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Sub corneal pustular dermatosis: A case Study

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Abstract
Subcorneal pustular dermatosis is also known as Sneddon-Wilkinson disease and is a rare neutrophilic dermatosis in which recurrent crops of sterile pustules appear in the most superficial layers of the skin. The majority of cases have been reported in adult females, generally over the age of 40. It is most common in middle-aged adults (particularly women) but can develop in children. The cause of subcorneal pustular dermatosis is still not known but IgA monoclonal gammopathy and multiple myeloma like factors are found to be responsible for occurrence. Phototherapy with psoralen with UVA (PUVA), broadband UVB, and narrowband UVB alone or in combination with dapsone and/or retinoids can be successful at controlling sub-corneal pustular dermatosis.

Keywords: Skin, sub-corneal pustular dermatosis, postules

Introduction

Sneddon and Wilkinson first described subcorneal pustular dermatosis (SPD) in the 1950s. SPD is a chronic and benign condition for which the primary concerns are minimizing morbidity, improving quality of life, and ruling out the presence of an associated internal disease. Subcorneal pustular dermatosis is characterised by numerous soft pustules at the skin surface. They usually appear on the trunk, particularly in the skin folds such as the armpits and groin [1].

Subcorneal pustular dermatosis dermnetnz.org/scaly/subcorneal-pustulosis.html www.orpha.net/consor/cgi-bin/OC_Exp.php?lng=en&Expert=48377

Pustules usually appear over a few hours and grow together to form round or wavy patterns. They may be mildly itchy or painful, but despite being pus filled, are not infected. The diagnosis of SPD is made based on the appearance of the pustules and the results of a skin biopsy [2].

Subcorneal pustular dermatosis: Comprehensive review and report of a case https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7330443/

Etiology

The cause of subcorneal pustular dermatosis is unknown. It is not caused by infection and is not contagious or cancerous. The pustules are thought to result from a signal pulling (recruiting) neutrophils to the affected areas of skin. Neutrophils are a type of white blood cell (WBC). Skin disorders that involve an abnormal recruitment of neutrophils are called neutrophilic dermatoses (NDs). While there are known triggers of neutrophil recruitment for some NDS, no specific triggers have been found to cause the pustule formation in SPD [2].

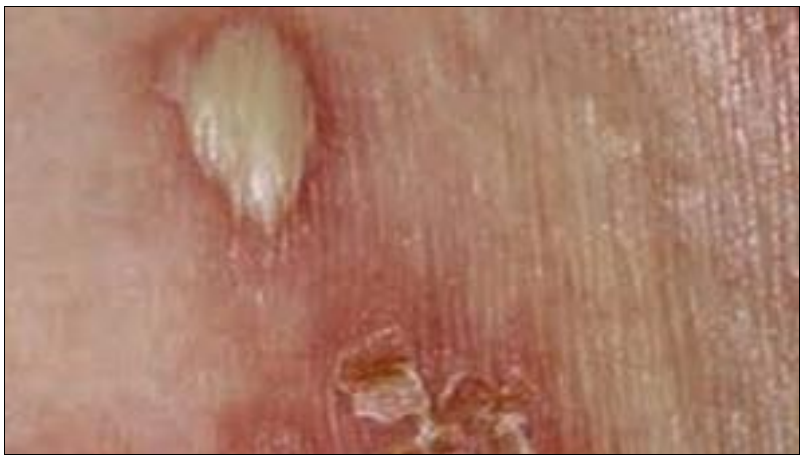

Fig 1: Sub-Corneal Pustular Dermatosis

Epidemiology

This condition is more common in middle-age and older women but has been reported to occur also in children. The incidence of SPD is unknown. There is no known ethnic or geographic predilection. More than 200 cases were reported in a review published in 1981.SPD is more common in females than in males (4:1).

Causes

The cause of sub-corneal pustular dermatosis is not known and also does not run in families. There is currently no

evidence it is inherited and it is not contagious. SPD may be associated with other diseases or health problems including Most often it occurs on its own, but has been linked to a variety of other diseases several autoimmune diseases, blood (hematologic) diseases, infections, and cancers.

How does it looks alike

It is a blistering skin condition with blisters developing on areas of normal looking skin or red and inflamed skin. Any area of skin can be involved, but the skin folds under the breast, underarm and the groin are common areas of involvement. It is less common for forearms and lower legs to be affected. The blisters develop quickly i.e. within hours and may be single or in clusters. The top of the blister comes off easily, and then a scab forms. When the skin heals, it is often slightly darker than before, but this will very gradually fade over weeks or months, and scarring does not usually occur. The bullae are flaccid and rupture easily, thus forming superficial crusts and scales. They heal often with mild hyperpigmentation. The lesions are mildly pruritic and not usually associated with constitutional symptoms; however, cases presenting with malaise, fever, or arthralgias [3].

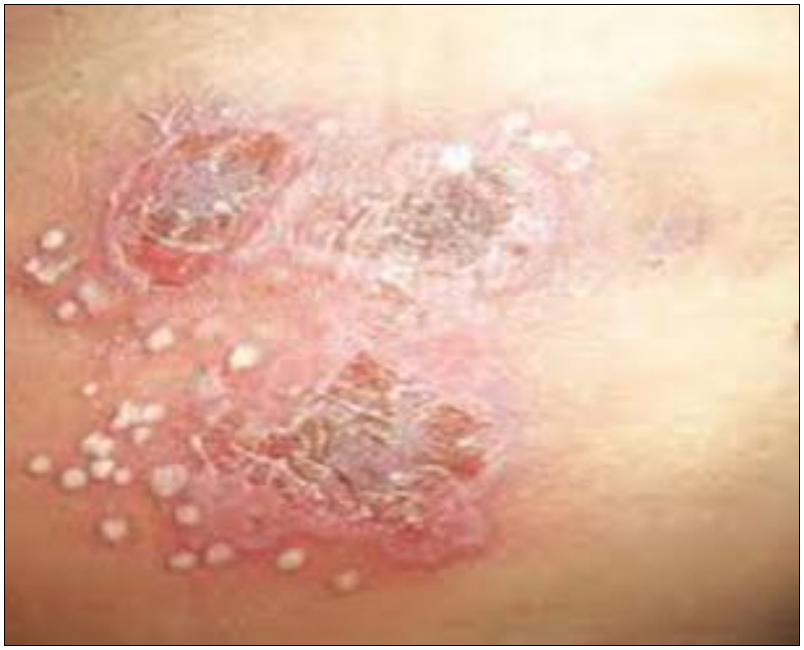

Fig 2: Depicts the appearance of Sub Corneal Postular Dermatosis

Investigations

The diagnosis can be made by looking at the skin, particularly in cases where there are the classical 'half and half blisters. A diagnosis is not always easy, as other blistering conditions, reactions to medicines and a form of psoriasis (pustular psoriasis).

- To confirm the diagnosis a Skin biopsy with immunofluorescence will be performed.
- Blood tests are likely to include a general screen such as Full blood count, Calcium levels and liver function tests, Auto antibodies, Bence–Jones protein, Serum protein electrophoresis to look for a gammopathy.

Treatment

Subcorneal pustular dermatosis usually clears over a period of about 4 weeks when treated with a tablet medication called dapsone.

Dapsone is often successful, with the lesions resolving over a month. The blisters can re-occur on stopping treatment therefore, intermittent treatment for months or even years with a low dose of dapsone is often required to keep the skin clear. Other therapies have been tried with mixed results.

- Steroid creams or steroid tablets can also be used with dapsone.
- Alternative tablet medications are sulphapyridine and sulphamethoxy pyridazine.
- Other treatments, for example acitretin, colchicine, tetracycline antibiotics, immunosuppressive medication or biological treatments can also be used successfully.

There are a few case reports detailing good response to phototherapy—psoralen and UVA (PUVA), narrowband UVB, or broadband UVB—in combination with dapsone or retinoids.

Both topical and systemic corticosteroids have been reported in isolated cases to provide some degree of control [4].

https://www.sciencedirect.com/science/article/pii/S2352647520300125?via%3Dihub

Case report

40-year-old women was admitted on 25may 2023 in our hospital with chief complaints of itchy eruption over the trunk, uppers libs and lower extremities over 1 months' duration. Patient was K/C/O hypothyroidism but there were no similar complaints, no personal or family history of psoriasis or any other skin disorder in the family. A complete blood count and the studies of serum biochemistry showed normal results; moreover, serum protein electrophoresis had negative results. The lesions initially developed over the trunk, uppers libs and lower extremities then they progressed up to involved almost the coverage of both the extremities. The perennial, lower abdomen and facial areas were spared, and no lymphadenopathy was

present. Till the report result patient was advised to apply coconut oil and mupirocin ointment but less relief was found and observed that patient is frequently itching on the affected areas

Histopathological testing revealed that a subcorneal pustule with numerous neutrophils, neutrophils infiltrating in the epidermis, mild intercellular oedema, and a dermal perivascular mixed inflammatory cell infiltrate with neutrophils. Bacterial and fungal stains and cultures from pustules also tested negative, and testing ruled out infections such as urinary tract infection. Serologic testing revealed level of TSH is high and T4 count is low and 8gram with exception of low haemoglobin level whereas reticulocyte count, liver and renal blood tests, and serum chemistries including calcium all were in normal ranges & no paraproteinemia was found. Direct immunofluorescence studies are negative for immunoglobulin A (IgA) intercellular accumulation.

On the basis of Physical Examination by dermatologist showed that well demarcated annular brownish plague undersize from 4 to 11 mm with few pustules noted that tended to form annular and serpiginous pattern and superficial crusts on the affected skin surface areas.

Taking into consideration a suspected diagnosis of SCPD, the patient was treated with levothyroxine & mupirocin topic ointment with (2%); moreover an incisional biopsy of a lesion was carried out. Further, Histopathology demonstrated a sub corneal vesiculo-bullous dermatitis; the pustule is located immediately below the stratum corneum and contains mainly neutrophils with few eosinophils. The underlying epidermis to the pustule shows slight intercellular oedema [5]. In the dermis, superficial blood vessels are surrounded by a nonspecific mixed inflammatory cell infiltrate consisting of neutrophils and mononuclear cells.

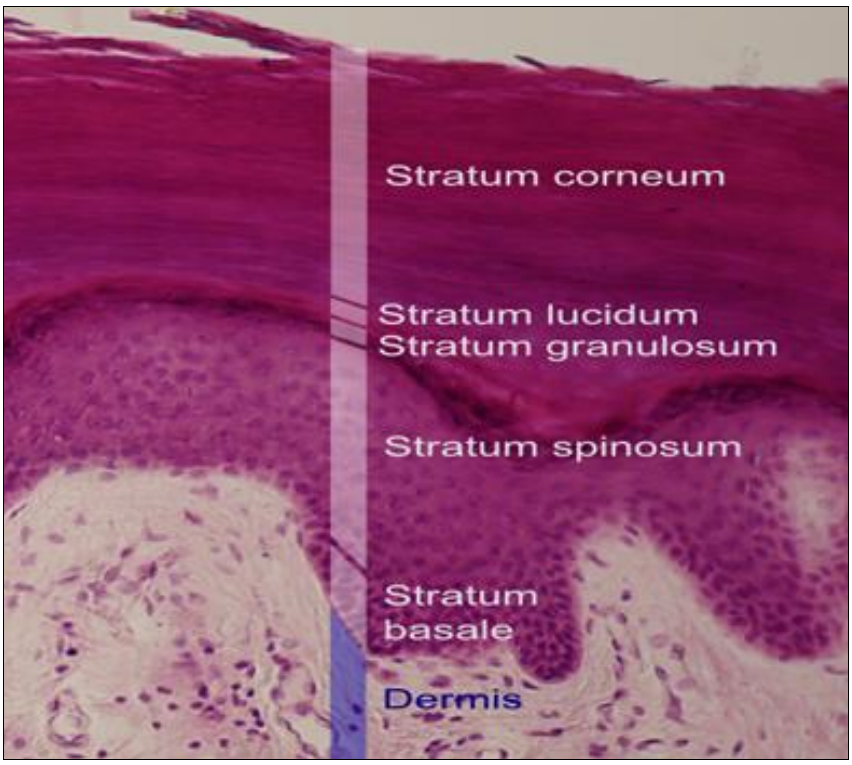

Fig 3: Sub corneal pustule immediately below the stratum corneum containing mainly neutrophils; the underlying epidermis show slight intercellular edema.

On the base of this finding, associated to histopathological features and the clinical date, a diagnosis of sub corneal pustular dermatitis (SCPD, Sneddon-Wilkinson disease) was made.

The patient was treated with 30 mg of oral Diaminodiphenylsulfone (Dapsone, 1 mg/kg/day), Supradyn tables, and mupirocin topical ointment The cutaneous lesions were almost completely healed within 2 weeks. After 4 weeks, treatment with Dapsone was continued on alternate days for another month, at the same daily dosage.

After 1.5 months, the patient was still recalled for the follow up at our hospital and observed that almost itchy had subsided and lesion had healed [6].

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