

- 7 Kenner BM, Rosen T. Cutaneous amebiasis in a child and review of the literature. *Pediatr Dermatol* 2006; 23: 231–234.
- 8 Loschiavo F, Guarneri B, Ventura-Spagnolo T, *et al*. Cutaneous amebiasis in a Iranian immunodeficient alcoholic: immunochemical and histological study. *Dermatology* 1997; 194: 370–371.
- 9 Bumb Ra, Mehta RD. Amoebiasis cutis in HIV positive patient. *Indian J Dermatol Venereol Leprol* 2006; 72: 224–226.
- 10 High WA, Bravo FG. Emerging diseases in tropical dermatology. *Adv Dermatol* 2007; 23: 335–350.
- 11 Pritzker AS, Kim BK, Agrawal D, *et al*. Fatal granulomatous amebic encephalitis caused by *Balamuthia mandrillaris* presenting as a skin lesion. *J Am Acad Dermatol* 2004; 50(Suppl. 2): 38–41.
- 12 Valverde J, Arrese JE, Pierard GE. Granulomatous cutaneous centofacial and meningocerebral amebiasis. *Am J Clin Dermatol* 2006; 7: 267–269.

Familial occurrence of calcifying epithelioma of Malherbe

Pilomatrixoma, also known as calcifying epithelioma of Malherbe, is a benign skin neoplasm of hair follicle origin. The greatest incidence is found in patients between 8 and 13 years of age.¹ The majority occur in the head and neck region (52%), followed by limbs (32%) and trunk (10%).^{1,2} No cases have been reported on the palms, soles, or genital region.^{1,3} There may be a history of previous trauma to the tumor site.⁴ There appears to be a 3:2 female-to-male incidence ratio, and Caucasians are primarily affected.^{1,5} However, familial incidence is very rare.^{1,5} We present a rare familial occurrence of pilomatrixoma in two siblings.

A 9-year-old girl presented with two painless, firm, slow-growing nodules of 1 year's duration on her left forearm and right arm. The lesions were between 1 and 1.4 cm in greatest diameter. Palpation showed two freely mobile subcutaneous tumors. Her brother, 8 years old, also presented with a painful nodule of two years' duration on his right cheek. Examination revealed a firm, 1.4 cm mass located in the right preauricular region. In both cases, the overlying skin was normal in appearance without evidence of ulceration or discoloration. There was no history of previous trauma to the areas. No other skin abnormalities were noted. Ultrasonography was requested in the second case where the lesion was located in the parotid region.

Total surgical excision of the tumors with clear margins was performed under local anesthesia, with a small skin incision and blunt and sharp dissection. The tumors were not attached to the surrounding tissues and were easily removed (Fig. 1). Histologic examination of both specimens showed typical features of pilomatrixoma (Fig. 2). The patients were examined by a Pediatric Neurologist who did not find any syndrome, anomaly, or other genetic disorder. Almost one and a half years postoperation, the boy remains without evidence of recurrence, but a new pilomatrixoma has been removed from the girl.

Pilomatrixoma is the most common superficial tumor in children; however, familial incidence is very rare.^{1,5} In the largest series of 1569 studied cases of pilomatrixoma,¹ only five showed familial occurrence. Hawkins and



Figure 1 One of the two pilomatrixomas removed from the right arm of the 9-year-old girl

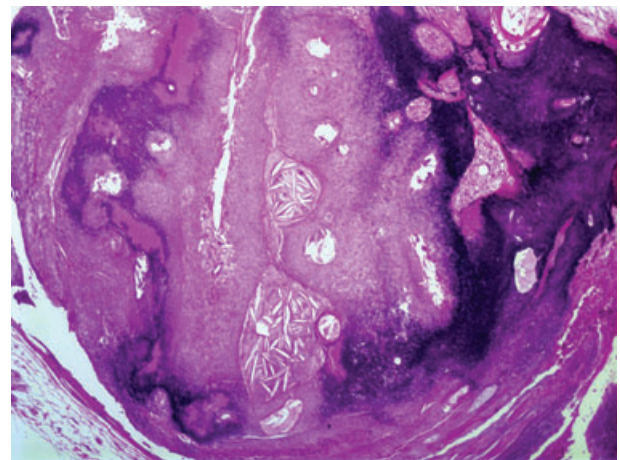


Figure 2 Pilomatrixoma with the presence of basaloid cells at the periphery and ghost cells in the center of the tumor islands (H+EX 4)

Chen⁵ in 1984 reported one more case and Demircan and Balic⁶ in 1997 found 2 of 15 studied cases with familial occurrence.

Pilomatrixomas are generally solitary tumors, but multiple tumors have been reported to occur in 2–3% of cases.^{1,2,6–8} Multiple tumors may be found in association with Gardner syndrome, myotonic dystrophy, xeroderma pigmentosum, or Turner's syndrome.^{7–10} Patients with myotonic dystrophy are more likely to have multiple tumors with familial occurrence. Some authors consider pilomatrixoma to be a cutaneous marker of myotonic dystrophy (Steinert's disease).⁸ Our patient had three tumors with no evidence of myotonic dystrophy.

Pilomatrixoma typically presents as a firm, slow-growing, subcutaneous mass that is well circumscribed with a general size <3 cm in diameter.⁷ However, there have been reports of giant pilomatrixomas. The diagnosis can usually be easily made on the clinical findings, which are confirmed by histopathologic examination.⁴ Radiologic imaging is of little diagnostic value apart from ultrasonography that demonstrates the superficial position, the continuity of the lesion with deeper structures and the degree of calcification.⁴ It is a fast, low-cost, noninvasive and more appropriate method of diagnosis for younger children.⁴

As spontaneous regression is never observed and malignant degeneration is rare, the complete surgical excision with clear margins is the treatment of choice,^{2,4,7} whereas for the extremely infrequent malignant variation, wide-field excision with margins of 1–2 cm is recommended.⁷

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References

- 1 Moehlenbeck FW. Pilomatrixoma (calcifying epithelioma): a statistical study. *Arch Dermatol* 1973; 108: 532.
- 2 Rotenberg M, Laccourreye O, Cauchois R, *et al.* Head and neck pilomatrixoma. *Am J Otolaryngol* 1996; 17: 133–135.
- 3 Knight PJ, Reiner CB. Superficial lumps in children: what, when and why? *Pediatrics* 1983; 72: 147–153.
- 4 Pirouzmanesh A, Reinisch JF, Gonzalez-Gomez I, *et al.* Pilomatrixoma: a review of 346 cases. *Plast Reconstr Surg* 2003; 112: 1784–1789.
- 5 Hawkins DB, Chen WT. Pilomatrixoma of the head and neck in children. *Inter J Pediatr Otorhinolaryngol* 1985; 8: 215–223.
- 6 Demircan M, Balic E. Pilomatrixoma in children: a prospective study. *Pediatr Dermatol* 1997; 14: 430–432.
- 7 Strobl H, Emshoff R. Pilomatrixoma of the cheek: report of case. *J Oral Maxillofac Surg* 1995; 53: 1355–1357.
- 8 Geh JLC, Moss ALH. Multiple pilomatrixomata and myotonic dystrophy: a familial association. *Br J Plast Surg* 1999; 52: 143–145.
- 9 Meenal PR, Vishalakshi V, Manoj A, *et al.* Pilomatrixoma in a case of familial xeroderma pigmentosum. *Indian J Dermatol Venereol Leprol* 2007; 73: 198–199.
- 10 Pielop JA, Metry D. Multiple pilomatrixomas in association with spina bifida. *Pediatr Dermatol* 2005; 22: 178–179.