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Characterizing pilomatricomas in children: a single institution experience☆

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Abstract

Background/Purpose: Pilomatricomas, or calcifying epitheliomas of Malherbe, are among the most common superficial cutaneous soft tissue lesions in children. Familiarity with the presenting signs and symptoms allows for the diagnosis to be made on physical examination alone in most patients, avoiding expensive and unnecessary diagnostic imaging.

Methods: A retrospective IRB-approved review of surgical pathology archives and medical records of all patients undergoing excision of pilomatricomas between 1982 and 2010 was performed to determine the characteristics of the pilomatricoma tumors. Data regarding gender, age, location, size of tumor, and histopathology were collected.

Results: There were 916 pilomatricomas resected in 802 patients. Fifty-five percent of the patients were girls (441 patients). The median age at the time of resection was 6 years (range 5 months to 18 years). Multiple lesions were found in 43 patients (5%). The most common location was head and neck ($n = 529$, 58%), followed by upper limbs ($n = 214$, 23%), trunk ($n = 130$, 14%), and lower limbs ($n = 43$, 5%). Information on size was available for 674 lesions; mean lesion diameter was 14.0 ± 7.4 mm. Twenty-eight patients (3%) had either recurrent ($n = 11$) or metachronous ($n = 17$) lesions resected at our institution, with a median interval of 12 months after initial resection (range 5 weeks to 5 years). No cases of pilomatrix carcinoma were observed.

Conclusion: The majority of pilomatricomas occur in the head and neck, although they can present in any location. Approximately 5% of children have multiple lesions. Pilomatricomas occur slightly more commonly in girls, and 66% of lesions occur in children < 10 years of age. Complete surgical excision is necessary to prevent recurrence. Recurrences and pilomatrix carcinoma are very rare if complete excision is achieved.

Pilomatricomas, otherwise known as "calcifying epitheliomas of Malherbe" were first described by Malherbe and Chenantais in 1880 as a skin tumor arising from sebaceous glands [1-3]. Dubreuilh and Cazenave first described the

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histopathologic features of basaloid epithelial cells islands and shadow (ghost) cells, and Turhan and Krainer discovered the origin of the tumor from hair cortex cells [1]. Pilomatricomas are benign skin neoplasms of childhood that present as deep, subcutaneous lesions lying between the dermis and the hypodermis (Fig. 1) [2]. They most commonly occur in the head and neck region, but have been reported in other areas of the body as well [1,4]. On excision these lesions are solid with a gray-tan appearance, and can appear to be in direct continuity with the overlying epidermis (Fig. 2) [5]. Pilomatricomas usually occur as isolated lesions, but have been reported to occur in association with polyposis–colorectal carcinoma predisposition syndromes (Gardner syndrome, Familial adenomatosis polyposis syndrome [FAP, APC gene, MYH (MUTYH) gene], and β-catenin gene mutations) and myotonic muscular dystrophy (Steinert disease) [2,6-12]. Less commonly, these tumors can occur in association with sarcoidosis, skull dysostosis, Rubinstein–Taybi syndrome, Churg–Strauss syndrome, Turner syndrome, Soto syndrome, fronto-parietal baldness, gliomatosis cerebri and trisomy 9 [1,6,13-16].

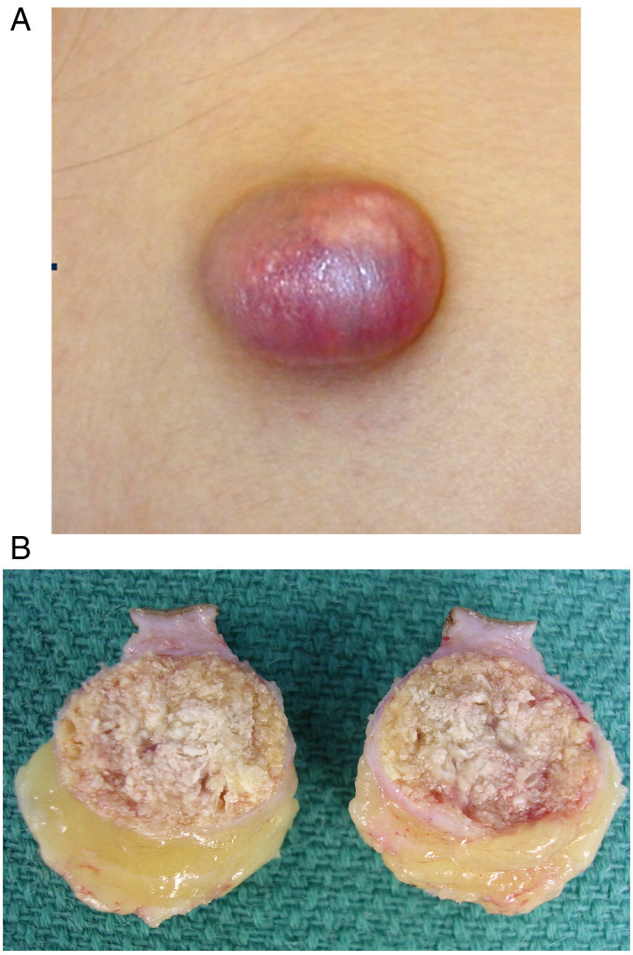

Fig. 1 Pilomatrixoma located on posterior neck (A) is well-circumscribed, present below the epidermis, and has a purple-blue hue with an intact overlying epidermis. Gross examination (B) of the excised pilomatrixoma reveals the close proximity of the tumor to the epidermis and involvement the entire dermis and extending into the subcutis. Note the irregular variegated and granular nature of the tumor's cut surface.

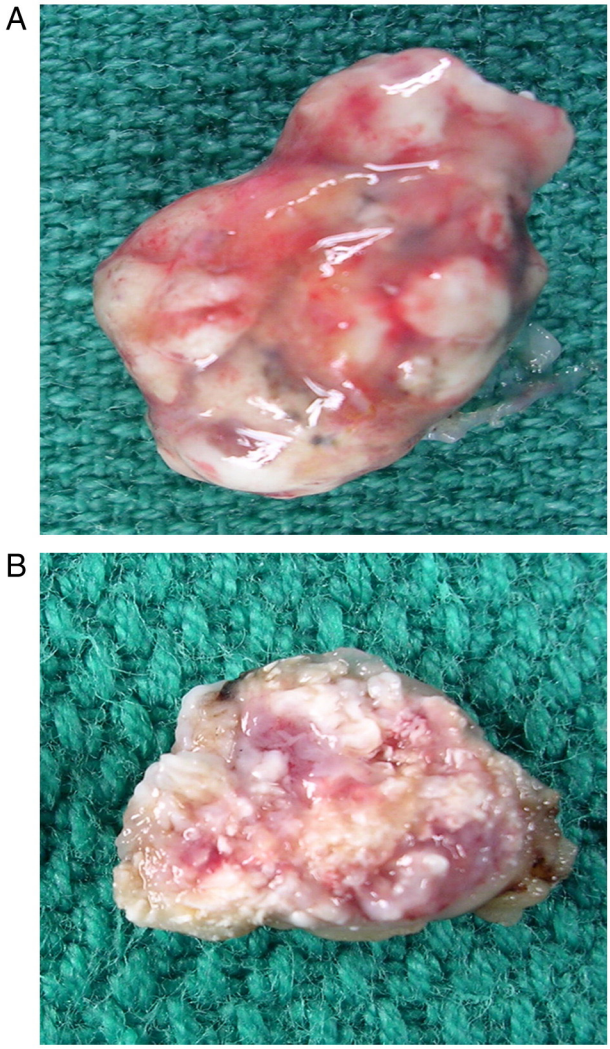

Fig. 2 Pilomatrixoma: gross appearance of lesion that was "shelled out," demonstrating irregular surface architecture (A), a thin membrane encasing the irregularly-shaped mass, and heterogenous nature of the lesional contents on cut surface (B). The irregular surface and lack of complete excision of the mass lead to local recurrence of the lesion in many cases.

After epidermoid cysts, pilomatricomas are the second most frequently excised pediatric skin mass, accounting for approximately 10% of all superficial masses evaluated by pathologists [5]. They may be misdiagnosed clinically as cysts (dermoid, trichilemmal, or sebaceous), tumors (parotid, giant cell, chondromas, eccrine spiradenomas, hydrocystomas, degenerating fibroxanthomas, osteochondromas, dermatofibroma, trichoepitheliomas, basal cell epitheliomas), and other cutaneous lesions (foreign body reactions, fat necrosis, or lymphadenopathy) [1,2,17]. Due to the broad differential diagnosis, a pre-operative diagnosis of pilomatricoma is correctly made in only 46% of cases [17].