Hard Subcutaneous Lumps—Pilomatricomas: A Report of Three Cases

Venkat Prasad, MD

Calcifying epitheliomas are uncommon tumors involving the hair matrix. Calcification is seen in most of these tumors.[1] More than 75% of cases occur in the head and neck region during the first two decades of life.[2] Treatment of choice is surgical excision. Calcifying epitheliomas can be multiple[3] and rarely manifest atypical changes or malignant behavior.[4] I describe three cases of calcifying epitheliomas in the chest wall, elbow, and arm of young adults.

Case Reports

Case 1

A 22-year-old man came to the clinic requesting excision of a chest wall lump. He had been well and denied any trauma, foreign travel, or eating raw pork. He gave no history of pain or discharge from the swelling. Findings on review of systems were completely normal.

He was a short man of average build. His temperature was 99.0°F, his pulse 80 beats per minute, and his blood pressure 110/70 mm Hg. He had no hypopigmented or hyperpigmented skin patches and no rash, and his hair distribution was normal. There was a 1.5 × 2-cm solitary, hard, well-defined, subcutaneous lump about 3 cm below the midclavicular point. The lump was nontender and freely mobile, with normal overlying skin, and was not fixed to skin or deeper tissues. The surface was nodular. Physical examination findings were otherwise unremarkable.

Excision of the total lump was easily accomplished following local infiltration with lidocaine. On cut section the lump was hard, irregular, and tan-brown with flaky calcific material. The histology report described it as calcifying epithelioma of Malherbe with extensive ossification.

Submitted, revised, 23 July 1999.

From Tri-County Community Health Center, Dunn, North Carolina. Address reprint requests to Venkat Prasad, MD, Tri-County Community Health Center, 33–31, Easy St, Dunn, NC 28334.

Case 2

A 29-year-old woman complained of a 6-month history of a painful lump on the inner aspect of the right arm. The pain was localized to the lump and had an aching, dull character with no radiation or throbbing. She denied any tingling, numbness, or weakness in the upper limb. There was no history of trauma, foreign travel, fractures, or eating raw pork. She had had an incision and drainage of the lump 2 months previously with no relief. Apparently, a thick whitish material had been expressed at the time of the incision and drainage. She had also noticed a nonhealing wound over the site of the incision. The woman was of average build. Her pulse was 74 beats per minute, blood pressure 104/68 mm Hg, respirations 18/min, and temperature 98.4°F.

The patient had a few acne scars and comedones on her face, but no other hypopigmented or hyperpigmented patches or rash. A single lump 2.5 × 1.5 cm was found on the medial aspect of her right arm about a hand's breadth above the medial epicondyle. The lump was subcutaneous, tender, and adherent to the skin. There was a 1-cm ulcer with a whitish base and sloping edges over the central part of the lump, but there was no increased warmth locally. The lump was not fixed to deeper tissues. There was no lymphadenopathy, and no neurologic deficit was evident. Bones and joints in the extremity were normal. Findings of a physical examination were otherwise unremarkable.

Local infiltration with lidocaine was given, and an ellipse of skin to include the ulcer was excised along with the entire lump. The lump was pale and white with keratinous material. A histology report described it as being consistent with calcifying epithelioma of Malherbe.

Case 3

An 18-year-old woman complained of a painful lump that had been over her right elbow since she was 2 years old. The lump had been static in size after an initial slow growth. The pain was sharp,

pricking, and localized. She had no tingling, numbness, or weakness in the upper limb. She denied any trauma, fractures, excessive strain or exertion of her arms, foreign travel, or eating raw pork. She had no history of any prolonged pressure over the area.

This lean, young girl had a temperature of 99°F, pulse of 84 beats per minute, blood pressure of 96/64 mm Hg, and respirations of 16/min. She had smooth skin with no rash or abnormal pigmentations. A single 3 × 2-cm lump was evident about 2 to 3 cm above the lateral epicondyle of the right arm. The lump was well-defined, mobile, hard, and nontender, and it was not fixed to skin or deep tissues. There was no increase in local temperature. Overlying skin was normal in color and texture. There was no neurologic deficit. Bones and joints in the extremity were normal.

Local infiltration anesthesia with lidocaine was used, and the entire mass was easily excised. It was a light pink, tan-brown, firm, ovoid mass. Histologically it was described as a calcifying epithelioma of Malherbe.

Discussion

Malherbe first described calcifying epithelioma or pilomatrixoma in 1880.[5] Moehlenbeck's analysis[3] of 140,000 skin tumors found an incidence of 0.12%.

Calcified epitheliomas or pilomatricomas must be included in the differential diagnoses of hard subcutaneous lumps. Other, subcutaneous lumps (Table 1) include lipomata, sebaceous cyst, calcified hematoma, parasitic cysts, tumoral calcinosis, foreign body granuloma, dermoid cyst, chondroma, osteoma cutis, and metastatic bone formation.[5] Pilomatricomas originate in the hair follicle, as confirmed by immunohistochemical and electronic microscopy studies. Marrogi et al,[2] reporting on 76 tumors from 69 patients, found their patients' average age to be 8 years, with a female-to-male predominance of 3:1. Although most of these tumors tended to occur in the head and neck regions, they have also been reported in the parotid gland,[6] in the eyelids,[7] and intraorally.[8]

The median size of these tumors is 1.2 cm[9] Lopansri and Mihm[10] reported that the tumor does not have to be large to show aggressive behavior. Yoshimura et al[11] reviewed 37 cases diagnosed during a 17-year period, and found a mean age of 23 years and a female-to-male ratio of 2.4:1. Only 2 patients (5%) had multiple tumors. The average size was 0.5 to 3 cm. The most common site was the preauricular region. They recommend complete excision, an opinion expressed universally in all studies reviewed, including any area of attached skin, because it is often impossible clinically to separate benign from malignant masses.

Lesions with typical features, such as cords and sheets of basaloid cells with sheets of ghost or shadow cells and sometimes squamous pearls or abortive hair follicles, rarely recur; those showing atypia, such as cell necrosis, mitotic figures, or prominent nucleoli, can be locally aggressive. Frank carcinoma has been observed in lesions that show necrosis, mitoses, large vesicular nuclei, and irregular borders.[2]

Zhang,[1] in his study of 73 cases, reported an incidence of 0.094%. Most of his patients (70.2%) were children or young adults, and 47 patients (64%) were female. Most of the tumors (88.5%) measured 0.5 to 1.5 cm. Ossification was found in 21%, whereas calcifications were seen in 86%.

Ichikawa et al[12] described a large 18 × 12 × 8-cm right upper arm pilomatricoma that was subcutaneous and calcified. After excision, there was no recurrence within a 3-year follow-up.

Pilomatricomas have been reported to be associated with myotonic dystrophy[13] and Gardner syndrome.[10]

McCulloch et al[14] described a case of a 25-year-old mentally disabled man who had 11 cutaneous scalp lesions excised within a 19-year period. None was examined histologically, as the clinical diagnosis of epidermal cyst was made. A fast-growing new lesion on his scalp, which became ulcerated and infected, was debrided without histologic examination of the lesion. One year later, after a recurrence of the problem, histologic examination of the material was obtained, and a diagnosis of aggressive pilomatricoma was made. The authors hypothesized that the case possibly represents one of multiple benign pilomatricomas that later turned aggressive. They emphasized the need for histologic confirmation of all lesions undergoing excision.

Death from local invasion and even metastatic lesions has been reported in cases of pilomatrical carcinoma.[15] Most of these cases, however, are benign, and simple excision results in cure. When the lesion is associated with atypical histologic features, close long-term follow-up is recommended.

Table 1. Subcutaneous Lumps and Nodules.

Lumps	Number	Distribution	Clinical Features
Lipoma	Single, can be multiple	Anywhere except palms, soles	Soft, lobulated, slips under examining finger; usually painless; variable size; no fixity to skin or tissues
Sebaceous cyst	Single usually; if multiple or in childhood—Gardner syndrome	Hair-bearing areas	Soft unless calcified, cystic, punctum, present; fixed to dermis; soft cheesy, keratinous contents
Calcified hematoma	Usually single	Anywhere	History of trauma with skin discolorations. Ill-defined edges, usually painless
Parasitic cyst	Single or multiple	Anywhere, at times intramuscular	History of third world travel or origin, eating raw pork, meats. Firm, cystic, well-defined, poorly mobile
Tumoral calcinosis	Single or multiple	Elbows, iliac crest	Hard, nodular, whitish chalky material
Foreign-body granuloma	Usually single	Anywhere	History of trauma, foreign body. Ill-defined edges, painful at times. Chronic
Dermoid cyst	Single	Midline, eyebrows	Congenital, cystic, smooth, well-defined, nontender; at times bony erosion palpable under cyst
Chondroma	Single or multiple	Chest wall	Painful, nodular, firm to hard
Osteoma	Single	Skull, femur (adductor magnus—rider's bone), knee (medial collateral ligament)	Smooth, painless, bony hard. If associated with multiple sebaceous cysts—Gardner syndrome
Pilomatricoma	Single	Head and neck, extremity	Hard, well-defined, nodular; 1–3 cm
Neurofibroma	Multiple	Anywhere, along nerve distribution	Appear at puberty, soft, sessile, or pedunculated. Button-holing on compression, dome-shaped, violet hue
Keratoacanthoma	Single or multiple	Limbs, trunk	Elderly. Possible viral cause. Start as 1–2-mm papule, then enlarges to 2 cm or more. Nodular with central keratin crater
Tuberous sclerosis	Multiple	Cheeks	Firm, pink- to flesh-colored, papular, 3 mm to several centimeters (adenoma sebaceum)
von Recklinghausen	Multiple	Generalized	Varying sizes, soft, pedunculated, nontender; autosomal dominant; café au lait spots, axillary freckling
Dermatofibroma	1–10	Limbs, trunk, especially anterior surface of leg	3–10-mm, raised, pink-brown, hard; nodules retract under the skin on compression
Pilar cyst	Single or multiple	Scalp, scrotum	Movable, cystic, varying size. Soft, nontender, cannot be moved separate from skin. Cheesy contents
Rheumatoid nodules	Multiple	Elbows, pressure areas	History of rheumatoid arthritis, 0.5–4.0 cm, hard, nodular
Tophaceous gout	Multiple	Ear helix, elbows, prepatellar	Firm, yellow; occasionally discharge chalky material. Contents monosodium urate. History of gout, hyperuricemia
Dermal metastases	Multiple	Anywhere	Firm, flesh-colored nodules can be pink, blue, or black. In men, associated with colon and lung cancer and melanoma. In women, associated with breast, colon, and lung cancer. Initial finding in some cases of kidney, lung, or ovarian cancer.

Conclusion

I report an uncommon subcutaneous tumor originating from the hair matrix in 3 patients. The sites were uncommon, occurring as they did on the chest wall in one and on the elbow and arm in the other two. Hard calcific nodules in subcutaneous areas merit inclusion of pilomatricoma in the differential diagnosis. Histologic confirmation of all excised lesions is important to avoid missing any atypical or aggressive tumors.

References

1. Zhang ZC. [Histopathology of 73 cases of calcifying epithelioma]. Chung Hua Chung Liu Tsa Chih 1991;13:313–5.

2. Marrogi AJ, Wick MR, Dehner LP. Pilomatrical neoplasms in children and young adults. Am J Dermatopathol 1992;14(2):87–94.
3. Moehlenbeck FW. Pilomatrixoma. Arch Dermatol 1973;108:532–4.
4. Sau P, Lupton GP, Graham JH. Pilomatrix carcinoma. Cancer 1993;71:2491–8.
5. Rotenberg M, Laccourreye O, Cauchois R, Laccourreye L, Putterman M, Brasnu D. Head and neck pilomatrixoma. Am J Otolaryngol 1996;17:133–5.
6. Costa P, Orlando A, Papadia F. [Pilomatrixoma of the parotid region]. Pediatr Med Chir 1993; 15:601–3.
7. Seitz B, Holbach LM. [Medial canthus pilomatrixoma in an 80-year-old patient]. Klin Monatsbl Augenheilkd 1993;203:444–5.
8. Vasilas A, Ganasouli D, Stylogianni E, Sklavounou A. [Calcifying epithelioma of Malherbe]. Stomatologia 1990;47:174–80.
9. Aguirre JM, Lopez Cedrun JL, Martinez-Conde R, Martin JC, Lopez Vincente J, Rivera JM. [Malherbe's calcifying epithelioma (pilomatrixoma) of the head and neck]. Rev Stomatol Chir Maxillofac 1991; 92:44–7.
10. Lopansri S, Mihm MC Jr. Pilomatrix carcinoma or calcifying epitheliocarcinoma of Malherbe: a case report and review of the literature. Cancer 1980;45: 2368–73.
11. Yoshimura Y, Obara S, Mikami T, Matsuda S. Calcifying epithelioma (pilomatrixoma) of the head and neck: analysis of 37 cases. Br J Oral Maxillofac Surg 1997;35:429–32.
12. Ichikawa T, Nakajima Y, Fujimoto H, et al. Giant calcifying epithelioma of Malherbe (pilomatrixoma): imaging features. Skeletal Radiol 1997;26:602–5.
13. Chiaramonti A, Gilgor RS. Pilomatricomas associated with myotonic dystrophy. Arch Dermatol 1978; 114:1363–5.
14. McCulloch TA, Singh S, Cotton DW. Pilomatrix carcinoma and multiple pilomatrixomas. Br J Dermatol 1996;134:368–71.
15. Mir R, Cortes E. Papantoniou PA, Heller K, Muehlhausen V, Kahn LB. Metastatic trichomatricial carcinoma. Arch Pathol Lab Med 1986;110:660–3.