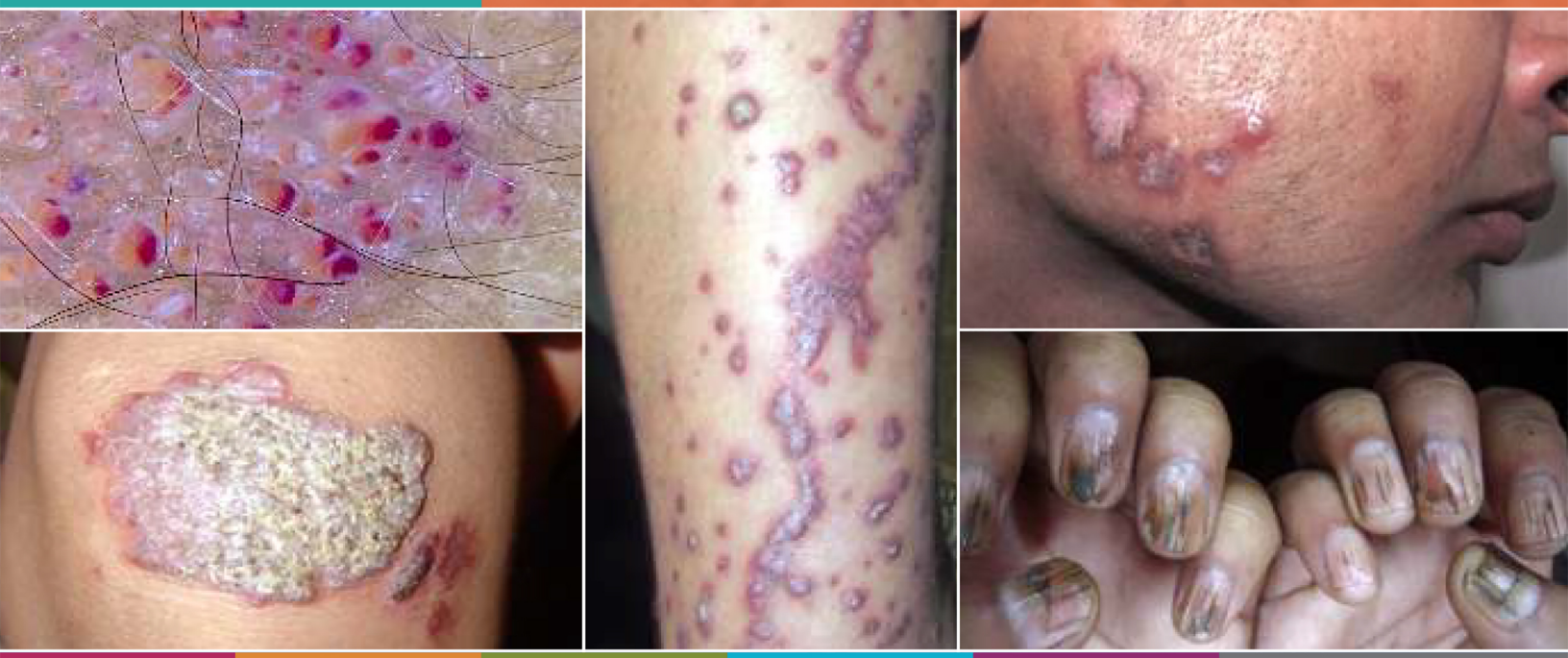
3rd Edition

Compendium of Dermatology *for* Examinations



Editors

Kabir Sardana
Surabhi Sinha
Seema Rani

CBSPD *Dedicated to Education*
CBS Publishers & Distributors Pvt Ltd

Chapter

26

Panniculitis

Seema Rani, Sinu Rose Mathachan, Kabir Sardana

These are a group of inflammatory diseases involving the subcutaneous fat.

CLASSIFICATION

A detailed classification is given in **Table 26.1** and we will focus on the most common entity that is erythema nodosum.

TABLE 26.1: Classification of the panniculitides

Histopathological type	Clinical condition
A. Predominantly septal panniculitides	
1. With vasculitis	
• Veins	• Superficial migratory thrombophlebitis
• Arteries	• Cutaneous polyarteritis nodosa
2. No vasculitis	
a. *Lymphocytes and plasma cells predominantly*	
• With granulomatous infiltrate in septa	• Necrobiosis lipoidica
• No granulomatous infiltrate in septa	• Deep morphea
b. *Histiocytes predominantly (granulomatous)*	
• With mucin in center of palisaded granulomas	• Subcutaneous granuloma annulare
• With fibrin in center of palisaded granulomas	• Rheumatoid nodule
• With large areas of degenerate collagen, foamy histiocytes and cholesterol clefts	• Necrobiotic xanthogranuloma
• Without mucin, fibrin or degeneration of collagen, but with radial granulomas in septa	• Erythema nodosum
B. Predominantly lobular panniculitides	
1. With vasculitis	
• *Small vessels*	
– Venules	• Erythema nodosum leprosum • Erythema induratum of Bazin
• *Large vessels*	
– Arteries	• Erythema induratum of Bazin

(Contd.)

TABLE 26.1: Classification of the panniculitides *(Contd.)*

Type	Clinical condition
2. No vasculitis	
a. *Few or no inflammatory cells*	
• Necrosis at the center of the lobule	• Sclerosing panniculitis
• With vascular calcification	• Calcific uremic arteriolopathy (calciphylaxis)
b. *Lymphocytes predominant*	
• With superficial and deep perivascular dermal infiltrate	• Cold panniculitis
• With lymphoid follicles, plasma cells and nuclear dust of lymphocytes	• Lupus panniculitis • Panniculitis associated with dermatomyositis
c. *Neutrophils predominant*	
• Extensive fat necrosis with saponification of adipocytes	• Pancreatic panniculitis (high serum lipase, calcium soap formation)
• With neutrophils between collagen bundles of deep reticular dermis panniculitis	• Alpha1-antitrypsin deficiency (splaying of neutrophils)
• With bacteria, fungi or protozoa	• Infective panniculitis
• With foreign bodies	• Factitious panniculitis
• Neutrophilic lobular panniculitis	• Subcutaneous Sweet syndrome
d. *Histiocytes predominant* (*granulomatous*)	
• No crystals in adipocytes	• Subcutaneous sarcoidosis • Traumatic panniculitis
• With crystals in histiocytes or adipocytes	• Subcutaneous fat necrosis of the newborn (associated hypercalcemia, localized nodules, favorable prognosis) • Poststeroid panniculitis (due to rapid steroid withdrawal in children) • Sclerema neonatorum (premature infants, diffuse hardening, poor prognosis) • Gouty panniculitis • Fungal panniculitis (zygomycosis, mucormycosis and aspergillosis)
• With cytophagic histiocytes	• Cytophagic histiocytic panniculitis (bean bag cells) • Subcutaneous panniculitis-like T cell lymphoma
• With sclerosis of the septa	• Sclerosing postirradiation panniculitis

ERYTHEMA NODOSUM

Most common panniculitis.
2nd–4th decade, Female > Male

Pathogenesis

- *Delayed hypersensitivity response (Th1 cytokine pattern) to various antigens.*

Causes

- Idiopathic—most common cause, followed by streptococcal infections, other infections (bacterial GI infections—*Yersinia*, *Salmonella*, *Campylobacter*), viral URIs, coccidioidomycosis, tuberculosis, and histoplasmosis).

- *Drugs* (estrogens/OCPs, sulfonamides, and NSAIDs)
- Sarcoidosis, Behçet disease, pregnancy
- IBD (Crohn > UC)

  Positive prognostic factor in sarcoidosis and coccidioidomycosis.

Clinical Features

Acute, tender subcutaneous nodules **(Fig. 26.1)** on pretibial areas (most commonly) bilaterally with overlying erythema resolving within 1–6 weeks by turning to bruise-like patches (erythema contusiformis), ulceration/scarring or atrophy is not seen, systemic symptoms—arthralgia, fever, malaise.

Chronic forms (subacute nodular migratory panniculitis/erythema nodosum migrans) can occur. Seen in women, mainly unilateral, migrating centrifugally, nodules (are less tender than EN).

Investigations

Box 26.1 lists the investigations required in erythema nodosum.

Histology

- Septal panniculitis with thickening/fibrosis of septae. No vasculitis.
- Neutrophils seen particularly in early lesions.
- Miescher microgranulomas: Small histiocytic aggregates surrounding a central stellate cleft; located in fat septa +/− thrombophlebitis (more common in EN-like lesions seen in Behçet disease).
- In later stages, the septa become fibrotic, partially replacing the fat lobules.

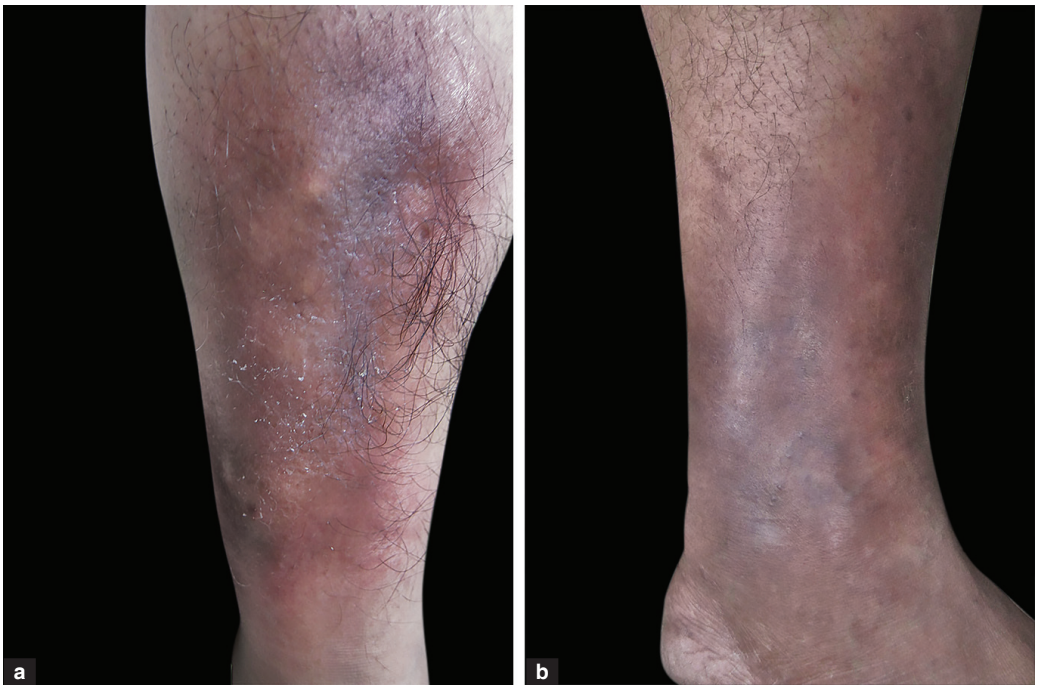


Fig. 26.1: Erythema nodosum

BOX 26.1: Investigations required in erythema nodosum

Complete blood count with ESR, CRP, throat swab culture, ASLO titers, Tuberculin skin test or IGRA, sputum for AFB, stool examination for cysts/ ova, chest X-ray, pregnancy testing in women of child-bearing age.

Treatment (Table 26.2)

TABLE 26.2: Depends upon etiology

- Rest and foot end elevation
- Aspirin, NSAIDs
- Supersaturated potassium iodide solution (SSKI)—2–10 drops (1 drop = 0.03 mL = 30 mg), three times per day in water or orange juice
- Colchicine (especially for Behçet disease)
- TNF-alpha inhibitors etanercept, infliximab (especially for IBD-associated)
- Others—thalidomide, cyclosporine, HCQS, dapsone, systemic corticosteroids (rarely used, after ruling out underlying infections)

BIBLIOGRAPHY

1. Luis Requena. Ponniculitis Griffiths EMC. Basker J. Bleiker J. Chalmers R. Creamer D. Editors. In: Rook's Textbook of Dermatology (9th ed). UK Blackwell Publisher. 2016. Part 8; Chapter 99: P2629–89.

Chapter

27

Perforating Dermatoses

Surabhi Sinha, Kabir Sardana, Sinu Rose Mathachan

INTRODUCTION

A group of disorders with perforation or elimination of dermal connective tissue components through the epidermis (also known as transepidermal elimination).

Transepidermal Elimination (TEE)

- Proposed by Mehregan in 1970.
- **Definition—**this is a pathologic dermoepidermal reactive phenomenon incited by exogenous substances or altered dermal constituents characterized by pseudoepitheliomatous hyperplasia of epidermis and/or follicular epithelium and formation of multiple transepithelial perforating channels, facilitating the extrusion of the altered dermal material or foreign substances to the exterior.
- TEE can occur as a primary disorder or secondary to other pathologic processes **(Table 27.1)**.

TABLE 27.1: Classification of TEE disorders

Classical conditions with TEE* (Primary)	Unspecified conditions with TEE (Secondary)
Elastosis perforans serpiginosa	Collagenoma perforant verruciforme
Reactive perforating collagenosis (inherited)	Chondrodermatitis nodularis helicis chronica
Acquired perforating dermatosis (acquired reactive perforating collagenosis—Kyrle disease)	Non-infective granulomatous disorders—perforating granuloma annulare, necrobiosis lipoidica, rheumatoid nodule, sarcoidosis
Perforating folliculitis	Infections—cutaneous TB, botryomycosis, chromoblastomycosis, schistosomiasis, leishmaniasis,lobomycosis, histoid leprosy
	Calcifying dermatoses—PXE, calcified hair follicle tumors, calcinosis cutis, osteoma cutis
	Foreign materials—silica, wood splinter

*Confusion over most terminologies. Acquired perforating dermatosis is now used for most acquired conditions earlier labeled as RPC/ Kyrle disease. Perforating folliculitis—this term is also going out of favor.

Epidemiology

Acquired perforating dermatosis—most common associated with **diabetes mellitus/chronic kidney disease** with/without dialysis.

Predisposing Factors

- Basic pathology: Transepidermal elimination of degenerated collagen/elastin/other connective tissue elements.
- **Diabetes mellitus**—strongly associated.
- **Chronic kidney disease**—strongly associated.

Clinical Features

Even though it is now believed that most of the older 'named' disorders can essentially be clubbed under APD, these dermatoses should still be known and are listed in **Table 27.2** and depicted in **Fig. 27.1**.

TABLE 27.2: Differential diagnosis of common perforating dermatoses

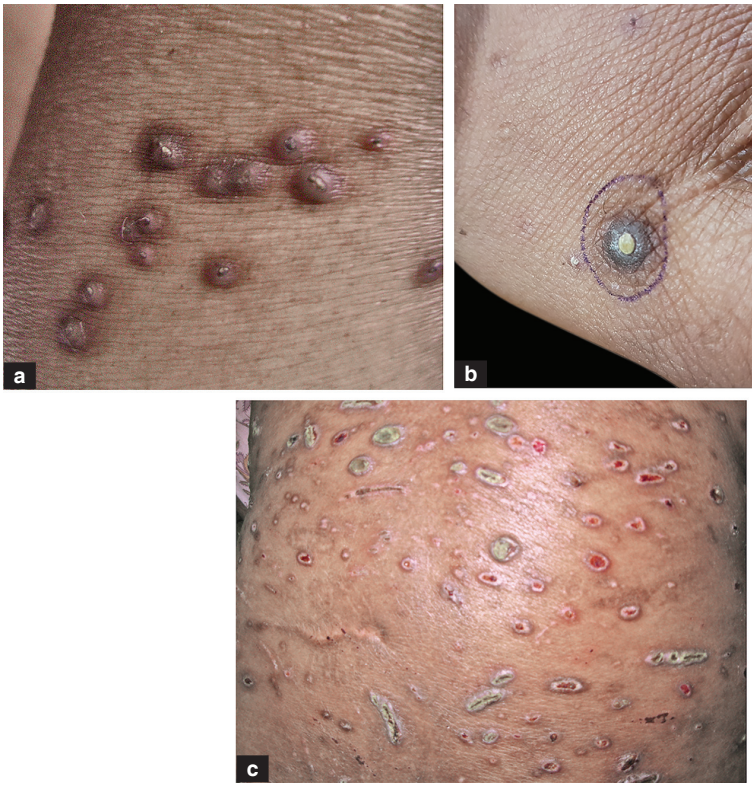
	Inherited EPS	Inherited/familial RPC	Acquired perforating dermatosis	Perforating folliculitis
Morphology	Non-follicular papules—linear/arcuate/serpiginous patterns with keratotic papules along rim	Small eroded papules with central hyperkeratotic plug	Pruritic dermatosis with keratotic dome-shaped papules with central crusts	Pruritic keratotic follicular papules
Age of onset	2nd decade	1st decade	5th–6th decades	3rd decade
Sites	Nape and sides of neck, face, upper limbs, lower limbs	Dorsae of hands, forearms, elbows, knees, lower legs, face	Extensors of limbs, trunk, head and neck	Hair-bearing parts of limbs
Precipitating factors/course	Scratching, insect bites May spontaneously resolve but tend to persist longer	Scratching, cold spontaneous resolution over 6–10 weeks	—	—
Koebner phenomenon	Occasionally +ve	Often +ve	Occasionally +ve	–ve
Associated diseases	**MAD PORES** • **M**arfan • **A**crogeria • **D**own syndrome • **P**enicillamine • **P**XE • **O**steogenesis imperfecta • **R**othmund–Thomson • **E**hlers–Danlos • **S**cleroderma	—	• **Diabetes mellitus** • **Chronic kidney disease** • Natalizumab therapy • Dermatomyositis • Congestive heart failure • Liver disease	None
Differential diagnosis	• Granuloma annulare • Tinea • Actinic granuloma • Perforating PXE • Familial inherited RPC • Porokeratosis of Mibelli	—	• Prurigo nodularis • Arthropod bites • Dermatofibroma • Folliculitis	—

(*Contd.*)

TABLE 27.2: Differential diagnosis of common perforating dermatoses *(Contd.)*

	Inherited EPS	Inherited/familial RPC	Acquired perforating dermatosis	Perforating folliculitis
Histopathology	Keratotic crusted plug surrounding epithelial hyperplasia **('crab-claw')** grabbing pink elastic fibers in superficial dermis (*VVG stain*: Elastic fibers stain black *vs* pink collagen)	Shallow cup-shaped epidermal invagination with degenerated collagen bundles—extruded through vertically oriented fine slits (VVG stain, Masson trichrome stain)	• Cup-shaped epidermal invagination with degenerated collagen bundles—extruded through vertically oriented fine slits • Resembles RPC most commonly (may also resemble perforating folliculitis or less commonly EPS)	Suppurative folliculitis Collagen fibers and elastin fibers and degenerated fibers surrounding follicles
Important Factoids	• Associated with inherited disorders of connective tissue and Down syndrome	• Sites of minor trauma • Collagen perforates out • Upper extremities	• Almost always a/w **diabetes** or **renal failure** (10% of dialysis patients) • Lower extremities (extensor) • Possible role of fibronectin/advanced glycation end product (AGE) modified collagen I and III in causing perforation	—

a/w, associated with; EPS, elastosis perforans serpiginosa



Figs 27.1: (a) Discrete papules and nodules with a central keratotic plug in APD; (b) Close up of lesions in acquired perforating dermatoses; (c) A case of APD with widespread lesions in a case with chronic renal failure

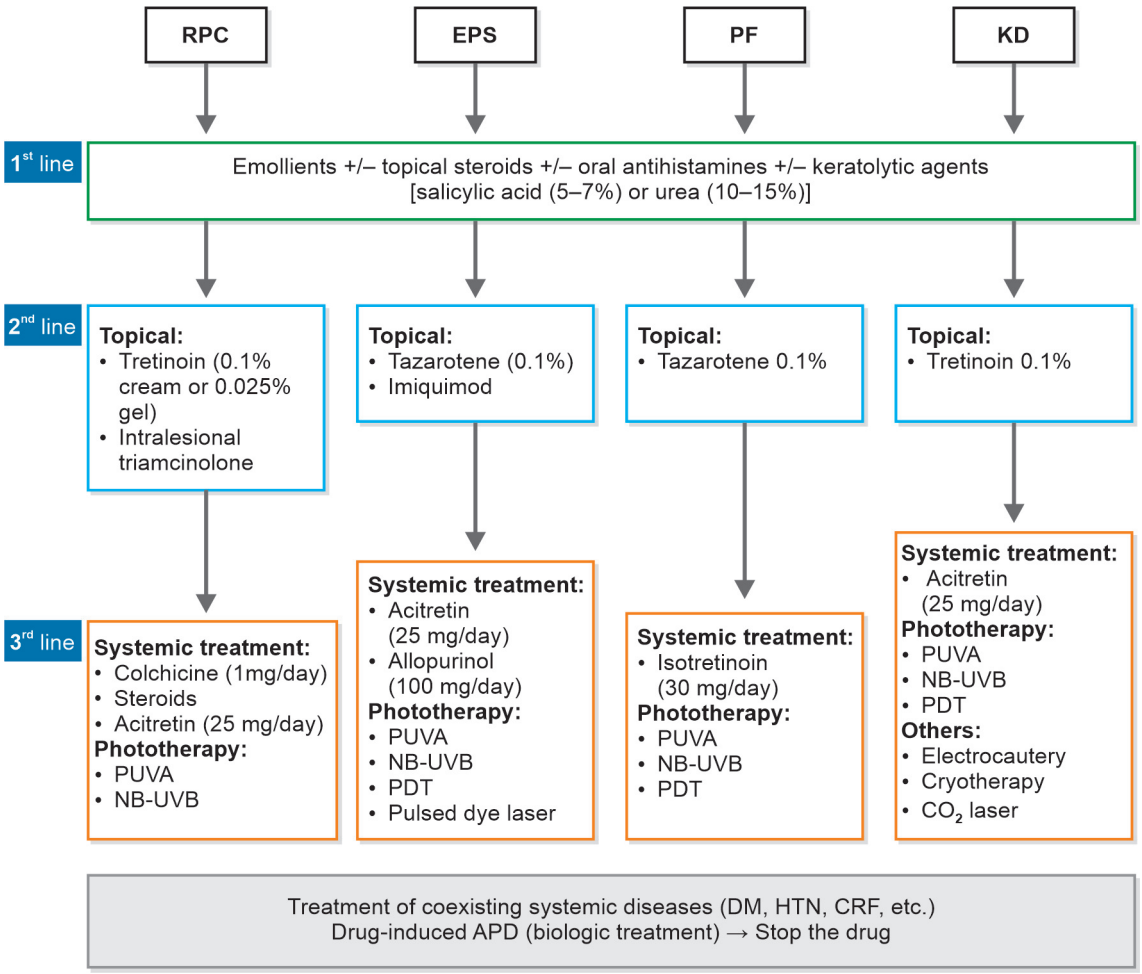
Treatment

Treatment of APD is listed in **Table 27.3** and a therapeutic algorithm for perforating dermatoses is shown in **Flowchart 27.1**.

TABLE 27.3: Therapy of acquired perforating dermatosis

First line	Second line	Third line
• Spontaneous resolution if kidney disease improves	• Oral isotretinoin • Allopurinol • Methotrexate • Rifampicin	• BB/NB UVB (most effective) • PUVA • PDT
• Topical tretinoin	• Emollients • Intralesional steroids • Topical steroids under occlusion • Topical tacalcitol	

Flowchart 27.1: A summarized therapeutic algorithm for perforating dermatoses



BIBLIOGRAPHY

Books

1. Dermatology (Bolognia). 4th Edn. Elsevier. Chapter 96.
2. Granulomatous, necrobiotic and perforating dermatoses. In. McKee's Pathology of the Skin. 4th Edition.
3. Rook's Textbook of Dermatology. 9th Edition. Wiley Blackwell. Chapter 96. Acquired disorders of dermal connective tissue.

Journals

1. García-Malinis AJ, Del Valle Sánchez E, Sánchez-Salas MP, Del Prado E, Coscojuela C, Gilaberte Y. Acquired perforating dermatosis: clinicopathological study of 31 cases, emphasizing pathogenesis and treatment. J Eur Acad Dermatol Venereol. 2017 Oct; 31(10):1757–63.
2. Shah H, Tiwary AK, Kumar P. Transepidermal elimination: Historical evolution, pathogenesis and nosology. Indian J Dermatol Venereol Leprol 2018; 84:753–7.