Dear Editor,

Pilomatrixoma, also known as Malherbe’s calcifying epithelioma, originates in pluripotent precursor cells of the hair follicles. The tumor was first mistakenly described by Malherbe and Chenanatis as a benign tumor of the sebaceous gland, then later recognized as related to the hair follicles.[1]

Pilomatrixoma is a slow-growing, dermal tumor, usually found on the head and neck region, and less frequently on the trunk and lower extremities. Most cases are detected in children and young adults, with a female predominance, but it may be seen at any age and in either gender.[2]

Clinically, pilomatrixoma typically presents as a painless, solitary, superficial, mobile lesion that easily slides over the underlying subcutaneous tissue. An irregular, hard structure can be palpated when there is significant calcification. A blue discoloration may indicate ulceration of the mass. Other characteristic features of the lesion are an average size of 10 mm or less, consistency ranging from firm to cystic, moderate pattern of growth, pink to purple hue with sub-epithelial yellowish tinge, and intact overlying skin with telangiectasic vessels.

Differential diagnosis should include epidermoid cyst, dermoid cyst, sebaceous adenoma or carcinoma, juvenile xanthogranuloma, capillary hemangioma, chalazion, and rhabdomyosarcoma.[3–5] Although pilomatrixoma grows slowly, it occasionally demonstrates rapid growth and can resemble keratoacanthoma of the skin.[6]

Ultrasonography (US) investigation is helpful to detect pilomatrixoma.[7] The accuracy rate for a round, well-demarcated, hyperechogenic mass with a dense posterior acoustic shadow is approximately 80%.[8] US examination and findings of expected features can confirm the diagnosis. Computer tomography and magnetic resonance imaging examinations also provide more detail of surrounding structures and the depth of the lesion.

Fine needle aspiration biopsy (FNAB) may reveal diagnostic clues for diagnosis of pilomatrixoma, but can also lead to misdiagnosis of carcinoma and result in aggressive surgery.[9] In a previous study, the diagnostic accuracy of FNAB for pilomatrixoma was found to be 44.4%.[10]

Histopathologically, pilomatrixoma has sharply demarcated dermal nodules surrounded by a capsule of fibrous tissue located in the dermis, extending into the subcutaneous fat. Shadow cells, also known as ghost cells, which evolve from basaloid cells, are dead cells that retain their cellular shape and appear as a central, unstained area that corresponds to the lost nucleus. The tumor often has calcification structures around the ghost cells, and there may be foreign body giant cells around the keratinized debris.

Pilomatrixoma will not spontaneously regress, so surgical excision of the tumor and the
overlying skin is the primary treatment modality. After total excision of the tumor, the recurrence rate varies from 1.5% to 6%.[11,12] Malignant transformation is only rarely reported, typically a low grade tumor, with higher recurrence potential in elderly patients.[13]

A 20-year-old male patient presented at our clinic with a mobile, slow-growing, asymptomatic 2x2 cm lesion, firm and cystic in appearance, on the posterior cervical triangle of the neck. Pathological examination revealed a solid, well-demarcated, tumoral lesion surrounded by fibrous pseudocysts. The tumor was composed of small, round, uniform, vesicular nuclei, and study of the nuclei revealed basaloid cells and ghost cells with eosinophilic appearance. The diagnosis was pilomatrixoma (Figure 1). No recurrence has been detected since excision in 1 year of follow-up.

Pilomatrixoma is a rare, benign form of dermal tumor, most often seen in females and adolescents, but one that may appear at any age and in either gender. Treatment is total excision. The differential diagnosis can be confusing for the clinician. Keep pilomatrixoma in mind when a firm, mobile, dermally located lesion is detected in the head and neck region.

Peer-review
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Authorship Contributions

Conflict of Interest
None declared.

REFERENCES
1. Lever W, Griesemer RD. Calcifying epithelioma of Malherbe; report of 15 cases, with comments on its differentiation from calcified epidermal cyst and on its histogenesis. Arch Derm Syphilol 1949;59:506–18. [CrossRef]