Pilomatricoma of the Shoulder Easily Identified by Color Doppler Ultrasound: A Case Report and Review of Literature

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Pilomatricoma is a benign skin tumor that develops from hair matrix cells. It most commonly occurs in the head and neck, followed by the upper extremities. Accuracy of preoperative diagnosis was low in previous studies and excisional biopsy was even performed frequently without imaging studies. We report a case of pilomatricoma of the shoulder that was easily diagnosed by ultrasound including color Doppler, which is a more useful imaging modality than computed tomography or magnetic resonance imaging scans not only because of its cost effectiveness but also because of the precise information obtained from mass contents.

Key Words: Pilomatricoma, Shoulder, Ultrasound, Color doppler, Calcification

Pilomatricoma, also known as pilomatrixoma or calcifying epithelioma of Malherbe, is a benign skin tumor that develops from hair follicle matrix cells.\(^1,2\) Pilomatricoma is most commonly developed in the head and neck and are usually found in the first two decades of life.\(^2,3\) The usual presentation is that of a hard, usually solitary, mobile, slow growing mass of the dermis or subcutaneous tissue that may cause ulceration or a discoloration of the superficial skin.\(^1\) However, previous study showed that the accuracy of clinical diagnosis has been low, ranging from 28.9% to 46%.\(^1,4\)

Sometimes, patients are treated without imaging studies by dermatologists and plastic surgeons. Recently, ultrasound (US) has been used by an imaging modality of choice on these types of superficial masses.\(^5,6\) We now describe a case of pilomatricoma of the shoulder that was easily diagnosed by US.

Case Report

A 3-year-old male presented with complaints of a painful right shoulder mass which was incidentally found 1 month previously. His past history was not significant. On physical examination, he had no gross deficits in muscle power. The mass was palpable over his right shoulder lateral aspect and was fingertip
sized, ovoid, firm and movable. The plain radiographs showed a 12×8 mm sized ovoid shaped radiopaque mass in the proximal humerus lateral aspect (Fig. 1). US showed 13×7×8 mm sized ovoid shaped, well defined, mixed echogenicity, heterogenous echotexture, peripheral hypoechoic rim and posterior shadowing. On color Doppler US images, the presence of peripheral vascularity was identified (Fig. 2). US findings suggest a pilomatrixoma or calcified epidermoid cyst.

The patient was taken to the operating room, where he underwent excisional biopsy of the mass. The mass was located in the fat tissue without a capsule but was clearly demarcated from the surrounding fat tissue. The mass was whitish and firm but brittle (Fig. 3). The pathology revealed typical basaloid cells and shadow cells with calcifications diagnosed as pilomatrixoma (Fig. 4).

**Discussion**

Pilomatrixoma is a tumor of the hair matrix that is almost always benign, with less than 20 cases of malignant transformation reported in the literature. Pilomatrixoma most often presents in patients before their early twenties, and especially among children. There is a slight female predominance (male/female ratio, 1/1.1 to 1/2.4). The head and neck is the most common (40~77%) location of development especially of periorbital lesion, followed by the upper extremities, the trunk and the lower extremities. It can occur in any hair-bearing skin and is usually located in the...
dermis or subcutaneous fat layer, however a deeply seated and invisible lesion has also occurred. Pilomatricoma commonly forms as a solitary mass, but multiple lesions were found for 2.3% to 10% of reported cases. The clinical presentation is that of a firm, mobile, well demarcated nodule. Histopathologically, pilomatricoma consist of irregular epithelial islands lined by basaloid (basophilic) cells with prominent nuclei at the periphery and are filled in the center with eosinophilic keratinized anucleated shadow (ghost) cells. Basaloid cells are usually less in number than shadow cells and sometimes there are no basaloid cells. Shadow cells are identified by the lost nuclei, which leaves a central unstained area. These are typical findings of pilomatricoma. Calcium deposition in shadow cells or stroma is a common feature. Granulomatous inflammation, dystrophic ossification, or melanin pigmentation can also be identified in regions where keratinized debris is plentiful. In our case, typical inner shadow cells and outer basaloid cells with calcium deposition were identified. The mass itself has no cystic contents and dystrophic ossification and melanin pigmentation were not found.

The differential diagnosis of pilomatricoma includes sebaceous, trichilemmal, dermoid and epidermoid cysts, calcified lymph nodes, metaplastic bone formation, foreign body granuloma, calcified hematoma, hemangioma, cutaneous osteoma, osteochondroma, trichoepithelioma and basal cell epithelioma. The diagnosis can be made by a careful clinical examination accompanied by a high suspicion. The accuracy of clinical diagnosis had been low, ranging from 28.9% to 46%. However, the high-resolution US based pre-operative diagnostic rate had increased by 76% to 82% in recent studies. From the US, pilomatricoma was described as well-
defined, hyper or isoechoic subcutaneous masses with posterior shadowing and a hypoechoic rim.\(^5\)\(^6\)\(^9\) In our case, the preoperative diagnosis was easily performed by the US. The mass was surrounded by a peripheral hypoechoic rim. Mixed echogenicity, heterogeneous echotexture, and posterior shadowing represent calcium deposition in the mass. On color Doppler US, the presence of peripheral vascularity was identified. Furthermore, US including color Doppler differentiated pilomatrixoma from a dermoid cyst, hemangioma, and benign lymph node hyperplasia.\(^5\) Epidermoid cysts appear as hypoechoic masses with inner anechoic foci rather than echogenic foci and do not show Doppler flow. Hemangioma has a lobulated contour and a soft consistency. The associated areas of calcification (phleboliths) of the hemangioma are round, and low-frequency Doppler flows are evident within anechoic vascular channels. Most lymph nodes have central Doppler flow at their fatty hilum, whereas pilomatrixoma tend to have peripheral Doppler flow. Spontaneous regression of pilomatrixoma has never been observed, and the treatment of choice is surgical excision.\(^1\)\(^3\) Malignant transformation has been rarely described. The recurrence rate of pilomatrixoma is very low in complete surgical excision,\(^1\)\(^3\) but incomplete excision may result in recurrence. Therefore, wide resection is recommended to minimize that risk.

Pilomatrixoma is one of the possible diagnoses of skin mass developed in an extremity, even though the head and neck are common sites of development. The US including color Doppler was a more useful imaging modality than the computed tomography or magnetic resonance imaging scans not only because of its cost effectiveness but also because precise information is obtained about the contents of mass.

**Conflict of interest**

No potential conflict of interest relevant to this article was reported.

**참고문헌**

모기질세포종은 모발의 기질세포로부터 발생하는 양성 피부종양이다. 이것은 주로 두 경부 및 상지에 호발한다. 수술 전 진단의 정확성은 낮으며 단순방사선 검사만 시행한 후 절제생검을 하기도 한다. 저자들은 컬러도플러 초음파를 이용해서 쉽게 진단할 수 있었던 견부에 발생한 모기질세포종에 대해 보고하고자 한다. 초음파를 이용한 진단은 비용효과적인 측면 뿐만 아니라 종양의 내부에 대해 자세한 정보를 알 수 있다는 점에서 전산화 단층촬영이나 자기공명영상보다 더 손쉽고 유용한 진단방법으로 생각된다.

색인단어: 모기질세포종, 견부, 초음파, 컬러 도플러, 석회화