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A CASE SERIES OF PILOMATRICOMA: IMAGING FEATURES WITH FINE NEEDLE ASPIRATION CYTOLOGY CORRELATION

Radiology

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ABSTRACT

Introduction: Pilomatricoma is a benign skin neoplasm arises from hair follicle matrix cells, which is not commonly encountered in general practice. They present as superficial masses of the head and neck especially in children.
Case Series: Five cases were included in the present study, of which four were males and one female. The sites involved were scalp, face and chest.
Conclusion: Although there have been several case reports in the literature describing the clinical & imaging features and diagnostic pitfalls of pilomatricoma, this lesion continues to cause difficulty in clinical diagnosis. The idea of reporting these cases is that, pilomatricoma is not rare in occurrence but rarely diagnosed because of lack of confirmation of excised swelling and it is imperative that it should be kept in the differential diagnoses of all superficial skin tumors by radiologists, dermatologists and surgeons.

KEYWORDS

Pilomarixoma, Pilomatricoma, Calcifying epithelioma of Malherbe, Basaloid cells, FNAC.

INTRODUCTION:

Pilomatricoma also known as Pilomatrixoma or calcifying epithelioma of Malherbe is a benign skin neoplasm, which is derived from hair follicle matrix cells. These tumors are typically present in the head and neck region, but also occur in the upper limbs and are rarely reported in other sites[1]. It is more common in females and usually presents during the first two decades of life (60%)[2]. It represents as an asymptomatic, solitary, firm to hard, freely mobile nodule of the dermis or subcutaneous tissue and generally exhibits no fixation to neighbouring tissues but have an osseous- or cartilage like hardness [3,4,5]. The size of the tumor rarely exceeds 3 cm. The overlying skin may exhibit a bluish discoloration or ulceration [6].

CASE SERIES:

Five cases were included in the present study. Four (80%) of these were male and one (20%) female. The site of involvement in these patients were different (Table 1). Two cases (40%) occurred in the scalp, two cases (40%) on the chest and one case (20%) on the face . All the patients presented with a painless swelling. On examination the swellings were non-tender and freely mobile.

Table-1: showing Age, Sex and Site distribution of Pilomatricoma cases.

Case	Age (in years)	Sex	Site involved
1	27	Male	Cheek
2	45	Male	Back
3	49	Male	Chest
4	35	Female	Scalp
5	20	Male	Scalp

CASE-1:

A 27-year-old male presented with a 3 month history of insidious onset of an isolated swelling over right cheek. The mass was painless, progressively enlarging, not associated with itching and discharge. He denied any history of trauma over the site and no history of fever, chills, fatigue, weight loss, numbness and tingling. Physical examination revealed a 2cm x 2cm, non-tender, firm mass over the right cheek. It was superficial and easily mobile.

Ultrasonography show a well circumscribed oval shaped heterogeneously hyperechoic mass lesion in the subcutaneous location which shows multiple foci of calcification within and hypoechoic peripheral halo. On colour doppler , lesion shows mild vascularity in the centre . (Figure 1 A ,1B)

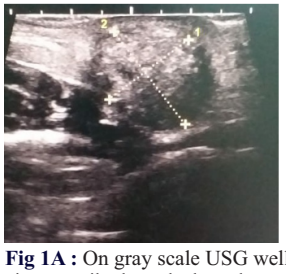


Fig 1A : On gray scale USG well circumscribed oval shaped heterogeneously hyperechoic mass lesion in the subcutaneous location which shows multiple foci of calcification within

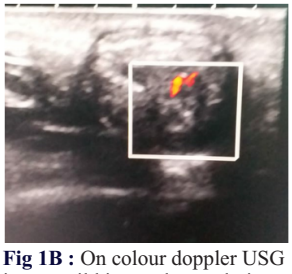
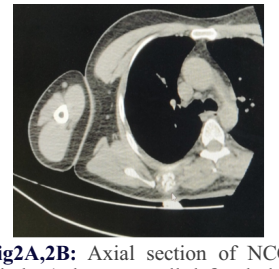


Fig 1B : On colour doppler USG image mild internal vascularity seen in the centre of lesion

CASE-2:

A 45-year-old male presented with a painless swelling over the right upper back since 1 year . On physical examination lesions are mobile, superficial and non tender. CT scan was done which shows a well-defined densely calcified oval shaped subcutaneous soft tissue lesion (Figure 2A,2B) .



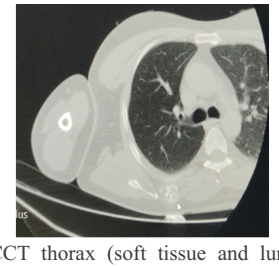


Fig2A,2B: Axial section of NCCT thorax (soft tissue and lung window) shows a well-defined densely calcified soft tissue lesion in the subcutaneous plane of right upper back.

CASE-3:

A 49 year old male presented with complain of cough & chest pain for which chest X-ray was done. Incidentally a calcified lesion is seen in the left upper chest wall below the clavicle (Figure 3A). On ultrasound a well defined predominantly calcified mass lesion showing posterior acoustic shadowing was noted in subcutaneous plane (Figure 3B).

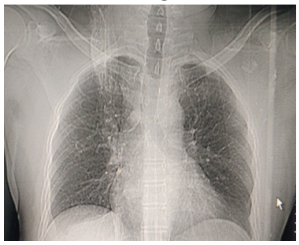


Fig 3A : Chest X-ray PA view shows a calcified lesion in the Left upper chest wall below the clavicle.

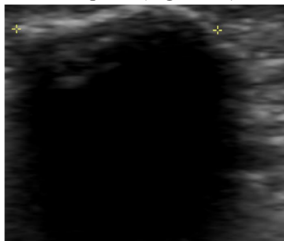


Fig 3B: Gray scale USG image shows a densely calcified subcutaneous lesion with posterior acoustic shadowing.

CASE-4 & 5:

Two patient, 35 year old female and 20 year old male presented with complain of headache for which CT brain was done . Incidentally nodular soft tissue lesion of approximate size 7mm x 7mm and 15mm x 12 mm with few calcific foci within is noted in the subcutaneous plane of scalp in the high frontal region. (Fig4A, 4B)

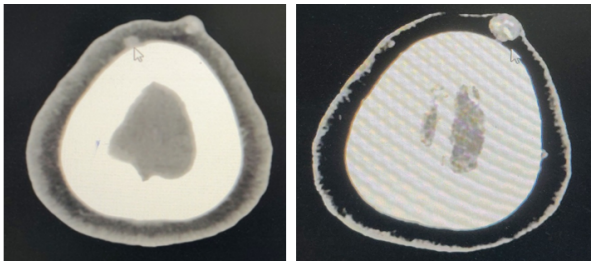


Fig 4A, 4B: Axial sections of CT brain shows two small partially calcified nodular lesions in the subcutaneous plane of scalp in the high frontal region.

Fine needle aspiration was carried out for the above mentioned lesions, slides were formed and sent for cytological examination which showed cell-rich aspirate containing predominantly basaloid cells in tight clusters, scattered squamous cells, anucleate squames, shadow cells, plenty of multinucleated giant cells and focal areas of calcification (Figure 5).

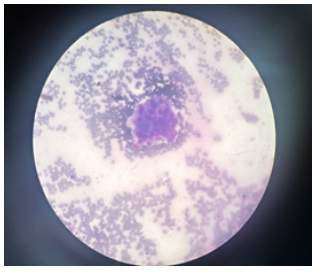


Fig 5: Photomicrograph of FNAC shows cluster of basaloid cells and scattered squamous cells characteristics of Pilomatricoma.

DISCUSSION:

Pilomatricoma, a benign neoplasm of the hair follicle, was initially thought to arise from sebaceous glands and was called calcifying epithelioma of Malherbe by Malherbe and Chenantais in 1880 [7]. In 1961, after histochemical and electron microscopic analysis of 228 such tumors, Forbis and Helwig found the cell of origin to be the outer root sheath cell of the hair follicle and proposed the name, pilomatricoma, now called pilomatricoma [8]. In 1973, Moehlenbeck reviewed 140,000 skin tumors and noted that pilomatricoma represented 0.12% of cases. Although pilomatricoma can develop in patients of any age, it occurs most often in children and young adults [9].

Diagnostic imaging is generally not obtained in the evaluation of pilomatricomas as they are usually superficial, small, and well-circumscribed. Plain radiographs in this case were unremarkable, but pilomatricomas may demonstrate foci of calcification. Ultrasound demonstrates a well-defined mass with inner echogenic foci and a peripheral hypoechoic rim or a completely echogenic mass with strong posterior acoustic shadowing in the subcutaneous layer [10]. Computed tomography (CT) demonstrates a sharply demarcated, subcutaneous lesion of soft tissue density, with or without calcification. MRI may reveal a rim-enhancing lesion with small areas of signal dropout which may be consistent with calcifications [11].

The histopathologic features of a pilomatricoma include a well demarcated tumor which is often surrounded by a connective tissue capsule. Generally, it is located in the dermal or subcutaneous layer. The tumor is composed of islands of epithelial cells made up of varying amounts of uniform basaloid matrical cells and often shows cystic change. Centrally, there is degeneration of these basaloid cells as the tumor matures. This is characterized by formation of anucleated ghost (or shadow) cells due to the central unstained areas of these cells. It is important to note, however, that these ghost cells, though quite specific, are not unique to pilomatricomas. Foreign body giant cells, keratin debris, and central calcifications are also characteristic. Calcification has been noted in 70 to 85 percent of cases. Wang et al. noted that 45 percent of cases of pilomatricoma were incorrectly diagnosed by fine needle aspiration cytology based on their review of multiple case reports and series. Nevertheless, in their study as well as other more recent studies, fine needle aspiration has been found to be quite accurate when two key components, basaloid cells and ghost cells, are visualized, as this has been found to be specific for pilomatricoma [12,13,14].

A rare malignant counterpart, pilomatrix carcinoma, has been described, and approximately 90 cases have been reported in the literature. It is locally aggressive and can recur. In several cases, it has demonstrated metastases. Many key features are similar between these benign and malignant counterparts; the primary differentiating characteristics include a high mitotic rate with atypical mitoses, central necrosis, infiltration of the skin and soft tissue, and invasion of blood and lymphatic vessels [15,16].

SUMMARY:

In summary, pilomatricoma is a benign skin neoplasm that usually presents as a single, slow-growing subcutaneous or intradermal nodule. It is usually less than 3 cm and commonly affects the face. Malignant transformation has been reported but it is very rare. The diagnosis is established based on histopathology. The treatment of choice is surgical excision with a clear margin with every effort to preserve the branches of the facial nerve. Recurrence with adequate treatment is uncommon.

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