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Pilomatrixoma (Calcifing Epithelioma of Malberbe), Case Study

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Abstract: Pilomatrixoma, also known as calcifying epithelioma of Malberbe, is a rare, benign skin tumor derived from hair follicle matrix cells. It predominantly affects young children but can occur across all age groups, with a slight female preponderance. This study presents the case of a 7-year-old female with a gradually enlarging mass on the left arm, diagnosed as pilomatrixoma following surgical excision and histopathological analysis. Pilomatrixoma typically manifests as a solitary, firm, skin-colored or purplish lesion, most commonly on the head, neck, or upper extremities. It is characterized by calcification within the lesion, often presenting the "tent sign," and is generally asymptomatic but can occasionally cause tenderness. Dermoscopy, imaging (e.g., ultrasound, X-ray), and biopsy are critical diagnostic tools, with histological findings revealing basophilic cells, shadow cells, and calcium deposits. The lesion in this case was excised surgically, a treatment that remains the standard to confirm diagnosis, alleviate cosmetic concerns, and prevent rare complications like malignant transformation. Although rare, pilomatrixoma may recur or become pilomatrix carcinoma. This research underscores the importance of early diagnosis and treatment of pilomatrixoma for favorable outcomes.

Keywords: Calcifing Epithelioma, leukocyte antigen, HLA-DRB1.

INTRODUCTION

The skin is the largest organ in the body, covering its entire external surface. The skin has 3 layers—the epidermis, dermis, and hypodermis, which have different anatomical structures and functions. The skin's structure comprises an intricate network that serves as the body's initial barrier against pathogens, ultraviolet (UV) light, chemicals, and mechanical injury. This organ also regulates temperature and the amount of water released into the environment. Skin thickness varies by body region and is influenced by the thickness of the epidermal and dermal layers. Hairless skin in the palms of the hands and soles of the feet is the thickest due to the presence of the stratum lucidum, an extra layer in the epidermis. Regions lacking this extra layer are considered thin skin. Of these regions, the back has the thickest skin because it has a thick epidermis.The skin's barrier function makes it susceptible to various inflammatory and infectious conditions. In addition, wound healing, sensory changes, and cosmesis are significant surgical concerns. Understanding the skin's anatomy and function is crucial for managing conditions across all medical fields (1). Adnexal structures such as hair follicles, sebaceous and sweat glands span both the epidermal and dermal layers and contain some keratinocytes in their ducts. In injuries where epidermis is lost, re-epithelialisation occurs from these structures as well as from the wound margins. The body can be envisaged as three-dimensional segments of tissue called angiosomes, each with an arterial supply and a venous drainage. Blood equilibrates and flows between neighbouring angiosomes via 'choke' vessels, which tend to be situated within muscles. Cutaneous arteries, direct branches of segmental arteries concentrated at the dorsoventral axes and intermuscular septae, perforate the underlying muscles or run directly within fascial layers to the skin from the deep tissues. The blood supply to the skin anastomoses in subfascial, fascial, subdermal, dermal and subepidermal plexi. The epidermis contains no blood vessels so cells there derive nourishment by diffusion. The venous drainage of the skin is via both valved and unvalved veins. Unvalved veins allow oscillating flow in the subdermal plexus between cutaneous territories, equilibrating flow and pressure. The valved cutaneous veins drain via plexi to the deep veins.

Function of the Skin

- Barrier to the environment enveloping the body and protecting against trauma, radiation and pathogens.
- Regulates temperature and water homeostasis.
- Organ of excretion for urea, sodium chloride, potassium and water, as well as sulphur-containing metabolites from drugs (e.g. dimethyl sulphoxide) or food (garlic, cumin).
- The skin has significant endocrine and metabolic functions and interactions.
- Skin cells contain receptors for and respond to: peptides, steroid sex hormones, thyroid

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hormones and neurotransmitters and they both produce (cholecalciferol) and metabolise (androgens) hormones and precursors to activate, potentiate and inactivate their functions.

- Sensory organ with multiple receptors for pain, pressure and movement (2).

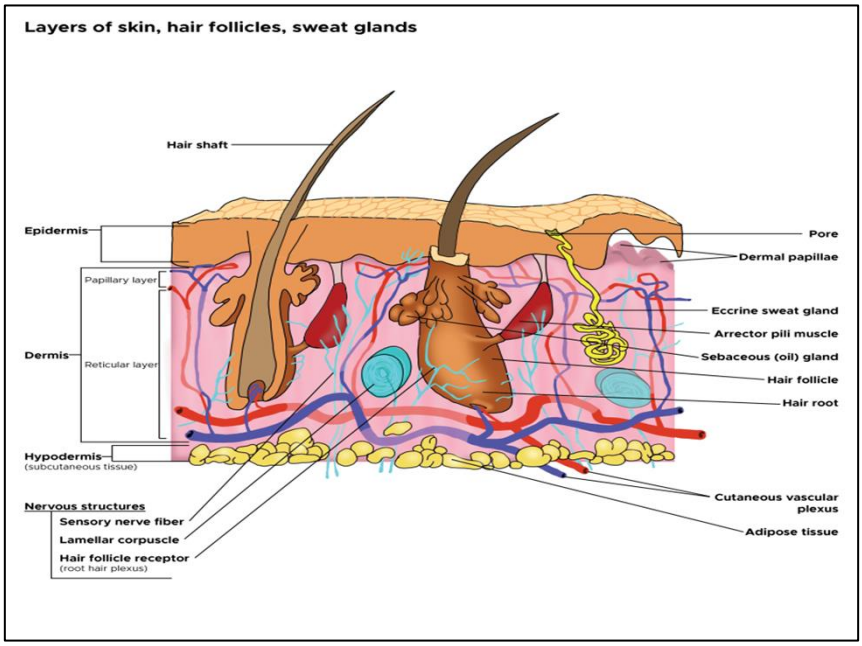

Fig 1: Cross Section, Layers of the Skin. This is a cross-section view of the hair follicles, hair roots and shafts, sweat glands, pores, epidermis, dermis, and hypodermis. The papillary and reticular layers are also included. The eccrine sweat gland is located.

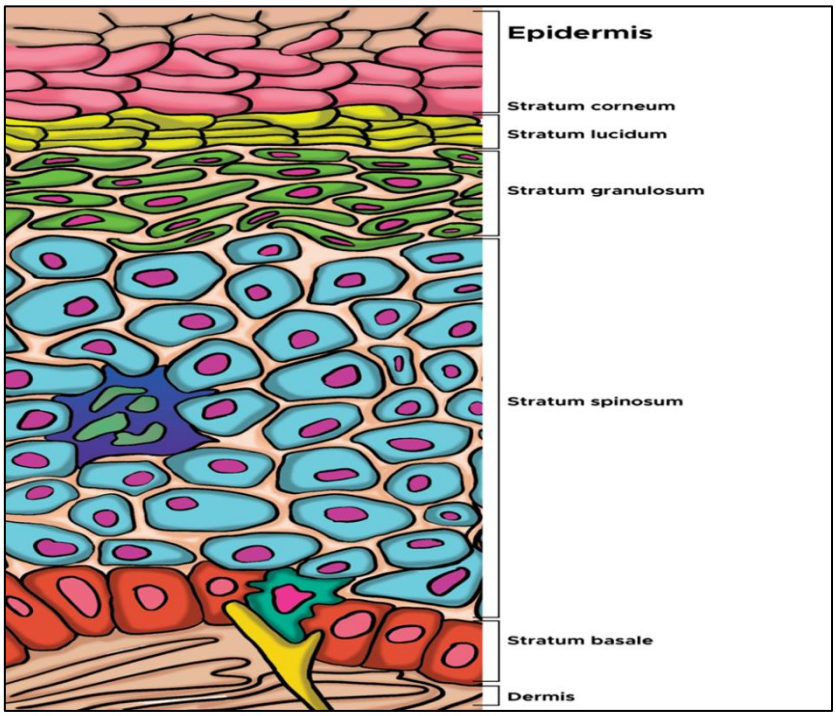

Fig 2: Cells of the Epidermis. The image shows stratum corneum, stratum lucidum, stratum granulosum, stratum spinosum, stratum basale, and dermis.

PATIENT AND METHODS

Seven years old female childe presented with mass in the left arm at lateral aspect since years which increased in size gradually and slowly, surgical excision was done for her under general anesthesia, and the patient was in good condition postoperatively, fig 3.

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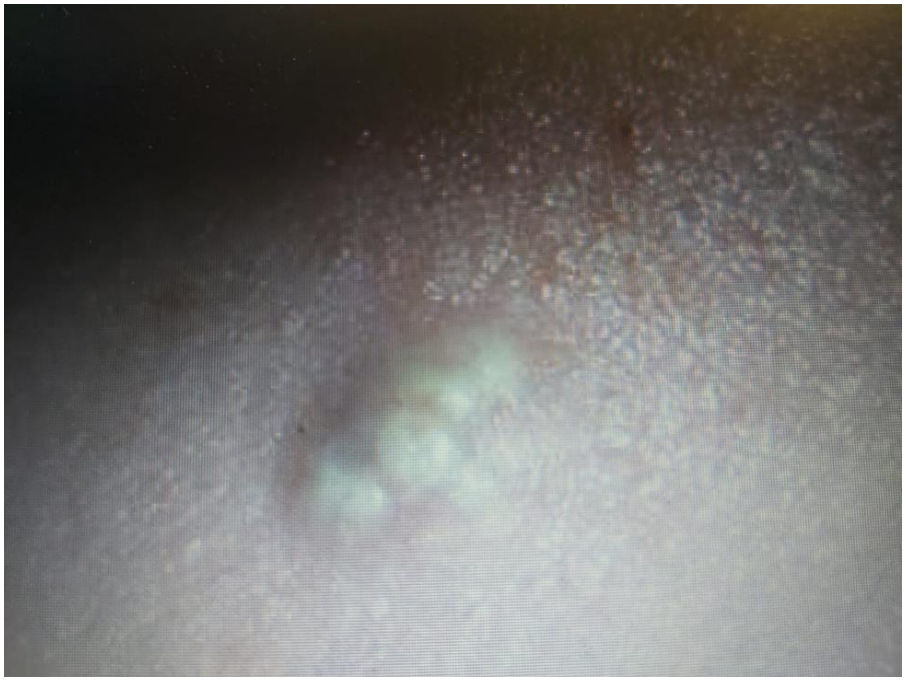

Fig 3: the lesion.

RESULT

- Gross section:- Single piece of tissue measuring 2×3×4 CM
- Cut section:-Revealed grey- white firm mass measuring 1.5 CM× 1CM
- Microscopical:- Section shows features consistent with benign skin adnexial tumour and consistent with PILOMATRIXOMA (Calcifing Epithelioma Of Malberbe).

DISCUSSION

Pilomatricoma is an uncommon, harmless, hair follicle benign tumour derived from hair matrix cells. It is also spelled 'pilomatrixoma', and sometimes known as 'calcifying epithelioma of Malherbe'. Pilomatrix carcinoma is a rare condition. A pilomatrixoma is often a single lump. But sometimes there may be more than one. Multiple pilomatrixomas have been observed, mainly in association with myotonic dystrophy, but also with other syndromes including Rubenstein-Taybi syndrome and constitutional mismatch repair deficiency (CMMRD) syndrome.. One cause is mutation of gen CTNNB1. Pilomatricoma is most often diagnosed in young children but may also affect adults. It appears to be slightly more common in females than males. pilomatricoma is now known to be due to a localised mutation in a hair matrix cell. An overactive proto-oncogene called BCL-2 suggests the normal process of cell death is suppressed, and mutations in CTNNB1 in most cases suggest loss of regulation of a protein complex called beta-catenin/LEF-1. Pilomatricoma presents as a single skin-coloured or purplish lesion.

- Usually found on the head and neck, but they may occur on any site.
- Don't usually cause any symptoms, but they may be tender.
- May be skin coloured, white or red.
- May be regular or irregular in shape.
- Most pilomatricomas are 5–10 mm in diameter.
- May remain stable for years or slowly grow in size up to several centimetres in diameter.
- Is characterised by calcification within the lesion, which makes it feel hard and bony, and often results in an angulated shape, 'tent' sign.

Complications of pilomatricoma are rare. However, occasionally they grow to giant size (several centimetres in diameter), and pilomatrix carcinoma (cancer) has been very rarely reported. If pilomatricoma is suspected, dermoscopy may be helpful, showing a central whitish or grayish-blue structureless area. Erythema and telangiectasia are sometimes observed. If the nature of the skin lesions is uncertain, ultrasound scan may be recommended. The scan of pilomatricoma is described as a doughnut within the dermis (mid layer of the skin) with a tail (the tail denotes calcification). The calcification may be detected by X-ray. Biopsy will help to establish the cause of the lesion. Alternatively the whole lesion can be removed, providing both diagnosis and treatment. The histology of pilomatricoma is striking.

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It may show a sharply demarcated tumour surrounded by a fibrous capsule or a poorly demarcated tumour without capsule. There are darkly stained 'basophilic' cells and 'shadow' cells with missing nuclei. Calcium deposits are found in most lesions.

Here are the most common symptoms of a pilomatrixoma:

- A small, hard lump beneath the skin, often on the face or neck
- The skin covering the lump looks normal or may have a blue color
- The lump is often painless, unless it becomes infected.

The symptoms of a pilomatrixoma can seem like other health conditions. Make sure your child sees their healthcare provider for a diagnosis. A pilomatrixoma is often diagnosed based on how it looks and feels. Diagnostic tests may include:

- Biopsy. A tissue sample is taken and looked at under a microscope.
- Imaging. An X-ray, ultrasound, CT scan, or MRI may be done.

Treatment may include surgery to cut out the lump. Surgery may be done if the pilomatrixoma is uncomfortable, is cosmetically bothersome, or to confirm the diagnosis. Most children don't face any serious complications. But pilomatrixomas can become cancer in rare cases. Pilomatrixomas can also come back after they are removed. Surgery to remove the lesion will result in a scar.

CONCLUSION

- A pilomatrixoma is a slow-growing, noncancerous (benign) skin tumor of the hair follicle.
- It is a small, hard lump beneath the skin. It's most often on the face and neck.
- The lump is often painless, unless it becomes infected.
- It's often diagnosed based on how it looks and feels, But a biopsy and imaging tests may also be done.
- Treatment includes surgery to remove the lump.

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