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Pilomatricoma in the Eyelid: A Case Report[1]

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The pilomatricoma (calcifying epithelioma of Malherbe) is a rare, benign, asymptomatic tumor arising from the hair follicular matrix cells. It is usually a solitary lesion and it is most commonly found on the face and upper extremities.

This tumor can occur in patients of any age, but about 60% of pilomatricomas are found in those patients less than twenty years old. The radiologic features of this neoplasm have rarely been described in the literature; in particular, the CT findings of pilomatricoma originating from the eyelid have not been published in Korea. We report here on a case of pilomatricoma that occurred in 69-year-old female; she had a hard subcutaneous mass on her right eyelid with overlying bluish skin.

Index words : Skin, neoplasms
Computed tomography (CT), pilomatricoma

Pilomatricoma is a benign, usually asymptomatic neoplasm that arises from the hair follicle matrix cells. The lesions usually appear as firm, solitary or multiple tumors of the dermis. It was initially described by Malherbe and Chenantais in 1880 as a calcifying tumor that originates from the sebaceous glands. Forbis and Helwig were the first to advocate the use of the term pilomatricoma for such tumors (1, 2).

Pilomatricoma is commonly found on the face and upper extremities and they demonstrate a peak age at the first and sixth decades of life. However, 60% of the cases occur within the first two decades of life (3 - 5).

The female to male ratio is about 3:2. Generally, pilomatricoma is not hereditary: however, multiple tumors occur in about 3% of the cases as an autosomal dominant inherited disorder (6 - 9).

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The diagnosis is usually based on palpation of a superficial, hard mass and it's confirmed by histopathologic examination.

On a review of the literature, the CT findings of pilomatricoma on the eyelid of an aged patient have not previously been reported in Korea, so we report here on a case of pilomatricoma in an elderly female.

Case Report

A 69-year-old female was referred to our hospital because of a painless mass on her right eyelid, which had been slowly growing for the past two years. Her past medical history was unremarkable. On the physical examination, the tumor appeared as about a 2 cm sized, movable, non-tender soft tissue mass with overlying bluish discolored skin. There was no evidence of ulceration or discharge. Orbit CT demonstrated a subtly enhancing, ovoid subcutaneous mass lesion in the right upper eyelid. The adjacent periorbital muscles seemed to be intact and the intraorbital organs were unremarkable.

The differential diagnosis included solid tumors such as pilomatricoma or sebaceous origin neoplasm as well as cystic tumors such as epidermoid or dermoid cyst because there was little contrast-enhancement of the tumor. But because the degree of enhancement was very small, the differential diagnosis included cystic tumor such as epidermoid or dermoid cyst.

There was no detectable abnormality in the complete blood count or blood chemistry. The mass, about 1.5 cm in diameter, was successfully excised.

Grossly, we found irregular friable soft tissue mixed with surrounding connective and adipose tissue. Histopathologically, it was a tumor showing two components of central shadow cells and basophilic cells, along with foreign body giant cells. There were tiny microcalcific foci with shadow cell nests (Fig. 5). The final pathologic diagnosis was a pilomatricoma.

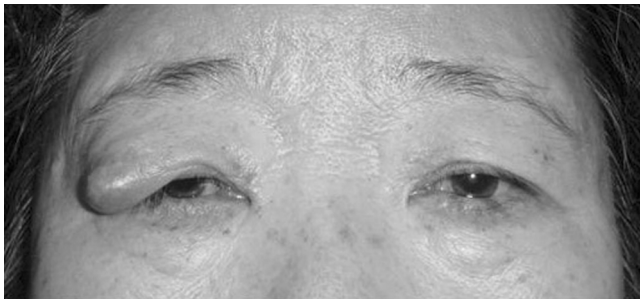


Fig. 1. A 69-year-old female has about a 2 cm sized, movable, nontender soft tissue mass with overlying bluish discolored skin in her right upper eyelid.

Discussion

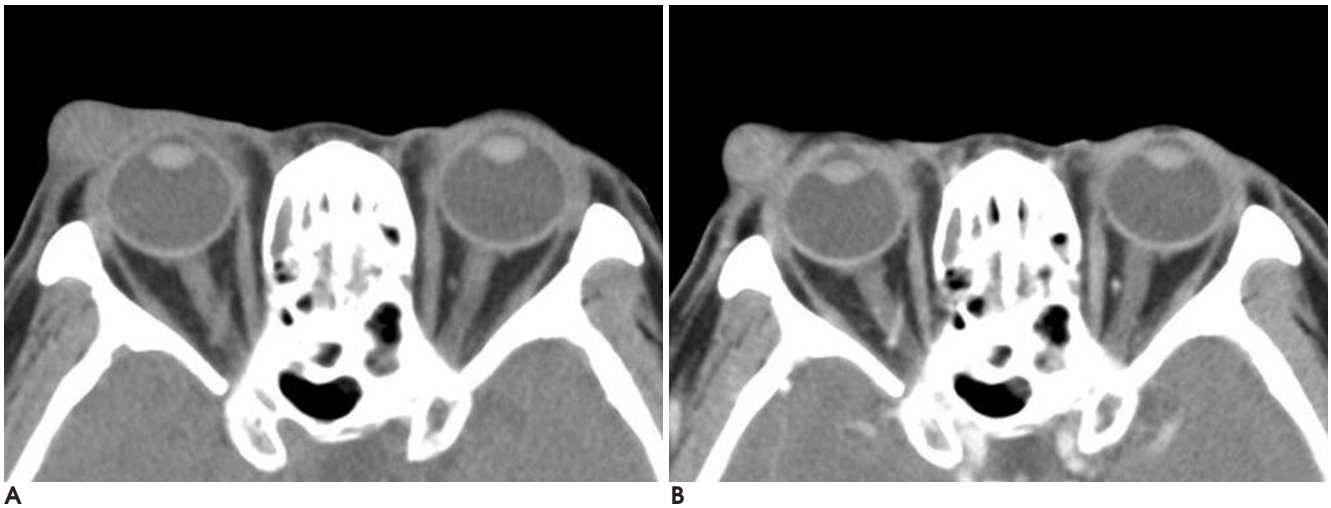
In 1880, Malherbe and Chenantais initially described a benign neoplasm of the skin that was thought to be derived from the sebaceous glands and arising in the subcutis, and they termed this a calcifying epithelioma (1, 3). In 1961, Forbis and Helwig found 500 pilomatrixomas in the files of the Armed Forces Institute of Pathology in 1961 and they renamed this neoplasm pilomatrixoma (2). In 1977, the name was changed to pilomatricoma to be more etymologically correct.

Moehlenbeck found 1569 cases in the literature in 1973 and this researcher described the low incidence of pilomatricoma; it represents about 0.12% of the neoplasms involving the integument (4).

Numerous studies have demonstrated that pilomatricoma most commonly occurs in the head and neck, and this location is followed by the upper extremity, trunk and lower extremity. The reported age range of the patients with pilomatricomas in the literature is from one week of the age to 77 years, with the greatest incidence between 8 and 13 years. The gender distribution is stated to be 3 females to 2 males (3, 4).

Pilomatricoma is not hereditary. However, family occurrences have been reported in about 10 cases (5). Gardner's syndrome and myotonic muscular dystrophy have been associated with cases of pilomatricoma (6).

The tumor is usually asymptomatic, but some cases have been associated with pain during episodes of in-



A B

Fig. 2. The precontrast axial CT (2A), and postcontrast axial CT (2B) images show a subtle enhancing, 1.5×1.0 cm sized, ovoid subcutaneous mass lesion without discrete calcification in the right upper eyelid.

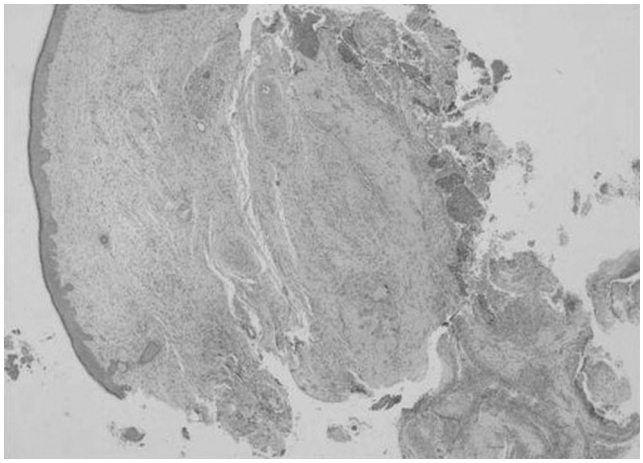


Fig. 3. (H & E ×40) A relatively sharply demarcated nodule in the lower dermis of the upper eyelid shows irregularly shaped islands of epithelial cells and prominent cellular stroma.

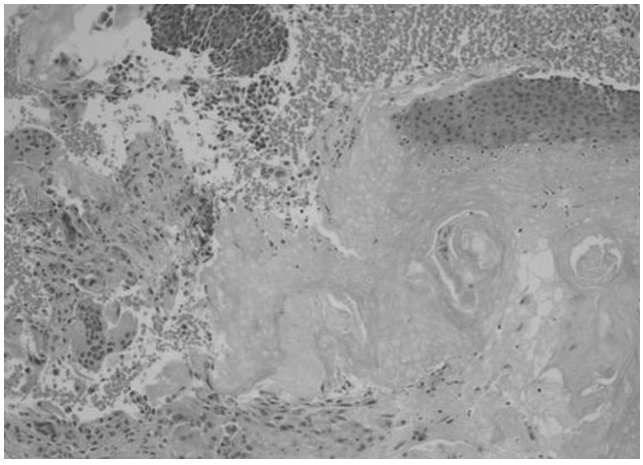


Fig. 4. (H & E, ×200) On higher magnification of the pilomatricoma, two types of cells, basophilic cells and shadow cells, compose the epithelial islands. The stroma adjacent to the shadow cells shows foreign-body reaction that contains many giant cells.

flammation or ulceration. Pilomatricoma is usually a solitary nodule, but multiple occurrences have been observed in 4% of cases.

A rare malignant variant with distant metastasis to the lung, bone, brain, skin and abdominal organs has been described (7).

The tumor is separated from the epidermis by a layer of fibrous tissue, so the tumor can usually be easily enucleated from the subcutaneous area (3, 8).

Grossly, pilomatricomas typically remain well circumscribed even when they extend into subcutaneous tissues. Histologic examination reveals polygonal basophilic cells that are nucleated and arranged either on one side or along the periphery of the tumor islands, and

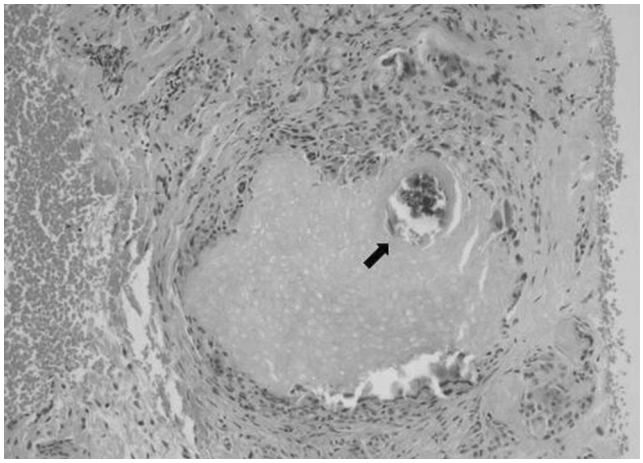


Fig. 5. (H & E, ×200) This tumor histologically shows a very focal microcalcification (arrow) within a shadow cell nest.

there are inflammatory cells as well as ghost or shadow cells formed by the keratinization of the basophilic cells. In some tumors, the transitional zone the between the basophilic cells and shadow cells was distinct, but in other tumors it occurred over multiple layers. Generally, as the lesion matures, the number of basophilic cells decreases and the percentage of shadow cells increases. Similarly, in our case, shadow cells were an absolutely majority microscopically. The reported incidence of calcification ranges from 69% to 85%, and this was seen in our case (9).

Plain X-ray films show nonspecific calcification, and ultrasonogram can demonstrate a well defined, round, hyper-echoic or hypo-echoic mass with a posterior dense acoustic shadow. Ichikawa et al. have reported on a case of giant pilomatricoma that was depicted at angiography as a hypervascular mass (8). Fink and Berkowitz reported on the cross sectional imaging of pilomatricoma; this showed a sharply demarcated, subcutaneous, opaque lesion that didn't enhance after injection of the contrast material (CT scan) or there were small areas of signal dropout that were consistent with the presence of calcification (MR scan) (8). Although many radiologic modalities can help diagnose pilomatricoma, such histopathologic examinations as fine needle biopsy should be done for confirmation.

However, the other entities to be considered in the differential diagnosis include cystic tumors such as epidermal inclusion cyst or dermoid cyst, malignant tumor such as squamous cell carcinoma or basal cell carcinoma, conjunctival origin tumor such as nevus, papilloma, carcinoma or melanoma, catilageneous origin tumor such as chondroma, and other benign conditions such as ossifying hematoma, foreign body reaction, etc.

Since the spontaneous regression of a pilomatricoma has never been reported, the treatment of choice is a surgical excision with clear margins (10). The recurrence rate is very low if the tumor is completely excised. Most recurrences arise due to incompletely excised neoplasm and to minimize this risk, wide resection is recommended (6, 10).

In summary, we suggest that if CT reveals a well-circumscribed, isoattenuated or hyperattenuated subcutaneous mass in the eyelid with little or no enhancement, whether or not sand-like calcification is visible, pilomatricoma should be included in the differential diagnosis. A histopathologic examination should be performed for confirmation.

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안검에 생긴 섬유기질종: 증례 보고[1]

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강 채 훈·유 동 수

섬모기질종(Malherbe씨 석회화 상피종)은 드문 양성 종양으로써, 모낭기질세포로부터 기인하는 무증상의 종양이다. 보통 단발성 질환이고, 안면부와 상지에서 가장 호발한다.

이 종양은 어느 연령에서나 발생할 수 있으나, 20세 이하에서 섬모기질종의 약 60% 정도가 발견된다. 이 종양의 방사선학적 소견은 드물게 보고되고 있으며, 특히 국내에서 안검부에서 발생한 섬유기질종의 CT 소견은 발표된 바가 없다. 저자들은 69세 여자 환자에서 푸른색 피부로 덮힌 우안검부 피하 종양으로 발생한 섬유기질종 1예를 보고하고자 한다.