eMedicine Specialties > Dermatology > Bullous Diseases

Familial Benign Pemphigus (Hailey-Hailey Disease)

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Updated: Aug 13, 2008

Introduction

Background

Familial benign pemphigus originally was described by the Hailey brothers in 1939.[1] It is a chronic autosomal dominant disorder with incomplete penetrance. Approximately two thirds of patients have a family history of the disorder. A history of multiple relapses and remissions is characteristic. Decreased numbers of desmosomes have been implicated in the pathogenesis of benign familial pemphigus. Therapeutic options are limited.

Among the eMedicine articles on pemphigus are the following:

- Pemphigus Erythematosus
- Pemphigus Foliaceus
- Pemphigus Herpetiformis
- Pemphigus Vulgaris

Additionally, the article Advances in Pemphigus Therapy, available from Medscape, may be of interest.

Pathophysiology

Keratinocytes are held together through desmosomes and adherens junctions. These junctions consist of calcium-binding transmembrane glycoproteins, which contribute to cellular adhesion. Many hypotheses exist concerning the pathogenesis of familial benign pemphigus, but the cause remains uncertain. An overall defect in keratinocyte adhesion appears to be secondary to a primary defect in a calcium pump protein, ATP2C1.

ATP2C1 encodes the secretory pathway Ca^{2+}/Mn2 ATPase (hSPCA1). Mutant proteins in familial benign pemphigus create a loss of sensitivity to Ca^{2+} and Mn^{2+} ion binding and transport. Low levels of Ca^{2+} within Golgi bodies impair protein processing. Gene expression may be affected in benign familial pemphigus, as may phosphorylation of adhesion molecules. Localized postzygotic mutation has caused segmental manifestations of familial benign pemphigus.[2,3]

Frequency

United States

No precise data are available on the incidence of familial benign pemphigus.

Mortality/Morbidity

Familial benign pemphigus causes discomfort but is not life threatening. Benign familial pemphigus lesions often begin during the teenage years and manifest as itchy and malodorous plaques.

Sex

Both sexes are affected equally.

Age

Familial benign pemphigus often manifests in the late teenage years or in adulthood (30s and 40s).

Clinical

History

A family history of benign familial pemphigus usually is present. Commonly, patients may not have symptoms until ages 30-49 years. Delayed diagnosis of familial benign pemphigus also is common, especially if the patient's lesions respond to topical corticosteroids, antibiotics, or antifungals.

Physical

With familial benign pemphigus, vesicles and erythematous plaques with overlying crusts typically occur in the genital area, as well as the chest, neck, and axillary areas (see Media Files 1-6). Burning and itching accompany the eruption, and a malodorous drainage occurs in some cases as a result of secondary infection. Symptoms related to staphylococcal and candidal overgrowth are common in familial benign pemphigus. Multiple asymptomatic longitudinal white bands on the fingernails also have been described. Involvement of mucosa is rare. The characteristic clinical appearance of familial benign pemphigus, as alvato da Windows Internet Explorer 8> Subject: Familial Benign Pemphigus (Hailey-Hailey Disease): [Print] - eMedicine Dermatology Date: Fri, 4 Sep 2009 00:36:51 +0200 MIME-Version: 1.0 Content-Type: multipart/related; type="text/html"; boundary="----=_NextPart_000_00B5_01CA2CF7.CB585AF0" X-MimeOLE: Produced By Microsoft MimeOLE V6.00.2900.5579 This is a multi-part message in MIME format. ------=_NextPart_000_00B5_01CA2CF7.CB585AF0 Content-Type: text/html; charset="Windows-1252" Content-Transfer-Encoding: quoted-printable Content-Location: http://emedicine.medscape.com/article/1063224-print

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Causes

Hailey-Hailey disease, or familial benign pemphigus, is hypothesized to result from a genetic defect in a calcium pump protein. The pump mutation is in *ATP2C1*, a gene localized on chromosome 3.[4] This gene defect is similar to the genetic defect in Darier disease, which also is a calcium pump defect, *ATP2A2*. The gene *ATP2C1* encodes the human secretory pathway Ca^{++} -ATPase hSPCA1, which is dysfunctional and causes abnormal calcium release from the Golgi apparatus and endoplasmic reticulum.

In addition to the primary gene defect in Hailey-Hailey disease (familial benign pemphigus), contributing factors are known that exacerbate the disease. These include heat, friction, and infection, resulting in separation of keratinocytes, especially in the intertriginous areas. Through ultrastructural studies of familial benign pemphigus lesions, characteristic changes in keratinocyte morphology have been described, including retracted tonofilaments, elongated membrane microvilli, and reduced numbers of desmosomes.

Differential Diagnoses

Erythrasma
Extramammary Paget Disease
Intertrigo
Pemphigoid Gestationis
Pemphigus Erythematosus
Pemphigus Foliaceus
Pemphigus Herpetiformis
Pemphigus Vulgaris
Pemphigus, Drug-Induced
Pemphigus, IgA
Pemphigus, Paraneoplastic
Pyoderma Vegetans
Tinea Corporis
Tinea Cruris

Workup

Laboratory Studies

- Microscopic examination reveals intraepidermal and suprabasilar acantholysis (see Media File 7). Elongated papillae (villi) extend into lacunae, and a single layer of basal cells lines the villi.
- Many layers of detached keratinocytes (acantholysis) exist and appear similar to a dilapidated brick wall (clumped acantholytic cells have only a few intact intercellular bridges holding the keratinocytes together).
- Unlike pemphigus vulgaris, direct immunofluorescence in familial benign pemphigus s negative. The Nikolsky sign is negative, and Tzanck preparation fails to reveal giant cells and syncytia formation characteristic of herpes virus infection.
- Serologic studies for familial benign pemphigus fail to reveal circulating autoantibodies, unlike pemphigus vulgaris in which antibody titers correlate with disease activity.

Histologic Findings

Histologic findings in familial benign pemphigus are suprabasilar and widespread acantholysis.

Treatment

Medical Care

Familial benign pemphigus waxes and wanes in intensity. Soothing compresses (aluminum acetate 1:40 dilution) followed by intermittent use of mild corticosteroid preparations (class V or class VI corticosteroids) and topical antibiotics (clindamycin or erythromycin) result in transient improvement. More widespread flares of familial benign pemphigus may require systemic antibiotics to suppress protease activation and acantholysis. Erythromycin and tetracycline are favored. Bacterial culture and sensitivity can help guide appropriate therapy.

In patients with refractory cases of familial benign pemphigus (Hailey-Hailey disease), dapsone, systemic corticosteroids, methotrexate, retinoids (isotretinoin or acitretin),[5] and etretinate have been tried and have been reported to be of value in some anecdotal reports. Most patients with familial benign pemphigus at the author's institution respond well to anti-infective therapy and short courses of corticosteroids, and other immunosuppressive agents have only rarely been helpful in the author's experience. Topical tacrolimus ointment has been a valuable addition to the treatment regimen and has been able to control familial benign pemphigus well, even without the adjunctive use of topical corticosteroids.

Topical tacrolimus ointment has been found to be helpful in familial benign pemphigus,[6] and photodynamic therapy with 5-aminolevulinic acid has been used for recalcitrant cases.[7]

Reports[8,9] indicate that low-dose botulinum toxin type A injection may be of benefit for familial benign pemphigus. Control of hyperhidrosis, which aggravates familial benign pemphigus (Hailey-Hailey disease), may be the mechanism for this off-label, novel approach.

Isolated reports of oral acitretin[10] therapy or intramuscular alefacept[11] leading to improvement in familial benign pemphigus warrant further study.

Surgical Care

Dermabrasion, carbon dioxide laser ablation, and pulsed dye laser therapy have been tried in the treatment of familial benign pemphigus, with variable success.[12,13,14]

Diet

To help minimize friction, it is recommended that patients with familial benign pemphigus maintain their weight at appropriate levels.

Activity

Instruct patients with familial benign pemphigus (Hailey-Hailey disease) to select cool and comfortable clothing that reduces heat, moisture, and friction. Patients should avoid fabrics or clothing styles that rub or irritate affected areas. Washing new shirts may soften the collars. In some cases, pain may limit physical activities.

Medication

The goals of pharmacotherapy in familial benign pemphigus are to reduce morbidity and to prevent complications. Reportedly,[8,9] off-label use of low-dose botulinum toxin type A injection may be of benefit to control hyperhidrosis, which aggravates familial benign pemphigus.

Immunosuppressants

Used in refractory cases. Ameliorate symptoms of inflammation (eg, pain, swelling, stiffness). Use of systemic immunosuppressive therapy for familial benign pemphigus is controversial. No large-scale studies offer a clear evidence-based approach to immunosuppressive therapy in the management of familial benign pemphigus. Because adverse effects can be severe and even fatal at times, such therapies must be initiated cautiously and with adequate informed consent.

Methotrexate (Rheumatrex, Folex PFS)

Antimetabolite that inhibits dihydrofolate reductase, thereby hindering DNA synthesis and cell reproduction.

Dosing

Adult

10-12.5 mg PO qwk

Pediatric

Not recommended

Interactions

Oral aminoglycosides may decrease absorption and blood levels of concurrent oral MTX; charcoal lowers MTX levels; coadministration with etretinate may increase hepatotoxicity of MTX; folic acid or its derivatives contained in some vitamins may decrease response to MTX; coadministration with NSAIDs may be fatal; indomethacin and phenylbutazone can increase MTX plasma levels; may decrease phenytoin serum levels; probenecid, salicylates, procarbazine, and sulfonamides, including TMP-SMZ, may increase effects and toxicity of MTX; may increase plasma levels of thiopurines

Contraindications

Documented hypersensitivity; alcoholism; hepatic insufficiency; documented immunodeficiency syndromes; preexisting blood dyscrasias (eg, bone marrow hypoplasia, leukopenia, thrombocytopenia, significant anemia); renal insufficiency

Precautions

Pregnancy

D - Fetal risk shown in humans; use only if benefits outweigh risk to fetus

Precautions

Monitor CBC counts monthly, and liver and renal function q1-3mo during therapy (monitor more frequently during initial dosing, dose adjustments, or when risk of elevated MTX levels, eg, dehydration); MTX has toxic effects on hematologic, renal, GI tract, pulmonary, and neurologic systems; discontinue if significant drop in blood counts; aspirin, NSAIDs, or low dose steroids may be administered concomitantly with MTX (possibility of increased toxicity with NSAIDs including salicylates has not been tested)

Corticosteroids

Have anti-inflammatory properties and cause profound and varied metabolic effects. In addition, these agents modify the body's immune response to diverse stimuli.

Prednisone (Deltasone, Orasone)

Glucocorticoid (adrenocortical steroid) absorbed easily into GI tract. May decrease inflammation by reversing increased capillary permeability and suppressing PMN activity.

Dosing

Adult

0.5-1 mg/kg/d PO prn for short periods

Pediatric

Administer as in adults

Interactions

Patients on corticosteroid therapy should not be vaccinated against smallpox or immunized because of possible neurologic complications and lack of antibody response; coadministration with estrogens may decrease prednisone clearance; when used with digoxin, digitalis toxicity secondary to hypokalemia may increase; phenobarbital, phenytoin, and rifampin may increase metabolism of glucocorticoids (consider increasing maintenance dose); monitor for hypokalemia with coadministration of diuretics

Contraindications

Documented hypersensitivity; viral infection; peptic ulcer disease; hepatic dysfunction; severe osteoporosis; diabetes; connective tissue infections; fungal or tubercular skin infections

Precautions

Pregnancy

C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Precautions

Reduced resistance and masked signs of infections; prolonged use may result in cataracts and glaucoma; psychic changes may be exhibited, such as mood swings and depression

Triamcinolone (Aristocort)

Treats inflammatory dermatosis that is responsive to steroids. Decreases inflammation by suppressing migration of polymorphonuclear leukocytes and reversing capillary permeability. Use 0.1% cream.

Dosing

Adult

Apply thin film bid/tid until favorable response

Pediatric

Apply as in adults

Interactions

None reported

Contraindications

Documented hypersensitivity; fungal, viral, and bacterial skin infections

Precautions

Pregnancy

C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Precautions

Caution in pediatric patients; do not use in decreased skin circulation; prolonged use, applications over large areas, and use of potent steroids and occlusive dressings may result in systemic absorption; systemic absorption may cause Cushing syndrome, reversible HPA-axis suppression, hyperglycemia, and glycosuria

Retinoids

Inhibit sebaceous gland function and modify keratinization.

Isotretinoin (Accutane)

Decreases sebaceous gland size and sebum production. May inhibit sebaceous gland differentiation and abnormal keratinization. Used to treat severe cystic acne.

Dosing

Adult

1 mg/kg/d PO

Pediatric

Not recommended

Interactions

Toxicity may occur with beta carotene coadministration; pseudotumor cerebri or papilledema may occur; may reduce plasma levels of carbamazepine

Contraindications

Documented hypersensitivity; pregnancy or potential pregnancy (unless proper contraceptives used); sensitivity to paraben (preservative in gelatin capsule)

Precautions

Pregnancy

X - Contraindicated; benefit does not outweigh risk

Precautions

Women should not be pregnant when on Accutane therapy; may decrease night vision; may be associated with development of hepatitis; occasional exaggerated healing response of acne lesions (excessive granulation with crusting) may occur; patients with diabetes may experience problems in controlling blood sugar while on isotretinoin; avoid exposure to UV light or sunlight until tolerance achieved; discontinue if rectal bleeding, abdominal pain, or severe diarrhea occur

Acitretin (Soriatane)

Retinoic acid analog, similar to etretinate and isotretinoin. Etretinate is primary metabolite and has demonstrated clinical effects close to those seen with etretinate. Mechanism of action is unknown.

Dosing

Adult

0.5-0.75 mg/kg/d PO

Pediatric

Not recommended

Interactions

Increases toxicity of MTX (avoid concomitant use); interferes with effects of microdosed progestin minipill; coadministration with alcohol may enhance synthesis of etretinate, which has much longer half-life than acitretin (>120 d)

Contraindications

Documented hypersensitivity

Precautions

Pregnancy

X - Contraindicated; benefit does not outweigh risk

Precautions

Do not use in severe obesity; women of childbearing age must be capable of complying with effective contraceptive measures; recommended that contraception be continued for at least 3 y after stopping treatment with acitretin; etretinate may form from acitretin, which takes approximately 2-3 y to clear from the body; caution if impaired renal or liver function; perform AST, ALT, and LDH tests prior to initiation of acitretin therapy at 1- to 2-wk intervals until stable and thereafter at intervals as clinically indicated

Etretinate (Tegison)

Not available in the United States. Retinoic acid analog. Used only after other medicines have been tried and failed.

Dosing

Adult

0.5-1 mg/kg/d PO divided bid

Pediatric

Not recommended

Interactions

Increases MTX toxicity (avoid concomitant use); interferes with effects of microdosed progestin minipill

Contraindications

Documented hypersensitivity

Precautions

Pregnancy

X - Contraindicated; benefit does not outweigh risk

Precautions

Do not use in severe obesity; women of childbearing age must be capable of complying with effective contraceptive measures; recommended that contraception be continued for at least 3 y after stopping treatment; etretinate takes approximately 2-3 y to clear from the body; caution if impaired renal or liver function; perform AST, ALT, and LDH tests prior to initiation of therapy at 1- to 2-wk intervals until stable and thereafter at intervals as clinically indicated

Antimicrobials

Used to eliminate microorganisms. Use for possible secondary bacterial infections. Also, some antimicrobials have immunomodulatory effects.

Erythromycin (E.E.S., E-Mycin, Ery-Tab)

Inhibits bacterial growth, possibly by blocking dissociation of peptidyl tRNA from ribosomes causing RNA-dependent protein synthesis to arrest. For treatment of staphylococcal and streptococcal infections. In children, age, weight, and severity of infection determine proper dosage. When bid dosing is desired, half-total daily dose may be taken q12h. For more severe infections, double the dose.

Dosing

Adult

250 mg PO q6h or 500 mg PO q12h

Pediatric

20 mg/kg PO 2 h prior to procedure, followed by 10 mg/kg 6 h later

Interactions

Coadministration may increase toxicity of theophylline, digoxin, carbamazepine, and cyclosporine; may potentiate anticoagulant effects of warfarin; coadministration with lovastatin and simvastatin, increases risk of rhabdomyolysis

Contraindications

Documented hypersensitivity; hepatic impairment

Precautions

Pregnancy

B - Fetal risk not confirmed in studies in humans but has been shown in some studies in animals

Precautions

Caution in liver disease; estolate formulation may cause cholestatic jaundice; adverse GI tract effects are common (give doses pc); discontinue use if nausea, vomiting, malaise, abdominal colic, or fever occur

Clindamycin hydrochloride (Cleocin)

Lincosamide for treatment of serious skin and soft tissue staphylococcal infections. Also effective against aerobic and anaerobic streptococci (except enterococci). Inhibits bacterial growth, possibly by blocking dissociation of peptidyl tRNA from ribosomes causing RNA-dependent protein synthesis to arrest.

Dosing

Adult

150-300 mg PO q6h

Pediatric

8-16 mg/kg/d PO divided tid/qid

Interactions

Increases duration of neuromuscular blockade induced by tubocurarine and pancuronium; erythromycin may antagonize effects of clindamycin; antidiarrheals may delay absorption of clindamycin; cyclosporine levels may decrease when administered concurrently; kaolin may reduce absorption

Contraindications

Documented hypersensitivity; regional enteritis; ulcerative colitis; hepatic impairment; antibiotic-associated colitis

Precautions

Pregnancy

C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Precautions

Adjust dose in severe hepatic dysfunction, no adjustment necessary in renal insufficiency; associated with severe and possibly fatal colitis; may result in colitis; occasionally results in overgrowth of nonsusceptible organisms (eg, yeast)

Dapsone (Avlosulfon)

Bactericidal and bacteriostatic against mycobacteria; mechanism of action is similar to that of sulfonamides where competitive antagonists of PABA prevent formation of folic acid, inhibiting bacterial growth. Anti-inflammatory mechanism of action most likely relates to inhibition of neutrophils through suppression of the halide-myeloperoxidase system.

Dosing

Adult

50-200 mg PO qd

Pediatric

2 mg/kg PO qd; not to exceed 100 mg/d

Interactions

May inhibit anti-inflammatory effects of clofazimine; hematologic reactions may increase with folic acid antagonists, such as pyrimethamine (monitor for agranulocytosis during second and third mo of therapy); probenecid increases dapsone toxicity; trimethoprim with dapsone may increase toxicity of both drugs; because of increased renal clearance, dapsone levels may decrease significantly when administered concurrently with rifampin

Contraindications

Documented hypersensitivity; G-6-PD deficiency; anemia

Precautions

Pregnancy

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Precautions

Sulfa allergy; history of liver, kidney, or heart disease; dapsone induces hemolysis and results in dose-related reduction in hemoglobin; myelosuppression and agranulocytosis have been reported, monitor patients frequently with CBC counts; methemoglobinemia, hepatitis, neuropathy, and headaches have been reported

Tetracycline (Sumycin)

Treats gram-positive and gram-negative organisms, as well as mycoplasmal, chlamydial, and rickettsial infections. Inhibits bacterial protein synthesis by binding with 30S and possibly 50S ribosomal subunit(s).

Dosing

Adult

250-500 mg PO q6h

Mild-to-moderate infections: 500 mg PO bid or 250 mg PO qid for 7-14 d

Severe infections: 500 mg PO qid for 7-14 d

Pediatric

<8 years: Not recommended

>8 years: 25-50 mg/kg/d (10-20 mg/lb) PO qid

Interactions

Bioavailability decreases with antacids containing aluminum, calcium, magnesium, iron, or bismuth subsalicylate; may enhance agents with neuromuscular blocking effect; can decrease effects of oral contraceptives, causing breakthrough bleeding and increased risk of pregnancy; tetracyclines can increase hypoprothrombinemic effects of anticoagulants

Coadministration with retinoids can cause increased intracranial pressure (coadministration contraindicated); administer tetracycline at least 1 h before or 4-6 h after colestipol or cholestyramine; if tetracycline administered concurrently with digoxin, monitor digoxin levels (dosage adjustment for digoxin may be required; risk of interaction may be reduced if given with Lanoxicaps)

Contraindications

Documented hypersensitivity; severe hepatic dysfunction

Precautions

Pregnancy

D - Fetal risk shown in humans; use only if benefits outweigh risk to fetus

Precautions

Photosensitivity may occur with prolonged exposure to sunlight or tanning equipment; reduce dose in renal impairment; consider drug serum level determinations in prolonged therapy; tetracycline use during tooth development (last one half of pregnancy through age 8 y) can cause permanent discoloration of teeth; Fanconi-like syndrome may occur with outdated tetracyclines

Pseudotumor cerebri has been associated with tetracyclines, therefore, possibility for permanent sequelae exists

Clindamycin phosphate solution 10 mg/mL (Cleocin T, Clindets, Clinda-Derm)

Lincosamide for treatment of serious skin and soft tissue staphylococcal infections when taken systemically. Useful in treatment of acne when applied topically. Inhibits bacterial growth, possibly by blocking dissociation of peptidyl tRNA from ribosomes causing RNA-dependent protein synthesis to arrest.

Dosing

Adult

Apply to affected area bid

Pediatric

Administer as in adults

Interactions

None with topical use; systemic use may enhance effects of neuromuscular blocking agents

Contraindications

Documented hypersensitivity

Precautions

Pregnancy

B - Fetal risk not confirmed in studies in humans but has been shown in some studies in animals

Precautions

Systemic (not topical) use has been associated with severe colitis; GI tract disturbances rarely have been reported with topical use; prolonged use may result in overgrowth of nonsusceptible organisms resulting in gram-negative folliculitis; discontinue if superinfection occurs

Ketoconazole (Nizoral)

Imidazole that inhibits the synthesis of ergosterol, thereby affecting cell membrane integrity and resulting in fungal cell death.

Dosing

Adult

Apply 2% cream to affected area qd/bid for 2-6 wk

Pediatric

Not established

Interactions

None reported

Contraindications

Documented hypersensitivity

Precautions

Pregnancy

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Precautions

Local reactions may occur including rash, irritation, burning, and pruritus; rarely, systemic absorption may occur

Astringents

Drying agents used in management of hyperhidrosis.

Aluminum chloride (Drysol)

Aluminum chloride hexahydrate 20% in absolute alcohol. Antiperspirant mechanism of action is not known, although creation of aluminum-containing casts within the sweat duct has been postulated.

Dosing

Adult

Apply to affected area hs for 2-7 consecutive nights, then prn; to prevent irritation, completely dry area prior to application

Pediatric

Not established

Interactions

None reported

Contraindications

Documented hypersensitivity

Precautions

Pregnancy

C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Precautions

External use only; not for application on irritated, broken, or recently shaved skin (some dermatologists use 5% aluminum acetate for hemostasis after shave biopsies)

Aluminum acetate 5% soak (Bite Rx)

Dissolve aluminum acetate tablets in water to attain a 1:20 solution. Has a drying effect on vesicular or wet dermatoses.

Dosing

Adult

Apply as compress for 20-30 min 4-6 times/d

Pediatric

Administer as in adults

Interactions

None reported

Contraindications

Documented hypersensitivity

Precautions

Pregnancy

C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Precautions

External use only

Follow-up

Further Outpatient Care

- Almost all patients with familial benign pemphigus can be treated successfully on an outpatient basis, but many patients may need to be excused temporarily from manual labor so that affected body folds can heal promptly under treatment.
- Familial benign pemphigus patients require regular evaluation to be sure that secondary infection is brought under control and that the adverse effects of topical corticosteroids (eg, cutaneous atrophy) are avoided.
- Individuals who receive intermittent courses of systemic corticosteroids should be evaluated for possible decrease in bone density and should be instructed regarding diet and therapy that may maintain bone density.
- One report describes squamous cell carcinoma arising in the setting of familial benign pemphigus. Biopsy specimens should be taken from any suspicious infiltrated areas.

Inpatient & Outpatient Medications

- Intracutaneous botulinum toxin A injection may be of benefit by inhibiting sweating. Remissions of up to at least 12 months have been achieved using only botulinum A toxin injection. Only a few isolated reports have been published,[8,9] and further work is needed to explore the benefits of this off-label procedure.

Deterrence/Prevention

- Maintaining a healthy weight and keeping the body folds cool and dry as much as possible help prevent flares of the disease.

Complications

- Cellulitis, abscess formation, scarring, and depression concerning the chronic and refractory nature of the skin condition are possible complications.
- Systemic corticosteroid therapy may result in the adverse effects of steroids, such as osteoporosis, cataracts, striae, ulcers, and others.

Prognosis

- Patients live long and productive lives. The skin disorder is more of a nuisance than a serious health threat.

Patient Education

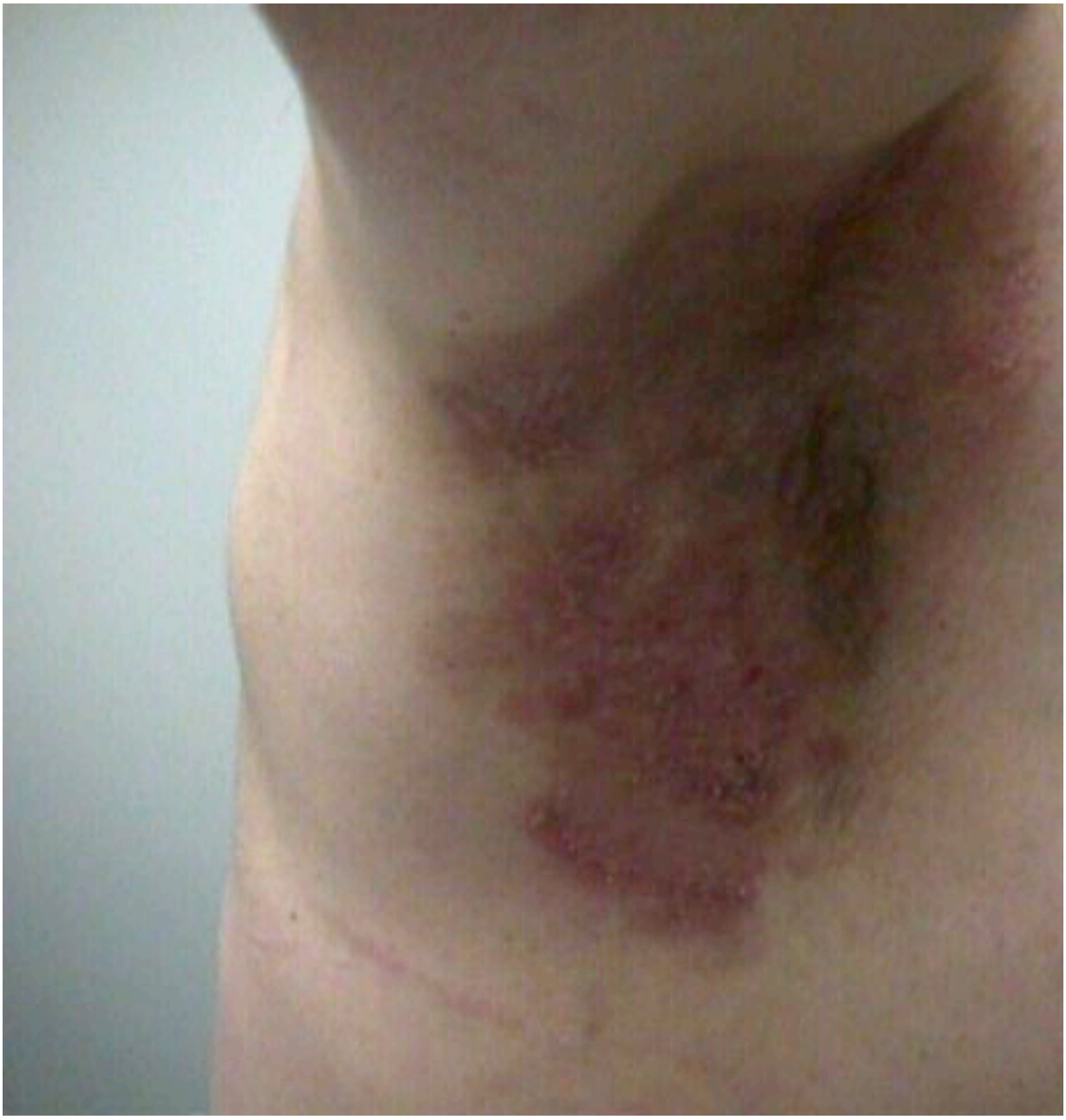
- Patients with familial benign pemphigus must be instructed to recognize flares in the disease promptly and to seek treatment for secondary infection before it becomes severe.

Miscellaneous

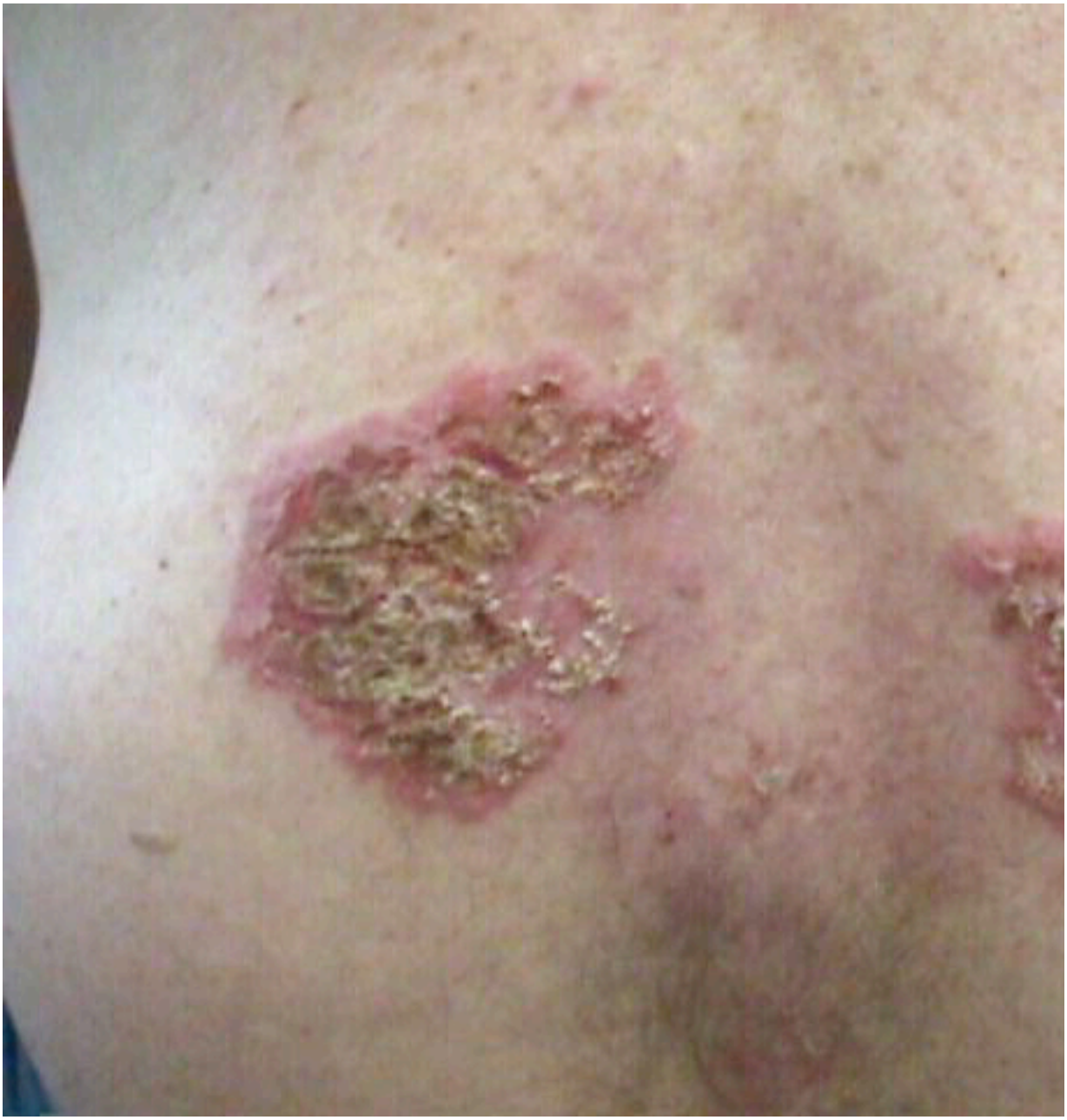
Medicolegal Pitfalls

- Failure to monitor adverse effects of systemic therapy is the most significant pitfall. Systemic corticosteroids should be used intermittently and as a last resort when topical therapy fails.

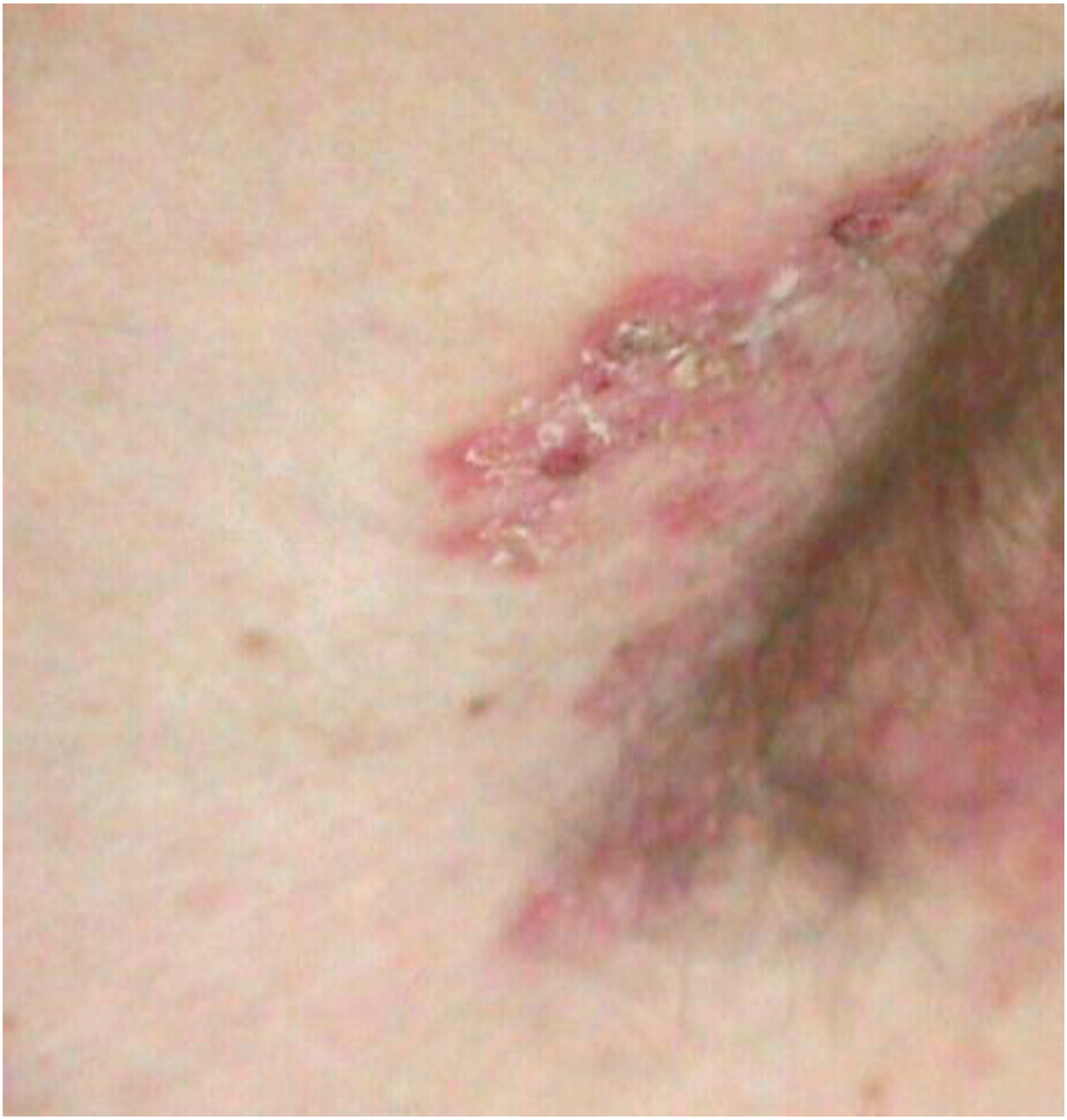
Multimedia



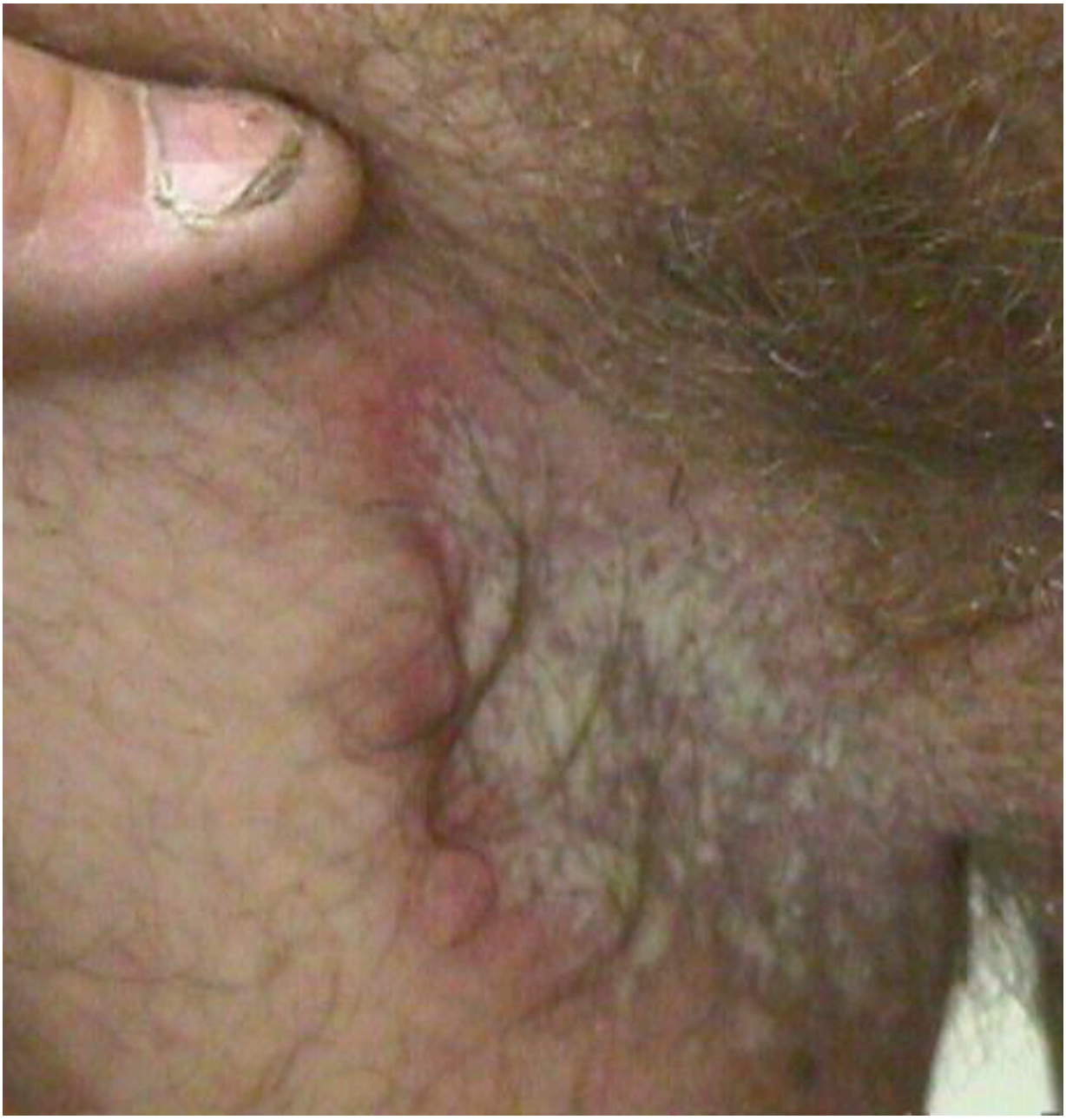
Media file 1: Right axilla with erosive erythematous plaques.



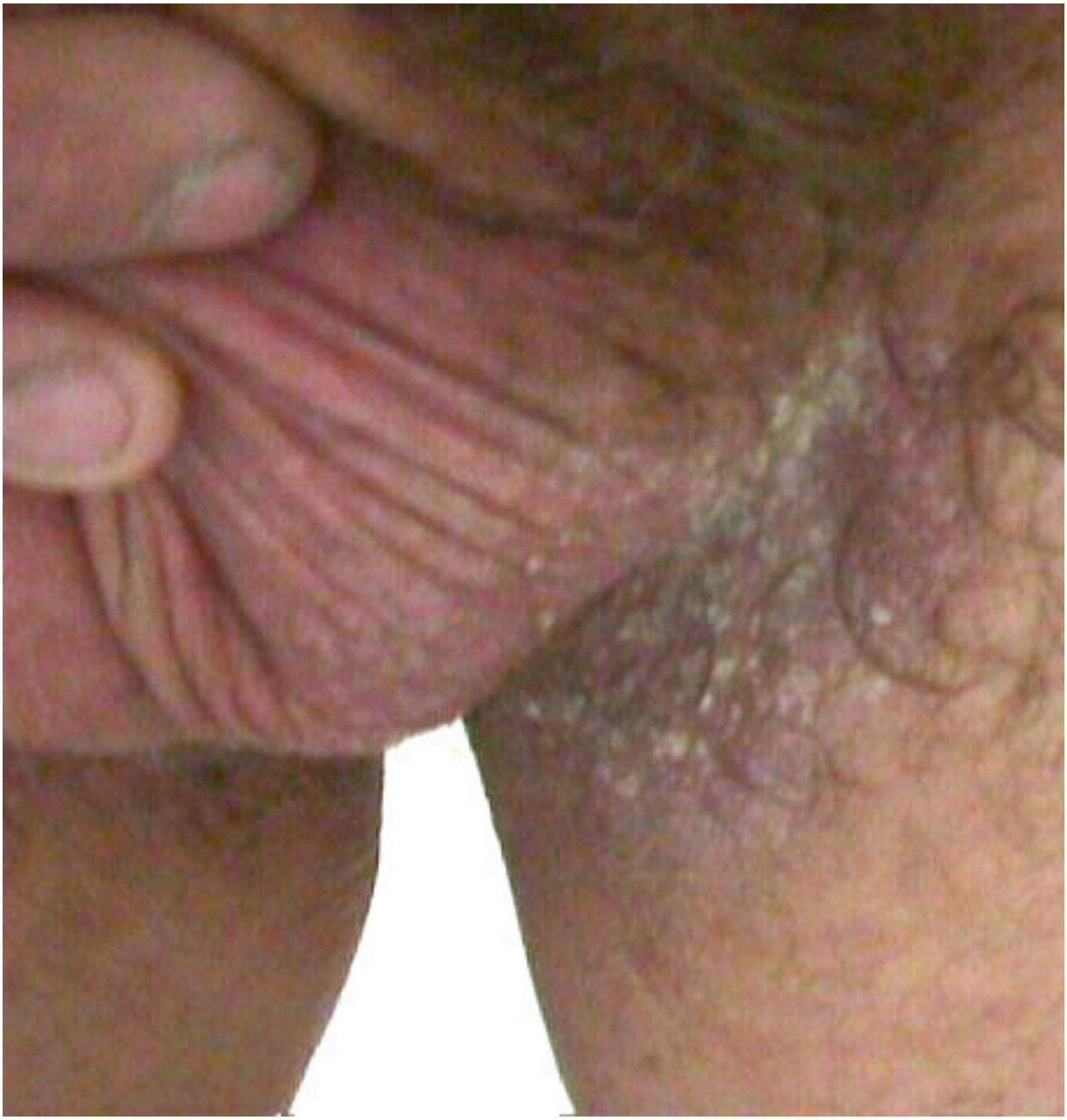
Media file 2: Lumbar back with erythematous plaques with impetiginized crust.



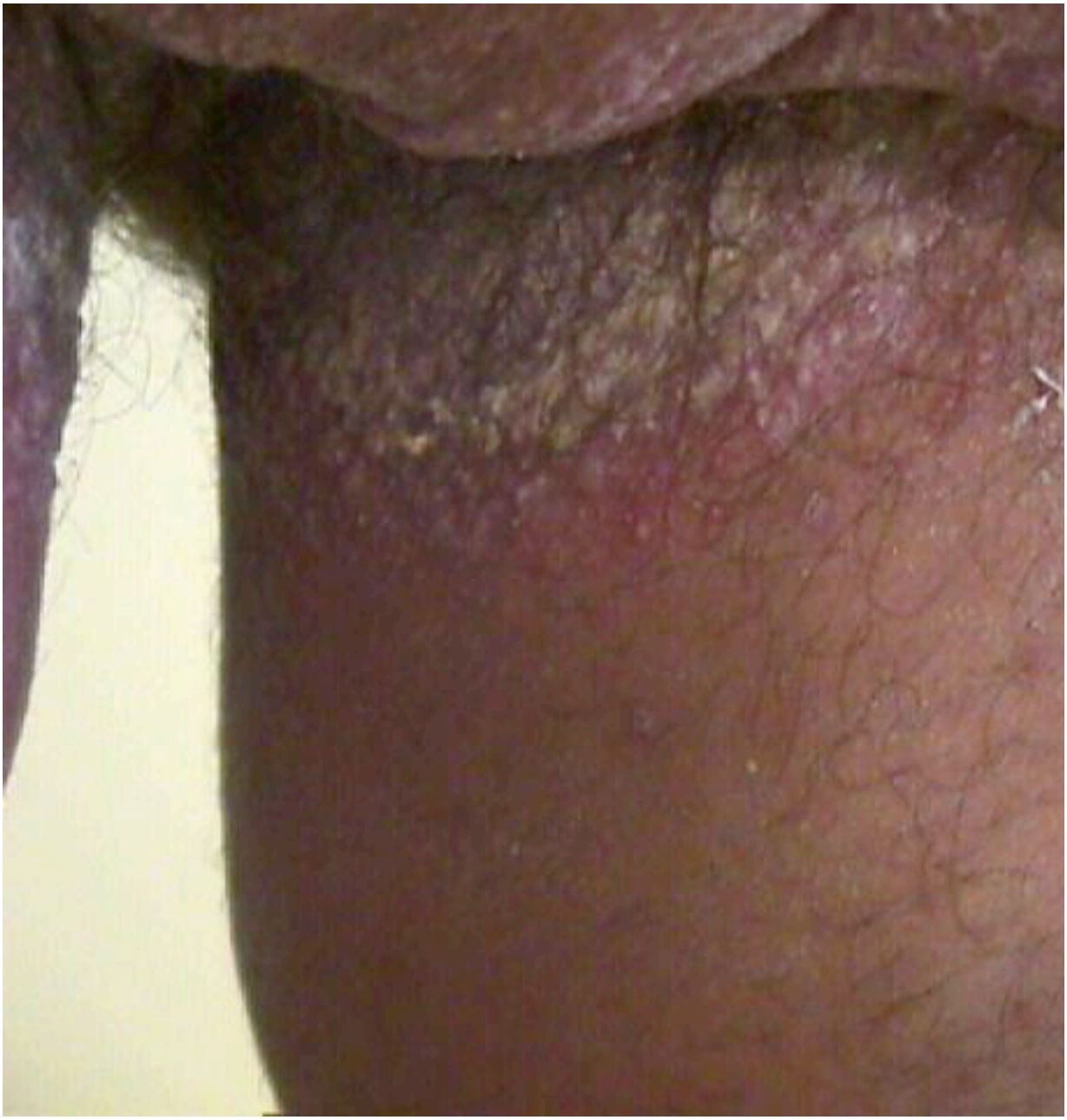
Media file 3: Left axilla with tender erythematous plaques.



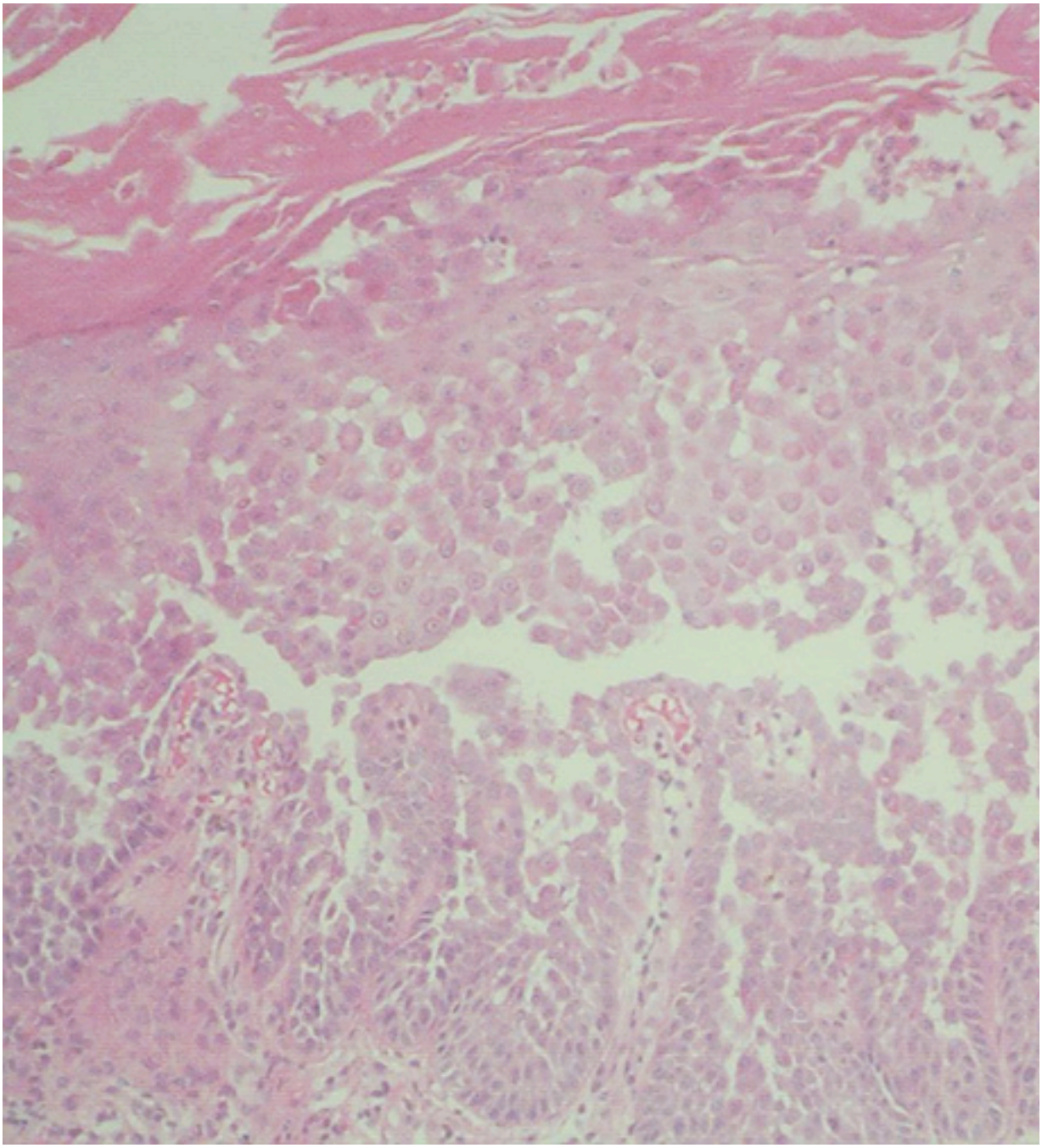
Media file 4: Eroded and crusted plaques in right groin at base of penis.



Media file 5: Erythema of scrotum and erosive plaques in left groin (simulated intertrigo).



Media file 6: Central groin with yellow crust over cleaned plaques.



Media file 7: Acantholysis at all levels of the epidermis (hematoxylin and eosin stain, original magnification X20).

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