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# Ectopic conditions in dermatology

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## I. Introduction

Most dermatoses have their origin within the skin or skin appendages presenting with characteristic clinical features. . Skin may be affected in many systemic conditions too because of alteration in the skin structure and/or physiology resulting from the common aetiopathogenesis. Very occasionally, we encounter a fewconditions which present within the skin but the pathology is not indigenous to that site. Such anomalous, non-indigenous presence of any condition within the skin is called ectopicwhich pose a difficulty in theirdiagnosis and management. A high index of suspicion and awareness of theectopic dermatologicconditions are indispensable pre-requisites.After thorough searching through standard dermatology textbooks and databases of pubmed, Scopus, Medline and Index medicus, we have compiled a list of such intriguing ectopic conditions. They have been classified based on their aetiologies(table 1) and their contents (table 2).

1. Cutaneous calcification: Deposition of calcium salts or calcification in skin can occur in various conditions. Broadly, it can be either calcinosis cutis (deposition of calcium phosphate) or osteoma cutis (deposition of calcium hydroxyapatite along with osteoblasts and osteoclasts). Calcinosis cutis has 4 main types: dystrophic, metastatic, idiopathic and iatrogenic.[1] Dystrophic form is commonly seen following trauma, connective tissue diseases such as systemic sclerosis and dermatomyositis and genetic diseases e.g., pseudoxanthoma elasticum and Ehler-Danlos syndrome. Hyperparathyroidism, renal failure and sarcoidosis are few causes of metastatic calcification. Idiopathic scrotal calcinosis also exists [**Figure 1**]. Osteoma cutis due to membranous ossification in dermis without cartilage which can be primary or idiopathic and secondary usually associated with chronic inflammatory diseases. Morphologically, idiopathic type may have 4 presentations: isolated, plate-like, widespread and multiple miliary osteoma cutis.[1]
2. Cutaneous endometriosis: The presence of endometrial tissues in skin accounts for lessthan 1% of all cases of endometriosis. Cutaneous endometriosis can be primary (without any prior surgical history) or secondary (due to iatrogenic implantation of endometrial cells after surgery. Umbilicus is the most common site of primarycutaneous endometriosis, known as Villar's nodule.[2] Vascular or lymphatic migration were proposed for primary forms.
3. Cutaneous meningioma: It is characterized by meningothelial elements present in the skin or subcutaneous layer and also known as Meningeal heterotopia, rudimentary meningoceleand meningeal hamartoma.It typically presents as a skin-colored nodule on the scalp over the midline, sometimes surrounded by a ring of long, dark, coarse hair (hair collar sign) and along the spine.[3]
4. Deep apocrine glands in Nevus sebaceous: In this condition, sebaceous glands are large and hyperplastic along with lack of terminal hair follicles and epidermal hyperplasia.[4] Ectopic apocrine glands with their dilated ducts are often located deep in the dermis.
5. Ectopic breast tissue:Breast develop from two mammary ridges or milk lineswhich extend from the anterior axillary folds to inguinal folds.Normally, all themammary ridges involute except in the pectoral region to form breasts. Persistence of any mammary ridge along the milk lineslead to ectopic breast tissue, also known as supernumerary or accessory breast.[5] It may have both glandular tissue and nipple-areola complex, called as polythelia. Polymastia term is used if there is only glandular tissue. In 2/3$^{rd}$ cases, it is found just below the inframammary crease. Axillary ectopic breast has about 20% incidence.Occasionally, fibroadenoma, carcinoma or phyllodes tumor may develop within ectopic breast.
6. Ectopic desmoglein 3 expression:Paraneoplastic pemphigus (PNP) is an autoimmune bullous disease of the skin and mucous membranes mediated by the IgG autoantibodies which target the desmoglein 3 (Dsg 3) and plakins.Interestingly, ectopic expression of Dsg3 has been reported in respiratory epithelia which may be a reason of significant pulmonary involvement in the form of bronchiolitis obliterans in PNP.[6]Any preceding lung injury can also lead to such ectopic expression of Dsg3.
7. Ectopic hair: In 'music box spine keratoderma', an idiopathic autosomal dominant keratinization disorder, spiny keratotic papules develop on margin of palms and soles, which represent ectopic hairs due to hair type keratinization.[7]
8. Ectopic hidradenitis suppurativa: Hidradenitis suppurativa (HS)is mainly seen in the flexures rich in apocrine glands. Humidity and friction are local predisposing factors. Rarely, it has been reported in areas

devoid of apocrine glands such as thigh, chest, abdomen, amputated limb, dorsal foot, eyelids, knee and scalp.[8]Knowledge of such presentations of HS is essential to avoid the misdiagnosis.

9. Ectopic nails: It is also called as onychoheterotopia which shows nail growth outside the digit's nail unit. It is mostly seen on the palmar aspect of 5th digit in its congenital form, sometimes being associated with Pierre Robin syndrome.[9]In acquired form, trauma may result in ectopic nail growth, commonly on the dorsal aspect of the digit**[Figure 2] [Figure 3**].
10. Ectopic teeth: Schinzel syndrome or ulnar-mammary syndrome is a rare inherited disorder characterized by bony abnormalities of upper limb and spine and apocrine glands dysfunction. In some cases, it may have ectopic upper canines.[10]Second example is Treacher Collins syndrome which has many dental anomalies including ectopic maxillary canine and 1st molar.[11]Other characteristic facial features are convex facies, prominent nose and retruded chin, downslanting palpebral fissures, upper eyelid coloboma, absence of the lower eyelashes, external ear malformations, macrostomia and cleft palate.
11. Ectopic-extramammary Paget's disease (E-EMPD): It is a very rare ectopic apocrine gland neoplasm affecting areas that usually lack apocrine glands such as the chest, abdomen, thigh, eyelids, face, and external auditory canal. Aberrant transformation of stem cells to abnormal keratinocytes with apocrine markers at those locations may be the reason. Intraepidermal Paget's cells and dermal lymphocytic infiltrates are the typical findings similar to the EMPD.[12]
12. Fibrofolliculoma/trichodiscoma: These are benign, folliculosebaceous neoplasms of perifollicular mesenchymal tissues which mostly present as slightly raised, round or oval, skin colored, dome shaped papules on head and neck. Syndromic form is more common (Birt-Hogg-Dube syndrome) in which multiple lesions are seen. Solitary, non-syndromic forms are rare. The typical histopathology shows central acanthotic, dilated infundibulum containing keratins (with no hair shaft formation) surrounded by fibroepithelial proliferations and multiple radiating, thin, anastomosing bands of basaloid ceils embedded in the mucinous stroma. Of note, mature adipocytes are often seen in the epithelial cords**[Figure 4]**.[13]Folliculosebaceous cystic hamartoma also shows dermal adipocytes.
13. Fordyce spots: These are ectopic sebaceous glands commonly seen as asymptomatic multiple, discrete, barely elevated, yellowish papules around the vermilion border of the lips and within the oral mucosa [**Figure 5**].Occasionally, it can be extensive requiring treatment.Some authors prefer to make the diagnosis of epidermisation of lip instead of Fordyce spots in some cases. Based on the dermoscopic examination, epidermisation of lip is better to be considered as closely set Fordyce spots.[14]
14. Hidradenoma papilliferum: It is a benign tumor of apocrine gland exclusively seen in females. It has also been described as ectopic forms on other sites such as head and neck, chest, and extremities which is attributed to the presence of heterotrophic and specialized version of apocrine glands in those areas.[15]
15. Nephrogenic rest: These are considered precursor of Wilms' tumor. It is usually seen as perilobar and intralobar lesions in the kidney. Rarely, it can be present in lumbosacral and sacrococcygeal region (frequently associated with spinal dysraphism), inguinal canal, and testis. In lumbosacral region, it may present as a soft polypoid mass covered by normal skin.[16]
16. Lingual thyroid:The ectopic presence of thyroid gland at the base of tongue is the result of failure of the normal descent of thyroid gland along a path from foramen cecum in the tongue to the final position.[17]It usually remains asymptomatic but may enlarge and cause dysphagia or dyspnea. Hypothyroidism is a common problem in this condition.
17. Nasal glioma: Due to abnormal closure of fonticulus nasofrontalis, ectopic rest of neuroglial tissue are left extracnially. It is mostly present on or within the nose as a round, firm papule. Other less common sites are face, scalp, lip, tongue, oropharynx,nasopharynx and orbit.[18]Intracranial connection is rare unlike an encephalocele.
18. Nevus lipomatosus cutaneous superficialis (NLCS): The presence of mature ectopic adipocytes in the dermis is the hallmark of this entity. The onset is at birth or in early childhood. Exact cause is unknown but there are multiple hypotheses such as adipose metaplasia in the course of dermal degenerative changes, developmental displacement of adipose tissues or adipogenesis from perivascular mononuclear cells differentiating into lipoblasts [**Figure 6**]. Clinically, it can be multiple, grouped skin-colored, pedunculated nodules or solitary dome-shaped sessile papule or nodule.[19]
19. Parameatal cysts:It is considered as embryological anomaly that is likely thought to develop from ectopic urethral mucosa or ectopic periurethral Littre's glands. It is locatedventrallyin midline anywhere from the urethral meatus to theperineum [**Figure 7**]. The mechanism is not clearly understood. Following hypotheses have been proposed about its origin: (1) the persistence of cystic spaces, during the process of preputial separation from glans, (2) urethral epithelial remnants left secondary to incomplete fusion of median raphe, (3) outgrowths of urothelium after primary closure of the raphe, (4) ectopic periurethral glands of Littreand/or (5) from blockage of paraurethral ducts which may occur secondary to infection or trauma in some cases.[20]

20. Umbilical polyp: It is a congenital ectopic remnant of intestinal mucosa at umbilicus due to incomplete distal closure of the omphalomesenteric duct, characterized by the presence of a firm, reddish, half-moon shaped, discharging polypoid lesion [**Figure 8**].[21]

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## Legends:

Figure 1:multiple scrotal calcified nodules
Figure 2:fully formed ectopic nail on dorsal aspect
Figure 3: partially formed ectopic nail
Figure 4:Fibrofolliculoma showing mature adipocytes in dermis
Figure 5:Fordyce spots on upper lip
Figure 6: Mature dermal adipocytes in Nevus lipomatosus cutaneous superficialis
Figure 7: Parameatal cyst
Figure 8:Umbilical polyp

**Table 1: Aetiological classification of ectopic cutaneous conditions**

| Idiopathic | Developmental origin | | | | Secondary to trauma, inflammatory or |
|---|---|---|---|---|---|
| | Endoder | Mesodermal | Ectodermal | Multidermal | |

| | mal | | | | | neoplastic conditions |
|---|---|---|---|---|---|---|
| Primary osteoma cutis<br><br>Idiopathic calcinosis cutis<br><br>Primary cutaneous endometriosis<br><br>Ectopic hair<br><br>Ectopic hidradenitis suppurativa | Umbilical polyp<br><br>Parameatal urethral cyst | Lingual thyroid<br><br>Subcutaneous nephrogenic rest<br><br>Nevus lipomatosus cutaneous superficialis | Surface ectoderm<br>Ectopic teeth<br><br>Ectopic nail<br><br>Deep apocrine glands in Nevus sebaceous<br><br>Fordyce spots<br><br>Ectopic EMPD<br><br>Ectopic hidradenoma papilliferum | Neuro-ectoderm<br>Cutaneous meningioma<br><br>Nasal glioma | Both ectodermal and mesodermal<br>Ectopic breast's tissue<br><br>Fibrofolliculoma /trichodiscoma | Secondary cutaneous endometriosis<br><br>Ectopic Dsg 3 expression in respiratory epithelia<br><br>Post-traumatic ectopic nails on dorsum of hands or feet<br><br>Secondary osteoma cutis<br><br>Dystrophic calcinosis cutis<br><br>Parameatal urethral cyst (after surgery) |

**Table 2: Classification based on the ectopic contents**

| Deposition or expression of acellular contents | Ectopic glandular tissue | Ectopic adipose tissue |
|---|---|---|
| i. Calcification - Osteoma cutis and Calcinosis cutis<br><br>ii. Keratinization - hair type keratinization in music box spiny keratoderma<br><br>iii. Ectopic Dsg 3 expression in respiratory epithelia | i. Apocrine glands –<br>a. deep apocrine glands in nevus sebaceous<br>b. Ectopic Hidradenoma pappiliferum<br>c. Ectopic Hidradenitis suppurativa<br>d. Ectopic EMPD<br><br>ii. Sebaceous glands – Fordyce spots<br><br>iii. Mammary glands – Ectopic breast<br><br>iv.Thyroid gland<br><br>v. Endometrial glands<br><br>vi.Periurethral Littre's glands | i. Nevus lipomatosus cutaneous superficialis<br>ii. Fibrofolliculoma/trichodiscoma<br>iii. Folliculosebaceous cystic hamartoma<br><br>Ectopic neuromeningeal tissue<br>i. Nasal glioma<br>ii. Cutaneous meningioma<br><br>Ectopic visceral tissue<br>i. Subcutaneous nephrogenic rest<br>ii. Umbilical polyp<br><br>Other ectodermal appendages<br>i. Ectopic teeth (mostly canines)<br>ii. Ectopic nails |