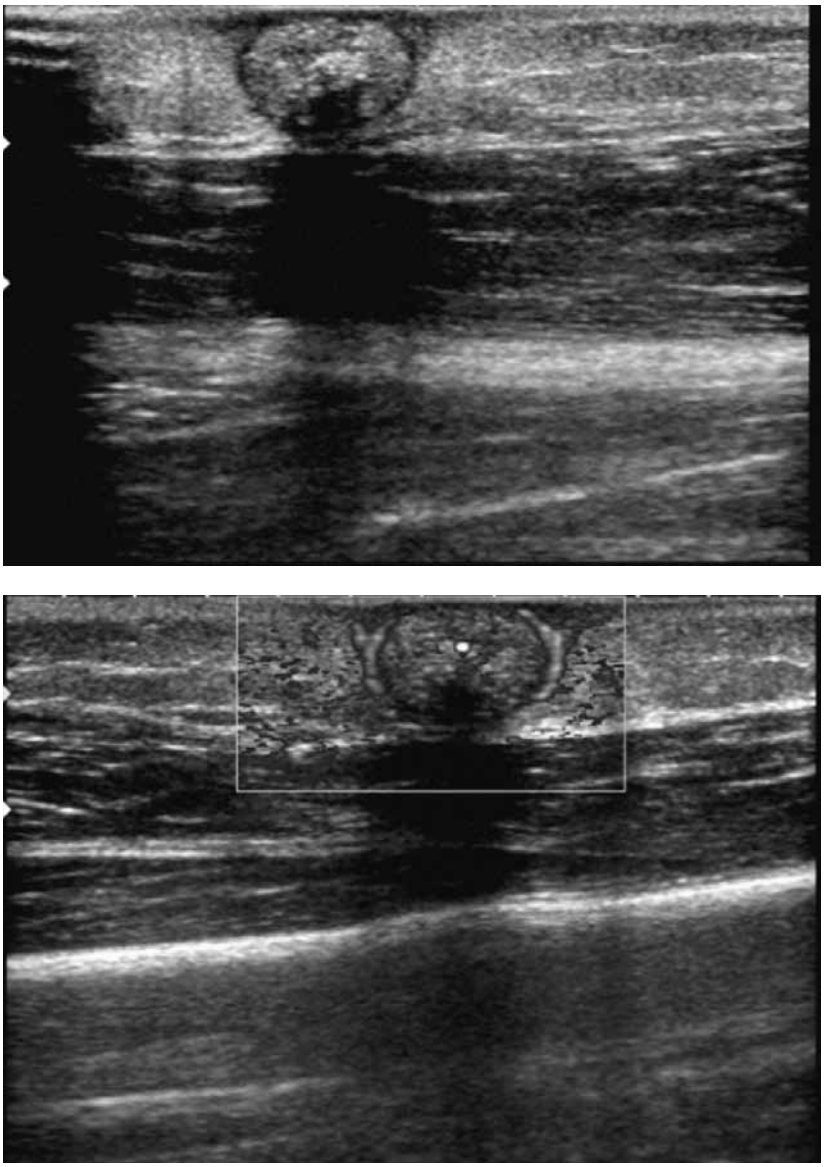

Fig. 1/2

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Work-up

Ultrasonography (2 D-image) (Fig. 1) demonstrates a well-circumscribed subcutaneous located lesion, overlying the triceps and the humerus. The lesion is isoechoic to subcutaneous fat and contains multiple small hyperechoic refl ections, which cause acoustic shadowing and thus represent calcifi cations. The mass is surrounded by a hypoechoic halo.

Ultrasonography (color-Doppler image) (Fig. 2) shows eminent vascular flow in the surrounding halo. Only moderate flow is seen in the center of the mass.

Radiological diagnosis

Pathological examination of the surgically excised mass revealed a pilomatricoma, also referred to as *pilomatrixoma or epithelioma of Malherbe*.

Discussion

Pilomatricomas are rare benign dermal neoplasms of the hair follicle with extension into the subcutaneous fat. Malherbe first described this lesion back in 1880 as a calcified epithelioma because it was thought to arise from sebaceous glands. With improving microscopic technology it became obvious that the origin of these masses was the outer root sheath of the hair follicle, hence the name 'pilomatrixoma'.

The most frequently involved locations of this benign lesion are the head and neck region, followed by the upper extremities, the trunk and lower extremities. These lesions can occur at any age, but most patients are younger than 20 years. Moreover, in children presenting with a superficially located tumor, it is the most frequent encountered diagnosis upon surgical excision. In most studies, females are affected up to twice as often as men.

Pilomatricoma frequently presents as a solitary calcifi ed lesion ranging from 0.5 to 3.0 cm in diameter; however, multiple lesions do occur, and these are associated with syndromes like Gardner and Steinert syndrome and with sarcoidosis and myotonic dystrophy.

The list of differential diagnoses is long and depends on the location of the lesion. If the lesion is located in the head and neck region, one can think of pre-auricular sinuses or branchial cleft remnants besides epidermal inclusion cysts, dermoid cysts, dystrophic calcifications, lipoma and pilomatrix carcinoma.

Pilomatrix carcinoma is a very rare malignant variant of pilomatricoma. It occurs mostly in middle-aged or elderly patients, and more often in men. Metastases have been reported but are also very rare.

Treatment of choice for both benign and malignant lesions is excision with clear margins.

Ultrasonography is the imaging modality of choice. It is a fast, non-invasive and accurate method for diagnosing pilomatricoma if most or all features are present. CT and MRI are occasionally used but less specific. MRI of a pilomatricoma has been described with intermediate signal on T1 and T2 and some enhancement, following administration of contrast medium.

Bibliography

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