Perforating Pilomatricoma Causing a Cutaneous Horn

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Pilomatricoma is a benign tumor differentiating towards the hair matrix. Perforation may rarely take place with extrusion of part of the contents.

A 3-year-old girl presented with a 5 × 6 mm sized, erythematous, elevated and indurated mass with a tiny horny projection on its surface, on her right upper arm. It had been present for one year. Histopathologically, there were several islands of epithelial cells in the upper to mid-dermis composed of basophilic, shadow and transitional cells. A few tumor islands adjacent to the epidermis were in the course of a characteristic transepithelial elimination process.

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Key Words : Horny projection, Perforation, Pilomatricoma

Pilomatricoma, calcifying epithelioma of Malherbe, is a benign tumor that tends to differentiate towards the matrix of the hair follicle. The lesion is usually a solitary, firm nodule and is found particularly on the head, neck and upper extremities. Histologically, the tumor is located in the reticular dermis and extends into subcutaneous fat. Perforation may rarely take place with extrusion of part of the contents[1].

Cutaneous horn is the clinical term for a hard, yellowish-brown conical hyperkeratotic excrescence which may be associated with many different pathological lesions including actinic keratosis, verruca, seborrheic keratosis and squamous cell carcinoma. On rare occasions, a trichilemmoma or a basal cell carcinoma is seen[1].

We present a rare case of perforating pilomatricoma causing a cutaneous horn.

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CASE REPORT

A 3-year-old girl presented with a cutaneous mass of one year duration on her right upper arm. It began as a tiny papule and became a firm, erythematous mass with a cutaneous horn. The mass had persisted for 1 year with no systemic signs or symptoms. Laboratory findings showed normal findings.

A physical examination disclosed a 4 to 5 mm round, erythematous, elevated and indurated mass with a tiny horny projection on its surface(Fig. 1). She had no symptoms and no history of trauma.

Histologically, the epidermis overlying the tumor mass showed hyperkeratosis, parakeratosis, hypergranulosis and acanthosis. Islands of epithelial cells were noted in the upper dermis(Fig. 2). They were composed of two basic cell types, basophilic and eosinophilic shadow cells. Basophilic cells lay close together, having a large nucleus showing several mitotic figures and scanty cytoplasm. Shadow cells of a pale-pink color had lost their nucleus and retained their cell membrane(Fig. 3). The basophilic and shadow cell components were detached from one another but we could also observe a zone of transitional cells. The stroma showed mild inflammatory infiltration. Foreign body reactions were absent. A few tumor islands adjacent to the epidermis were in the course of an elimination process through characteristic hyperplastic follicular

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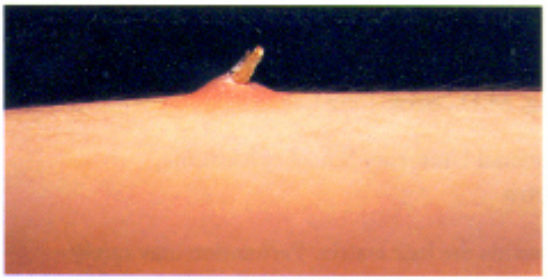

Fig. 1. A 5×6 mm sized, erythematous, elevated and indurated mass with a tiny horny projection on its surface.

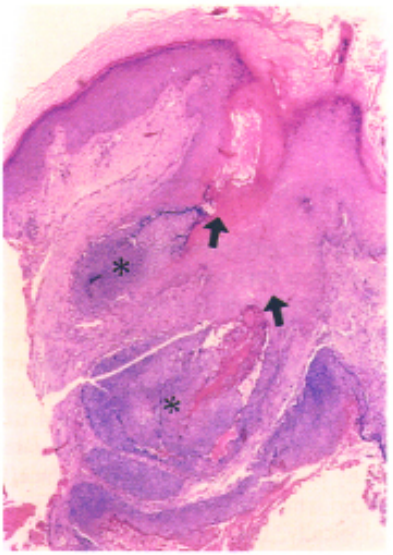

Fig. 3. Island of pale-pinkish shadow cells, which had lost their nucleus but retained their cell membrane (H&E, ×100).

epithelium-like channels(Fig. 4). Calcium deposits were not detected with the von Kossa stain in the tumor nests.

The diagnosis of perforating pilomatricoma manifesting as a cutaneous horn was made. No recurrence of the lesion was noted for 9 months after excision.

DISCUSSION

Pilomatricoma is a firm dermal tumor, usually showing an essentially normal overlying epidermis that tends to differentiate towards hair cortex

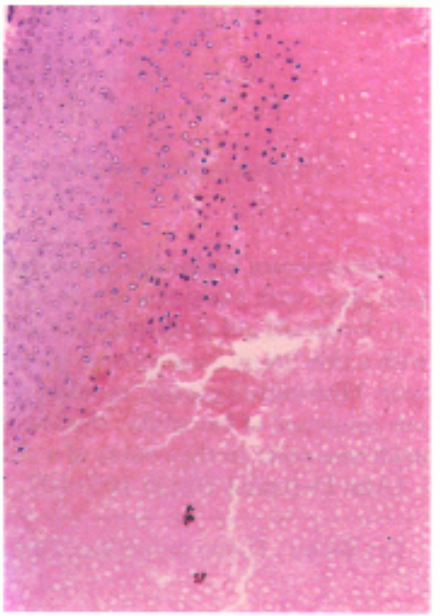

Fig. 2. Cell nests of pilomatricoma(*) in the course of removal through hyperplastic follicular epithelium-like channels(→) (H&E, ×40).

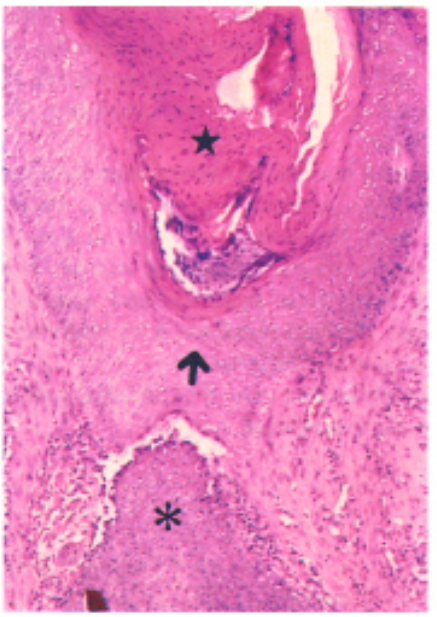

Fig. 4. A basophilic cell nest(*) is eliminated through a characteristic channel(→). A funnel shaped invagination of the epidermis is filled with parakeratotic cells(★) which are difficult to distinguished from transitional cells. (H&E, ×200).

cells[1,2]. It was first described in 1880 by Malherbe and

Chenantais[1] as a calcified epithelioma of the sebaceous glands. In 1942 Turhan and Krainer[1] reported that tumor cells differentiate towards hair cortex cells. They are located most commonly on the head, neck or on the upper extremities[1,2]. The tumor is frequently seen in children, about 40% of it occurring by the age of 10, 60% before the age of 20[1]. It is not hereditary but a few instances of familial cases and an association with myotonic dystrophy have been reported[1,2]. Malignant changes were reported in several cases, which appeared chiefly in large, old pilomatricomas[2]. The size varies from 0.5 to 5 cm in diameter[1]. Our patient had no signs of myotonic dystrophy, and no family history of similar lesions.

Perforating pilomatricoma is a rare clinical entity and, to our knowledge, eight cases have been reported in the English literature[3-10]. Clinically, the size, age and location of perforating pilomatricoma are similar to those of the classic form. However, prior case reports of the former showed the following features: a relatively short time to reach the diagnosis[10], a reddish to blackish brown exophytic cutaneous mass with surface alterations, such as keratinous cores[4], black dots[6], or areas of squamous crusting[9]. In 1986 Uchiyama et al.[7] reported a case of perforating pilomatricoma, showing a 6× 5× 5 mm sized, firm, blackish-brown, cutaneous horn-like appearance on the left upper arm. Our case was a similar, but was unique in that a sharp spike-like keratotic mass projected from the tumor surface.

Histologically, pilomatricoma is usually located in the reticular dermis and extends into subcutaneous fat[1]. In perforating forms, the tumor is situated in the relatively upper portion of the dermis, which suggests that a superficial location may be one of the causative factors in transepithelial elimination[5,8,9]. As shown above, the tumor mass in our case was also situated in the upper and mid-dermis. Transepithelial elimination is usually regarded as a process of extruding material from the dermis through the epidermis without causing ulceration. In this phenomenon, the transeliminating component acts as a mechanical irritant that eventually causes hyperplasia of the epidermis and the follicular epithelium. The epithelial hyperplasia encloses the pathologic tissue, which is gradually brought towards the surface and is finally eliminated with the keratinocytes[11]. In our case, transepithelial elimination was associated with epithelial hyperplasia but, there were also the other reported cases in which epithelial hyperplasia was absent[3,10].

Calcification is common in pilomatricoma, occurring in 75% of all cases[1], but the von Kossa stain was negative in the present case. Even areas of ossification are also seen in 15% to 20% of cases[1].

We report this case because the patient presented with a rare perforating pilomatricoma that showed a prominent cutaneous horn.

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