Pilomatrix Carcinoma in a Child

Sir,

An 8-year-old boy presented with a single, well-defined, asymptomatic, erythematous, firm, fleshy, lobulated tumour, measuring $80 \times 60 \times 40$ mm, over the right pre-auricular region. The tumour was of 2 months duration. The surface was eroded in many places (Fig. 1). The lesion had appeared spontaneously as a small red papule, which grew rapidly. Its surface ulcerated, occasionally discharging chalky white granules and purulent material. There were no constitutional symptoms. He had been treated with topical and oral antibiotics with no improvement. There was no history of any surgical intervention. General physical and systemic examinations were within normal limits. There was no regional lymphadenopathy. Clinical diagnoses of mycetoma and aggressive appendageal tumour were considered. Results of a haemogram, liver and renal function tests, chest X-ray and urinalyses were normal. X-ray of the skull revealed a soft tissue swelling over the right mandibular region with no gross evidence of calcification or underlying bone involvement. Gram's stain and KOH preparation of the purulent discharge and the granules did not reveal any microorganism. Culture of the pus and chalky granules were negative for bacteria and fungi.

A scalpel biopsy from the edge of the lesion showed features of pilomatricoma. Wide excision of the mass, followed by split thickness skin grafting was carried out under general anaesthetic. During surgery no gross sign of invasion into the surrounding structures was found. Histopathological examination of the excised tumour revealed irregular islands of epithelial cells in a cellular stroma and multiple cystic spaces containing abundant necrotic debris, focal calcifications and amorphous material. The shadow cells were situated within the cystic spaces. Gradual transition of basaloid to shadow cells was seen in most places; however, the transition was abrupt in some areas. In places, the shadow cells also showed keratinization. The basophilic cells formed sheets and islands and invaded the reticular dermis and subcutis. These cells were uniform in appearance; however, in some places the cells showed crowding with loss of polarity, moderate pleomorphism, and high nucleocytoplasmic ratio, hyperchromasia, prominent nucleoli and high mitotic ratio. In view of the large size of the tumour, its rapid growth, ulceration and the microscopic findings above, a final diagnosis of pilomatrix carcinoma was made. The patient is under follow-up for 1 year and has not developed any recurrence or metastasis.

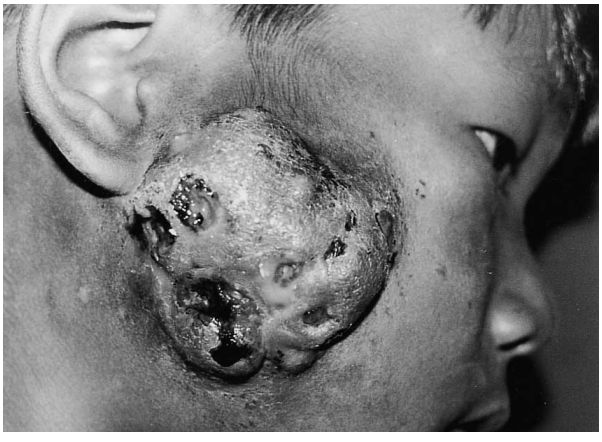

Fig. 1. A well-circumscribed lobulated and ulcerated tumour over the right pre-auricular region.

DISCUSSION

The currently accepted term for the condition described here is pilomatricoma. It is the most common hair-follicle tumour, and is often seen in children, presenting as an asymptomatic, solitary, firm, deep dermal or subcutaneous nodule, 5–30 mm in diameter, on the head, neck or upper extremities.

Malignant variants of pilomatricoma have been reviewed (1, 2). A review of 20 cases by Sau et al. (1) in 1993 revealed that pilomatrix carcinoma presents as a solitary, asymptomatic, dermal or subcutaneous mass with a predilection for the posterior neck, back and pre-auricular area. The scalp was the most common site in 14 patients, followed by the face in 6, neck in 3, arm, back, and buttocks in 2 cases each, and the inguinal area in 1 patient, among 30 cases of malignant pilomatricoma reviewed by Tateyma et al. (2) in 1992. The tumours varied in size from 10 to 100 mm (mean 46 mm) in diameter. The largest tumour reported so far was $200 \times 150 \times 80$ mm in size (3). Contrary to its benign counterpart, pilomatrix carcinoma has no predilection for people under 20 years of age. Sau et al. (1) found that it is more common in men, with a male : female ratio of 4 : 1 and a mean age of 45 years, whereas Tateyma et al. (2) reported an almost equal male : female ratio and a median age of 57 years. Pilomatrix carcinoma is a locally invasive tumour with a tendency to recur, especially if it is inadequately excised. Sau et al. (1) reported local recurrences in 59% of 17 patients on follow-up. The tumour rarely metastasizes. To date, only 4 cases of pilomatrix carcinoma with visceral metastases have been reported (4).

The patient described here had many unusual features, such as young age of onset (8 years), rapid progression over a period of 2 months, perforation and elimination of chalky white granules from the surface. To date, only 2 cases have been reported in patients under 20 years of age (5). The youngest case reported was 4 years old (5). Our patient's tumour was probably the fastest growing pilomatrix carcinoma, as it attained its presenting size of $80 \times 60 \times 40$ mm within 2 months and ulcerated due to rapid growth. Features of malignancy were revealed only after complete excision of the tumour and were missed on scalpel biopsy.

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Arun Joshi[1], Shatrughan P. Sah[2], Chandra Shekhar Agrawal[3], Mary Jacob[1] and Arun Agarwalla[1]
Departments of [1]Dermatology and Venereology, [2]Pathology and [3]Surgery, BP Koirala Institute of Health Sciences, Dharan, Nepal.

Topical Calcipotriol in Childhood Psoriasis

Sir,
Between January 1994 and March 1997, 20 children (13 males and 7 females) ranging in age from 2 to 13 years affected by mild, stable psoriasis were treated with calcipotriol ointment 50 μg/g (Psorcutan pomata, Schering).

Exclusion criteria included children with acute guttate psoriasis and with, or with past history of erythrodermic or pustular psoriasis. None of the children had any systemic treatment in the 2-week period before or during the trial and none of them had applied or were applying topical steroids in these two periods. In all patients, the area of skin treated did not exceed 10% of the total body surface area. We did not use calcipotriol ointment on the face, scalp, genitalia, nails or hands and feet, even though in six patients one or two of these areas was also affected.

The treated lesions were one or more erythemato-squamous nummular patches, 20–50 mm in diameter, in 15 patients, round guttate patches, less than 10 mm in diameter, in 4 patients and inversus psoriasis with a single patch on the right axillary fold in 1 patient. The lesions were located on the limbs (12 cases), the trunk (8 cases) or both (3 cases).

After receiving informed consent from the children's parents we proposed medication with calcipotriol ointment once a day (at night) without occlusion for the first week of treatment. In the morning a urea-based cream or a simple emollient was applied on the basis of scaliness value. If no irritant cutaneous reactions, such as redness or burning, were observed at the end of the first week we suggested twice daily applications of calcipotriol ointment for 8 weeks or until complete remission. Patients were examined every 2 weeks during the trial period.

Clinical assessment used a score system that considered the morphology and number of the lesions: in every patient we chose the most typical or the only patch of psoriasis and from this we measured the intensity of erythema, thickness and scaliness. Moreover, at the start and at every 2-weekly consultation we counted the exact number of patches. We considered results as: no change (25% improvement in number and morphology of lesions); slight improvement (25–50%); marked improvement (50–70%); or clearance (over 75%). In this manner, the value of severity at baseline was arbitrarily considered to be 100% in every patient and at each consultation any improvement was evaluated relatively for each case. No other available score was found because the PASI score at baseline was already too low to show any improvement at subsequent consultations.

RESULTS

After 8 weeks we observed a marked improvement in nine cases. Clearance was noted in six patients after 3–5 weeks of the therapy. No change was seen in two, and three withdrew because of local irritation. The maximum amount of ointment used during the 8-week trial never exceeded a single tube (30 g).

CONCLUSION

Our trial was limited to minimal childhood psoriasis, which shows remissions and relapses characterized by only one or few patches confined to a single site of the body and involving less than 10% of the cutaneous surface (1). In our opinion calcipotriol ointment is effective and safe in the treatment of such patients and it is a good alternative to the use of topical steroids. Similar results have been reported in the literature in other studies on children (2–4).

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Annalisa Patrizi, Iria Neri, Laura Rizzoli and Claudio Varotti
Department of Clinical and Experimental Medicine, Division of Dermatology, University of Bologna, Via Massarenti 1, IT-40138 Bologna, Italy.