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Hide and Seek of skin diseases: A Case Study

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ABSTRACT

Kyrle's disease is a rare acquired perforating dermatosis occurring in adults and is common in females (6:1). It is usually associated with Diabetes mellitus and Chronic renal disease. It typically presents as hyperkeratotic papules and nodules with keratotic plug on the lower extremities. Here we are presenting 47 year old male with kyrle's disease because of its rarity. Sebaceous cysts are commonly encountered in clinical practice. But all these cysts are not sebaceous cysts. There are several mimickers of sebaceous cysts which has to be differentiated histopathologically so that appropriate treatment and follow up can be advised. This article highlights diagnostic importance of sebaceous cysts and we are presenting 5 cases of sebaceous cysts mimickers – Eccrine hidradenoma, Malignant eccrine poroma, Calcinosis cutis, Chondroidsyringoma and fungal infection. Hyperpigmented skin lesions are extremely common and it has many causes. It can be due to melanocytic lesions, keratinocytic lesions, reactive lesions. In this article, we are presenting 4 cases of hyperpigmented vascular lesions – Blue rubber bleb nevus, 2 cases of pyogenic granuloma and Angiokeratoma. This is to consider vascular lesions as differential diagnosis while evaluating hyperpigmented skin lesions. Commonest skin lesions like epidermoid cyst and hyperpigmented skin lesions should be properly evaluated and are subjected to histopathological evaluation for accurate diagnosis so that appropriate treatment can be provided.Kyrle's disease is presented for its rarity.

Keywords: Kyrle's disease, Eccrinehidradenoma, Malignant eccrine poroma, Calcinosis cutis, Chondroidsyringoma, Blue rubber bleb nevus &Angiokeratoma.

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INTRODUCTION

The perforating dermatoses represent a group of skin disorders characterized by the "perforation" or elimination of dermal connective tissue through the epidermis. Perforating disorders may be divided into primary and secondary disorders. Kyrle disease is a rare acquired perforating dermatosis occurring in adults are mostly seen in setting of diabetes, chronic renal failure, and hyperuricemia [3]. Sebaceous cysts are commonly encountered in clinical practice. But all these cysts are not sebaceous cysts. There are several mimickers of sebaceous cysts which has to be differentiated histopathologically so that appropriate treatment and follow up can be advised. This article highlights diagnostic importance of sebaceous cysts. Hyperpigmented skin lesions are extremely common and it has many causes. It can be due to melanocytic lesions, keratinocytic lesions, reactive lesions.The present study emphasizes the importance of exploration by histopathological examination in common skin lesions.

MATERIAL AND METHODS

This is a case series study. All gross specimens were received in 10% formalin in the department of Pathology,Vinayaka Mission's KirupanandaVariyar Medical College& Hospitals, Salem. Tissue is processed by standard operating procedure and stained by hematoxylin and eosin. Special stain (PAS) is done wherever necessary.

RESULTS AND DISCUSSION

Case Reports

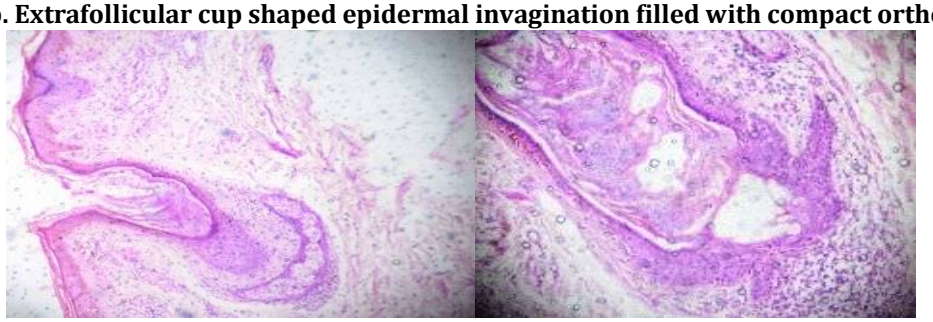
Case 1: 47 year old male with c/o itchy skin lesion all over the body for one year. O/E – Multiple hyperpigmented, hyperkeratotic papules seen over both LL. Diagnosis : ? PrurigoNodularis/ ?Perforating folliculitis.Shave Biopsy done.

Gross: Skin with soft tissue bits measuring 0.5cm in diameter in aggregate

Microscopy: Sections studied show epidermal hyperplasia, focal extrafollicular cup shaped epidermal invagination filled with compact orthokeratosis with changes of parakeratosis, dyskeratosis, basophilic degenerative material.

Dermis shows lymphohistiocytic infiltration. Diagnosed as Kyrle's disease.

Fig-1a & 1b. Extrafollicular cup shaped epidermal invagination filled with compact orthokeratosis

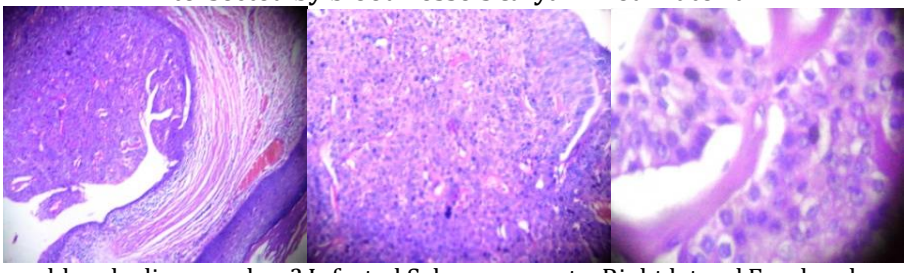


Case 2:Clinical details: Swelling Forearm – Infected Sebaceous cyst. **Gross:** Cystic soft tissue measuring 4 x 3 x 1.5cm with overlying skin ellipse measuring 4 x1.5cm.

C/S: Unilocular cyst with inner wall showing focal papillary excrescences along with thickened area and tiny cystic cavity measuring 2 mm filled with keratinous material.

Microscopy: Sections studied show cystic lesion partly lined by single to multilayered cuboidal epithelium with foci of nodular collection of tumor cells intersected by blood vessels &hyalinized material. Along with this cystic cavity lined by stratified squamous epithelium with distinct granular layer filled with keratinous material is noted. Diagnosed as Eccrine Hidradenoma with cystic change with Epidermal inclusion cyst.

Fig. 2a, 2b, 2c. Single to multilayered cuboidal epithelium with nodular collection of tumor cells intersected by blood vessels &hyalinized material

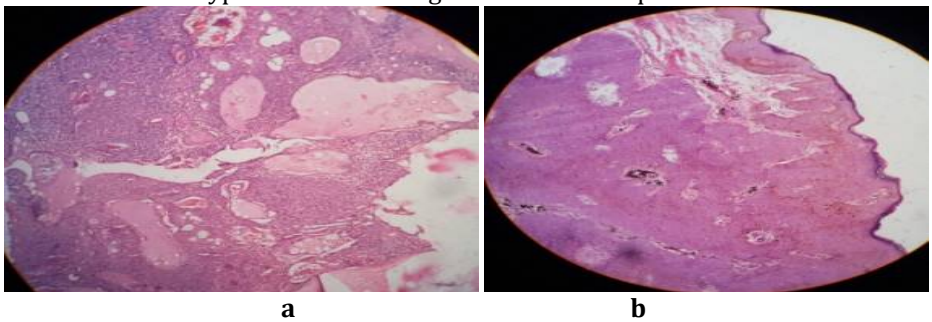


Case 3: 70 year old male diagnosed as ? Infected Sebaceous cyst – Right lateral Forehead.

Gross: Received specimen as multiple pieces, one of which is covered by skin, largest measuring 5 x 1 x 1cm, smallest measuring 0.6 x 0.5 x 0.2cm.

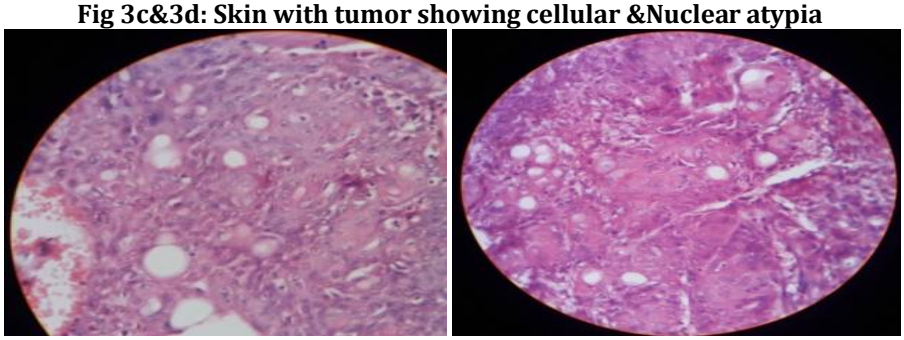
C/S: Grey white with slit like spaces.

Microscopy: Ductal lumen, cystic spaces filled with eosinophilic secretions, Extensive squamous differentiation with Nuclear atypia was noted. Diagnosed as Eccrine porocarcinoma**.**

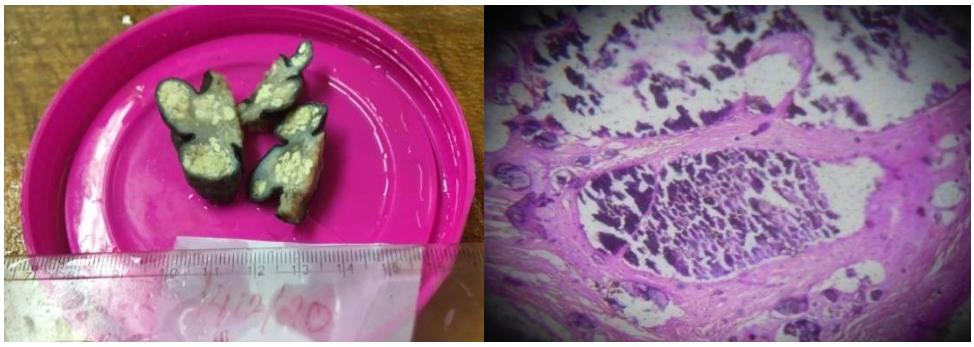


a **b**

Fig 3 (a and b) : Ductal lumen, cystic spaces filled with eosinophilic secretions.
Fig 3c&3d: Skin with tumor showing cellular &Nuclear atypia



Case 4: 35 year old male with papillomatous growth in the inter-gluteal cleft, diagnosed as ?Sebaceous cyst / Papilloma.
Gross: Soft tissue with skin measuring 4 x 3.5 x 2cm. Multiple skin colored nodules ranging in size from 0.5 to 2.5cm in diameter.
C/S: Multiple **s**olid yellow white foci were seen.
Microscopy: Sections studied show tissue lined by stratified squamous epithelium. Underlying dermis shows multiple islands of calcified nodules composed of basophilic material surrounded by mononuclear inflammatory cells, histiocytes& multinucleated giant cells.
Diagnosed as **Calcinosis cutis.**



a **b**

Fig 4 (a) Multiple solid chalky yellowish white areas; (b) Nodules composed of basophilic material

Case 5: 38 year old female with c/o swelling in right thigh since 2 years. Diagnosed as Infected sebaceous cyst.
Gross: Single grey brown soft tissue mass measuring 3.5 x 2 x 1cm with strip of skin measuring 4x 1cm.
C/S: Grey brown, reddish & grey white areas noted.
Microscopy: Section studied show dermal tumour composed of sheets, ducts, clusters of cuboidal epithelial cells. Cells are embedded in the chondromyxoid stroma. Diagnosed as chondroidsyringoma**.**

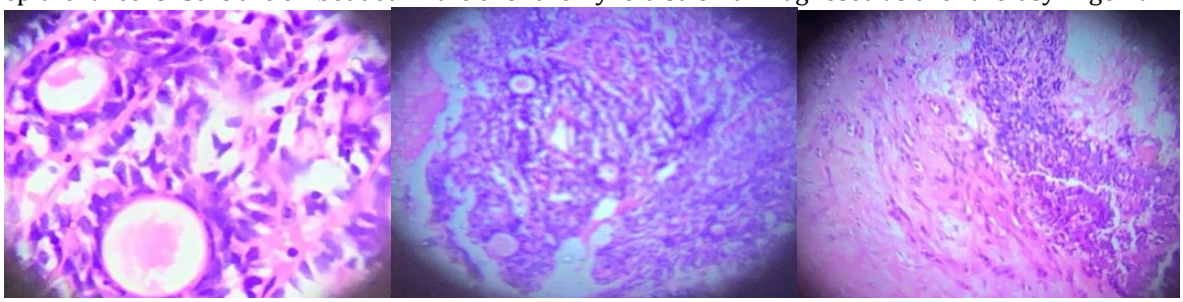
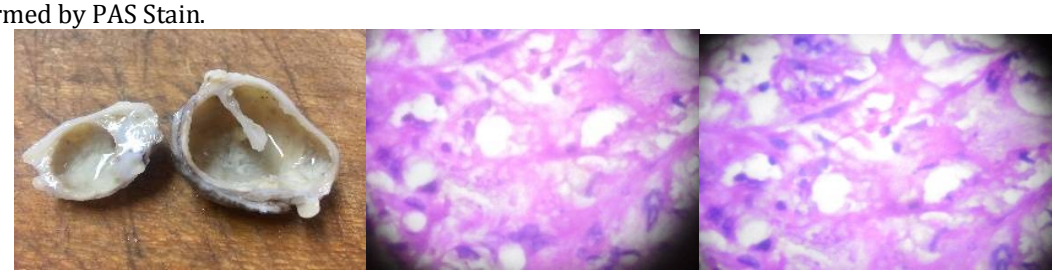


Fig 5 Sheets, ducts, clusters of cuboidal epithelial cells against chondromyxoid stroma

Case 6:75 years female with painful swelling in left leg, clinically diagnosed as Sebaceous cyst.
Gross: Single cystic mass measuring 4 x 3 x 3cm with overlying skin measuring 3.5 x 1cm.
C/S: Multiloculated cyst with presence of solid area and creamy material is noted.
Microscopy: Section studied show skin with epidermis &dermis. Underlying stroma shows a cystic lesion with its wall lined by granulation tissue with giant cell reaction composed of lymphocytes, multinucleated giant cells & newly formed blood vessels. Along with this are seen few septate & branching fungal hyphae

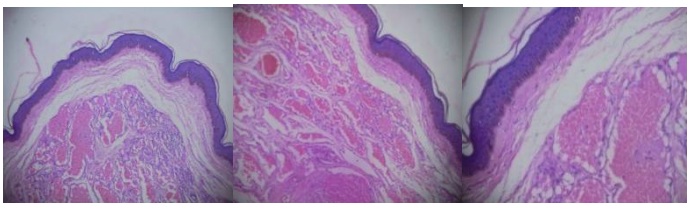
admixed with areas of necrosis. Diagnosed as Fungal abscess (Phaeomycosis) with giant cell reaction. Confirmed by PAS Stain.



a **b** **c**

Fig 6: C/S – Cystic lesion, HPE – Necrosis with fungal hyphae.

Case 7: 40 year old male with complaints of asymptomatic hyperpigmented lesions over trunk & face since 10 years.
O/E : Multiple skin colored, reddish blue plaques present over lower back, face with few pedunculated plaques over back & thigh.
Clinical Diagnosis: ?Pyogenic granuloma/?BRBNS/?Kimura disease/?Glomangioma. Punch biopsy done.
Gross: A Single grey white soft tissue bit with skin measuring 0.5cm in diameter.
Microscopy: Tissue with its surface lined by stratified squamous epithelium with hyperkeratosis and diffuse thinning. Superficial dermis showed presence of irregular vascular space containing RBC's & fibrinous material. The spaces were lined by single layer of thin endothelial cells. HPE features were suggestive of **Blue**
Bleb Rubber syndrome.



a **b** **c**

Fig 7: Irregular vascular spaces containing RBC's & fibrinous material (a,b&c)

Case 8 :21year old male with complaints of skin lesion over the trunk & genital region since 5 years.
O/E : Multiple erythematous to hyperpigmented papules over lower back, around umbilicus, scrotum & both the thighs were seen.
Diagnosis : ? Vascular tumor. Punch biopsy done.
Gross : Single grey white soft tissue bit with skin measuring 0.5cm in diameter.
Microcopy: Epidermis with orthokeratosis, focal elongation of the rete ridges. Dilated, thin walled, congested capillaries were seen in the papillary dermis. Based on the clinical findings & HPE features the case was diagnosed as Angiokeratoma Corporis Diffusum**.**

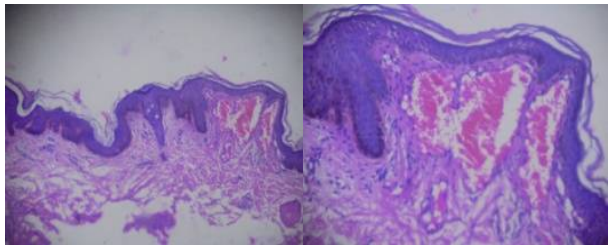
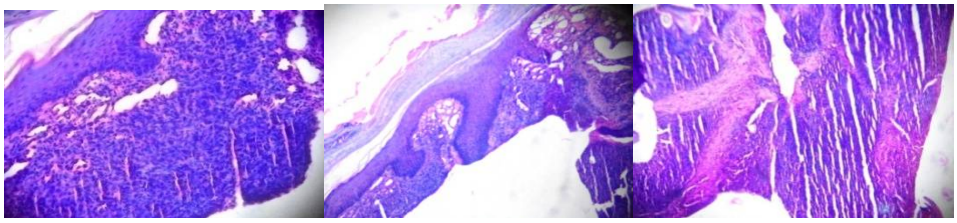


Fig 8 (a and b) : Orthokeratosis, elongated rate ridges and dilated vessels

Case 9: 8 years old male with complaints of raised skin lesion over left side of scalp for past 2 months. Lesion was initially small, then gradually increased in size.
O/E: Well defined pedunculated plaque was seen over left retroauricular area which bleeds on touch.
Gross: Single grey white soft tissue measuring 0.6x0.3cm.
Microscopy: Section studied shows epidermis with hyperkeratosis and Parakeratosis with presence of numerous vascular spaces lined by endothelial cells and proliferation of fibroblasts in the dermis. Diagnosed as Pyogenic granuloma.



a **b** **c**

Fig 9 (a, b, c) : Hyperkeratosis, parakeratosis, vascular spaces lined by endothelial cells

Case 10: 33 year old female with complaints of Painful skin lesions with bleeding over the left arm for the past two months.
O/E: Single pedunculated / lobulated erythematous mass of size, 1x1cm was present over extensor aspect of left arm. Shave biopsy was done.
Gross: Single Grey white soft tissue bit measuring 0.5x0.3x0.2cm.
Microscopy: Section studied shows epidermis with dermis. Dermis shows lobular arrangement of small to medium sized capillaries, lined by single layer of flat to plump endothelial cells separated by fibromyxoid stroma. Diagnosed as Pyogenic granuloma**.**

DISCUSSION

Kyrle's disease is a rare acquired perforating dermatosis occurring in adults and is common in females (6:1). It is usually associated with Diabetes mellitus and Chronic renal disease[1].It typically presents as hyperkeratotic papules and nodules with keratotic plug on the lower extremities. In our case, it is seen in male with acquired perforating dermatoses with no systemic association is also noted. Nodular hidradenomas are benign adnexal tumors that usually present as painless solitary nodules mimicking epidermoid cysts. Head and neck is most commonly involved. In our case it is seen in forearm, These neoplasms occur primarily in adults and in females. The recurrence rate is around 10 percent[2].Malignancy can arise from these lesions. Eccrine Poromas are benign adnexal neoplasms that arise from terminal portions of sweat gland duct. Poromas occur predominantly on the soles and palms and clinically present as painful nodule. Eccrine poromas (20%) can transform to malignancy[3](eccrine porocarcinomas). The diagnosis of these tumors on a clinical basis is very difficult. Hence to diagnose and to identify malignant transformation histopathological examination is utmost important.Calcinosis cutis is a condition in which calcium deposits occur in the skin. The etiology may be idiopathic or due to abnormal calcium or phosphorus metabolism It usually present as firm nodular lesion involving the skin that may mimic sebaceous cyst. Chondroidsyringoma is a benigntumor of the sweat gland that resembles pleomorphic adenoma (mixed tumor) of the salivary gland. It is usually seen in adults with male predominance[4].Chondroidsyringoma with cystic change can clinically resembles sebaceous cyst. In our case there is no malignancy. Cutaneous fungal infections can present as cystic lesion resembling sebaceous cyst[5].In these situations, microscopic examination is mandatory for proper diagnosis.
Blue rubber bleb nevus syndrome (Bean syndrome) is a rare angiomatous condition with an estimated incidence of 1:14000 births, in which venous malformations occur on the skin, soft tissue and GI tract[6]. Clinically they appear as multiple hyperpigmented painful lesion. It is an important condition due to the potential for significant bleeding which can be fatal, but our case presented as an asymptomatic lesion. Angiokeratoma corporis diffusum (ACD) is an asymptomatic benign vascular lesion that can occur localized or generalized. It presents as red to brown papule or nodule with verrucous surface in healthy individuals or in persons with enzyme deficiency[7]. However, our patient was a healthy individual with generalized pattern of lesion distribution.Pyogenic granuloma or lobular capillary haemangioma commonly present as elevated, dark red lesion with ulceration involving the skin and mucousmembranes [8]. It composed of 3 phases, in our case it is a cellular phase, the other cases it is mostly vascular phase. Hence vascular lesions serve as important differential diagnosis in the evaluation of hyper pigmented skin lesions.

CONCLUSION

Kyrle disease is presented because of its rarity. Commonest skin lesions like epidermoid cyst and hyperpigmented skin lesions should be properly evaluated and are subjected to histopathological evaluation for accurate diagnosis so that appropriate treatment can be provided.Clinicians must be aware of this condition and should also rule out malignant chondroidsyringoma. A careful histopathologic examination is the key to chondroidsyringoma.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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