Multiple Pilomatricomas, Sternal Cleft and Mild Coagulative Defect

Sir,

Pilomatricomas are usually solitary nodules, but multiple occurrences have been observed in 2–4% of cases (1, 2). The association of multiple lesions with myotonic dystrophy is well recognized (3); familial cases have also been described in association with Gardner's syndrome and sarcoidosis (4, 5). We report here the case of a girl with multiple pilomatricomas associated with sternal cleft and mild polyfactorial coagulative defect.

CASE REPORT

A 9-year-old girl, with negative familial anamnesis, had a 4-year history of 4 nodular lesions 4–10 mm in diameter, localized on her thigh, buttocks and right cheek. The lesions had moderately enlarged over the last years and were asymptomatic except for the 1 on the cheek which periodically underwent ecchimotic halo after minimal trauma (Fig. 1). The lesions were freely movable and very hard on palpation. The tent sign was present only in the cheek lesion. After birth, the girl had undergone an operation for sternal cleft. Anamnesis also revealed a history of chronic pharyngitis and alopecia areata of the eyebrows. A general predisposition to ecchymosis after minimal trauma was present.

Laboratory examinations demonstrated an international normalized ratio (INR) of 1.33 (normal 0.85–1.20); an activated partial thromboplastin time (APT) of 37"9 (normal, 23" – 37"0); fibrinogen of 351 mg/dl (normal, 150–400); and antitrombin III 95% (normal 80–120%). Calcium–phosphorus metabolism was within limits. Ecography of the lesions revealed their calcific nature; histology demonstrated classic pilomatricomas. The patient was treated with vitamin K orally for 2 periods of 20 days. Abdominal ecography was normal. After 1 year, no new elements had appeared and the patient was in good general condition.

Fig. 1. Clinical appearance of pilomatricoma with ecchimotic halo on the cheek.

DISCUSSION

Pilomatricoma is a benign cutaneous tumour derived from the hair matrix. Multiple tumours are rare; there is evidence to suggest that patients with multiple pilomatricomas, presenting in a familial pattern, have a high probability of myotonic dystrophy. This association most likely represents a further pleiotropic effect of myotonic dystrophy gene. Multiple lesions seem to have no special clinical or histological features that permit their differentiation from isolated lesions (6).

Cleft sternum is a rare congenital deformity due to partial or total failure of sternal fusion at an early stage of embryonic development. Sternal clefts can be classified as superior, inferior, or complete. (7). Sternal clefting is known to be associated with vascular dysplasia, such as cervicofacial hemangiomata, ectopia cordis, visceral malformations, sacral and genitourinary defects, palatoschisis (8, 9). Familial cases have been reported.

The polyfactorial coagulative defect of our patient was quite mild, but we believe that it may be taken into account in the syndrome.

To the best of our knowledge, the association of multiple pilomatricomas with sternal cleft and polyfactorial coagulative defect has never been reported. The literature teaches us that the association of very rare conditions in the same patient may not be casual. We therefore propose this association as a new entity, whose pathogenetic mechanism is obscure.

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