We should support in the future the possibility that pilomatrixoma could be hereditary and present without evidence of another autosomal dominant disease.

An 8-year-old male was presented with a painless subcutaneous, palpable oval-shaped neck mass present for the last 3 months. No signs of inflammation were detected and sonography revealed a subcutaneous hypoechoic mass with internal calcifications. The mass was excised and sent for pathological examination, which revealed the typical sheets of “Ghost” cells along with signs of calcification and basaloid cells with small uniform nuclei and scant cytoplasm. The features were compatible with pilomatrixoma and there were no indications of malignancy. The patient is recurrence free at 6 months of follow up.

Results: Despite their typical calcification in sonography pilomatrixomas are not always easy to differentiate, especially in paediatric patients, from other head and neck more common masses such as dermoid cysts and other cystic lesions. Pilomatrix carcinomas are extremely rare. No accurate way to exclude malignancy without an histological examination.

A pilomatrixoma or calcifying epithelioma of Malherbe is an uncommon skin tumor derived from the hair matrix, typically occurring in the head and neck area and upper extremities and are more common in females. There are two peaks of occurrence with age. The first is before the 3rd decade and the 2nd during the 6th and 7th decades. Pilomatrixomas depending their location may be difficult to clinically differentiate from other subcutaneous masses, such as dermoid cysts or other cystic lesions.

TFI has a distinctive pattern that consists of interconnecting plates of monomer - phic keratinocytes arising as an extension from the basal epidermal layer. 48,76-78 small, abortive follicular structures may connect to the plate from below.

We should support in the future the possibility that pilomatrixoma could be hereditary and present without evidence of another autosomal dominant disease.