Diagnostic criteria in dermatology

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Diagnostic criteria are a set of signs, symptoms and tests which are developed for use in routine practice for the care of individual patients. They need to be broad and must reflect all possible features and severity of a disease.

Diagnostic criteria in dermatology provides dermatologists, residents, medical students and other physicians with the most up-to-date and evidence-based diagnoses for dermatologic diseases and syndromes. For certain dermatoses in dermatology, evidence-based diagnostic criteria have been developed to improve the diagnostic acuity as an accurate diagnosis is essential for proper patient management. Diagnostic criteria in dermatology are recommended for certain dermatosis which is grouped under the heading tabulated in Table 1.

Neutrophilic dermatoses

Sweet syndrome (SS)

Su and Liu[1] in 1986 proposed a set of major and minor criteria for the diagnosis of Sweet syndrome. Von den Driesch[2] in 1994 published a modification of these diagnostic criteria, according to which both major criteria and two minor criteria were required for Sweet syndrome to be diagnosed while in 1996, Walker and Cohen[3] proposed individual diagnostic criteria for drug-induced Sweet syndrome.

The diagnosis of Sweet syndrome is based on a set of criteria that requires the presence of two major and at least two minor criteria.

- For "classical" and "malignancy associated" Sweet syndrome: both major/minor criteria should be present.

Major criteria

- Abrupt onset of painful erythematous papules, plaques or nodules.
- Histopathology showing dense neutrophilic infiltrate without evidence of leucocytoclastic vasculitis.

Minor criteria

- Fever > 38°C (100°F).
- Association with underlying malignancy (haematological/visceral), inflammatory disease/ pregnancy/precede by infection/vaccination.
- Excellent response to treatment with systemic corticosteroid/potassium iodide.
- Abnormal laboratory value (3/4):
  1. ESR >20 mm/h
  2. Positive CRP
  3. TLC >8000/UL
  4. Neutrophils >70%

For "drug-induced Sweet syndrome: the presence of all five criteria is required.

- Abrupt onset of painful erythematous papules, plaque/nodule.
- Histopathology showing a dense neutrophilic infiltrate without evidence of leucocytoclastic vasculitis.
- Fever > 38°C (100°F)
- Temporal relationship between drug ingestion and onset of lesion or temporally related recurrence after oral cholinergic.
- Temporally related resolution of the lesion after drug withdrawal or treatment with systemic corticosteroids.[3]

Pyoderma gangrenosum

In 2004, Su *et al.* proposed diagnostic criteria where both major criteria and at least two minor criteria (out of four) are required to establish the diagnosis.[4]

Major criteria

- Rapid progression of painful + necrolytic cutaneous ulcer with irregular, violaceus and undermined border.
- Exclusion of other causes of ulceration.

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Table 1: Selected dermatological, conditions with diagnostic criteria

Group of disorders	Various dermatological conditions
Neutrophilic dermatoses	Sweet syndrome, pyoderma gangrenosum, Behcet's disease
Vasculitis	Takayasu's arteritis, Churg–Strauss syndrome, Kawasaki disease
Connective tissue disorders	Systemic lupus erythematosus, dermatomyositis, systemic sclerosis
Infective disease	Scabies, Gianotti-Crosti syndrome,
Dermatitis	Atopic dermatitis, irritant contact dermatitis
Immunobullous disorders	Paraneoplastic pemphigus, epidermolysis bullosa acquisita
Leprosy	Lepra reaction
Psoriasis	Psoriatic arthritis
Miscellaneous	Polycystic ovarian syndrome relapsing polychondritis, functional itch disorder, systemic mastocytosis, Ehler-Danlos syndrome, Henoch-SchönleinPurpura, Sezary syndrome, Vogt-Koyanagi-Harada disease, DRESS

Minor criteria

- History suggestive of pathery or clinical finding of cribriform scarring.
- Systemic disease (Inflammatory bowel disease, polyarthritis, myelodysplasia, monoclonalgammaopathy) are associated with PG.
- Histopathology (sterile dermal neutrophils+mixed inflammation+lymphocytic vasculitis).
- Treatment response (rapid response to systemic corticosteroid treatment).

Behcet's disease[5]

Diagnosis of Behcet's disease is based on International Study Group criteria.

The presence of oral aphthosis is mandatory + 2 other items among

- Genital aphthosis,
- Skin manifestation-pseudofolliculitis, erythema nodosum,
- Ocular manifestation-anterior uveitis, posterior uveitis, retinal vasculitis,
- Pathergy phenomenon.

(Vascular manifestation-superficial phlebitis, deep vein thrombosis, large vein thrombosis, arterial thrombosis and aneurysm is not included).

International criteria for Behcet's disease[5]:

- Genital aphthosis and ocular manifestation-two points each.
- Oral aphthosis, skin manifestation, pathergy phenomenon and vascular manifestation- one point each.

Three or >3 points are needed for diagnosis.

Vasculitis

Takayasu's arteritis[6]

American College of Rheumatology criteria (three out of six needed for the diagnosis):

- Age of onset <40 years.
- Angiographic evidence of narrowing or occlusion of the entire aorta or its branch.
- Bruit over subclavian artery or aorta.
- Brachial artery pulse decrease.
- Claudication of an extremity.
- Difference of more than 10 mm Hg systolic BP between two limbs.

Churg–Strauss syndrome

- Asthma or allergic rhinitis.
- Peripheral eosinophilia of more than 10% of total white blood cell count.
- Clinical evidence of vasculitis in one or more extra pulmonary organs like skin, gastrointestinal tract or central nervous system.

All three features required for presumptive clinical diagnosis.[7]

Kawasaki disease

Kawasaki disease research committee guidelines (Japanese guidelines) for diagnosis of Kawasaki disease (2002).[8]

Five of the following six criteria:

- Fever persisting ≥ 5 days.
- Bilateral conjunctival congestion.
- Changes in lips and oral cavity.
- Polymorphous exanthema.
- Changes in peripheral extremities.
- Acute non purulent cervical lymphadenopathy.

American Heart Association guidelines, for diagnosis of Kawasaki disease [9]

Epidemiological case definition (classic clinical criteria) (Patients with fever for at least 5 days and 4 principal criteria can be diagnosed with Kawasaki disease when coronary artery abnormalities detected by 2D echocardiography or angiography.)

- Fever persisting for at least five days.
- Presence of at least four principal features:

Changes in extremities

- Acute: erythema of palms, soles; oedema of hands, feet.
- Subacute: periungual peeling of fingers, and toes in weeks 2 and 3.

1. Polymorphous exanthema.
2. Bilateral bulbar conjunctival injection without exudate.

3. Changes in lips and oral cavity: erythema, lips cracking, strawberry tongue, diffuse injection of oral and pharyngeal mucosae.
4. Cervical lymphadenopathy (>1.5 cm diameter), usually unilateral.

- Exclusion of other diseases with similar findings such as viral infections (e.g., measles, adenovirus, enterovirus, Epstein-Barr virus), scarlet fever, staphylococcal scalded skin syndrome, toxic shock syndrome, bacterial cervical lymphadenitis, drug hypersensitivity reactions, Stevens-Johnson syndrome, juvenile rheumatoid arthritis, Rocky Mountain spotted fever, leptospirosis, mercury hypersensitivity reaction (acrodynia).

Revised criteria for Kawasaki disease[10]

- In the absence of coronary artery disease, all six criteria must be present for a diagnosis.
- In the presence of coronary artery disease at least four out of six criteria necessary for diagnosis.
- High-grade persistent fever unresponsive to antibiotics >5 days.
- Bilateral non purulent conjunctivitis.
- Mucous membrane changes.
- Polymorphus exanthema.
- Changes in extremities.
- Acute non purulent cervical lymphadenopathy.

Connective tissue disorders

Systemic lupus erythematosus

Systemic lupus international collaboration clinics criteria.[11]

Clinical criteria

- Acute or subacute cutaneous lupus.
- Chronic cutaneous lupus.
- Oral/nasal ulcer.
- Non-scarring alopecia.
- Inflammatory synovitis with physician observed swelling of 2 or more joints or tender joints with morning stiffness.
- Serositis.
- Renal: urine protein/creatinine (or 24 hours' urine protein) representing >500 mg of protein/24 hours or red blood cell cast.
- Neurologic: seizure, psychosis, mononeuritis multiplex, myelitis, peripheral or central neuropathy, cerebritis (acute constitutional state).
- Haemolytic anaemia.
- Leucopenia <4000/mm at least once or lymphopenia (<1000/mm at least once).
- Thrombocytopenia (1 lakh/mm at least once).

Immunologic criteria

- above the laboratory reference range.
- Anti-dsDNA above laboratory reference range (except ELISA: twice above laboratory reference range).
- Anti-Sm.
- Antiphospholipid antibody: lupus anticoagulant:
  1. False positive test for syphilis.
  2. Anticardiolipin- at least twice normal or medium high titre.
  3. **Anti-beta 2** glycoproteins.
- Low complement–low C3, low C4, low CH50.
- Direct coombs test in absence of haemolytic anaemia.

At least four criteria including one clinical & one immunological or biopsy-proven lupus nephritis with antinuclear antibody/dsDNA).

American College of Rheumatology criteria for systemic lupus erythematosus:[12]

- Discoid rash.
- Malar rash.
- Recurrent oral ulcer.
- Photosensitivity.
- Non-erosive arthritis.
- Serositis.
- Haematological: anaemia, leucopenia, thrombocytopenia.
- CNS: psychosis, seizure.
- Renal: proteinuria >0.5 gm in 24 hour or RBC cast.
- Antinuclear antibody positivity.
- Immunology: Anti-dsDNA antibody, anti-Smith antibody, false positive Venereal Disease Research Laboratory.

EULAR criteria 2019[13]

All patients must have antinuclear antibody titer of at least 1:80 and 10 points from the criteria [Table 2].

Dermatomyositis

The Bohan and Peter criteria for dermatomyositis and polymyositis[14] includes:

- First, rule out all other forms of myopathies.
- Symmetrical weakness, usually progressive, of the limb-girdle muscles with or without dysphagia and respiratory muscle weakness.
- Muscle biopsy showing necrosis of type I and type II muscle fibres; phagocytosis, degeneration and regeneration of myofibers with variation in myofiber size; endomysial, perimysial, perivascular or interstitial mononuclear cells.
- Elevation of serum levels of muscle-associated enzymes (creatinine kinase, lactate dehydrogenase, transaminases and aldolase).
- Electromyography triad of myopathy.
  a. Short, small, low-amplitude polyphasic motor unit potentials.
  b. Fibrillation potentials, even at rest.
  c. Bizarre, high-frequency repetitive discharges.
- Characteristic rashes of dermatomyositis.

a. Polymyositis definite: All first four elements,
b. Probable: 3 of first 4,
c. Possible: 2 of first 4.
d. Dermatomyositis definite: rashes plus three others,
e. Probable: rashes plus two others,
f. Possible: rashes plus one other.

Systemic sclerosis[15]

If the total score is >9, it is definite scleroderma [Table 3].

The criteria are not applicable to patients with skin thickening sparing finger or to patients who have scleroderma-like disorder that better explain their manifestation (e.g., nephrogenic sclerosing fibrosis, generalized morphea, eosinophilic fasciitis, scleromyxoedema, erythromelalgia, porphyria, lichen sclerosis, graft versus host disease, diabetic cheiropathy).

Infective conditions

Scabies[16]

Suggestive features

- Distribution of lesions.
- Morphology- burrows are pathognomic, but are seen less frequently.
- Nocturnal pruritus.

Table 2: EULAR criteria

Clinical domain	Criteria	Score
Constitutional domain	Fever	2
Cutaneous domain	No scaring alopecia as EULAR criteria.	2
	Oral ulcer.	2
	Subacute cutaneous/discoid ulcer.	4
	Acute cutaneous lupus.	6
Arthritic domain	Synovitis in at least two joints or tenderness in at least two joints & at least 30 minutes of morning stiffness.	6
Neurologic domain	Delirium psychosis seizure	2
		3
		5
Serositis domain	Pleural or pericardial effusion	5
	Acute pericarditis	6
Hematologic domain	Leucopenia	3
	Thrombocytopenia	4
	Autoimmune haemolysis	4
Renal domain	proteinuria > 0.59 g/24 hrs	4
	Class 2 or 5 lupus nephritis	8
	Class 3 or 6 lupus nephritis	10
Immunology domain	**Criteria**	**Score**
Anti-phospholipid antibody domain	Anti-cardiolipin igG > 40 GPL or anti -B2GP1 igG > 40 unit or lupus anticoagulant	2
Complement protein domain	Low C3 or low C4	3
	Low C3 and low C4	4
Highly specific antibody domain	Anti - dsDNA antibody	6
	Anti-smith antibody	

- Contact cases.
- Response to specific therapy.
- Skin biopsy in inflammatory and nodular.

Diagnostic features

- Identification of mite.
- Microscopic study of a skin scraping for mite, eggs, scybala (faeces).
- Skin biopsy- multiple sections (usually required) may demonstrate mite.

Gianotti crosti syndrome[17]

A patient is diagnosed as having Gianotti crosti syndrome or papular ac rodermatitis if:

- On at least one occasion /clinical encounter exhibits all the positive clinical feature.
- On all occasions /clinical encounters related to rash, he/she does not exhibit any of the negative clinical features.
- None of the differential diagnoses is considered to be more likely than the Gianotti crosti syndrome on clinical judgement.
- If lesional biopsy is consistent with Gianotti crosti syndrome.

Positive clinical features

- Monomorphic flat-topped, pink–brown papules/papulovesicle 1–10 mm in diameter.
- At least three of the following four sites: cheek, buttocks, extensor surface of the forearm, extensor surface of legs.
- Being symmetrical.
- Lasting for at least 10 days.

Table 3: Diagnostic criteria for systemic sclerosis

Items	Sub-items	Score
Skin thickening of finger of both hands, extending proximal metacarpus phalangeal joints (sufficient criteria).	–	9
Skin thickening of finger (only count as higher score)	Puffy finger	2
	Sclerodactyly of finger	4
Finger tip lesion (Only count as higher score)	Digital tip ulcer	2
	Finger tip pitting scar	3
Telangiectasia	–	2
Abnormal nail-fold capillary	–	2
Pulmonary arterial hypertension and /or interstitial lung disease	Pulmonary hypertension	2
	Interstitial lung disease	2
Raynaud's phenomenon	–	3
SSC- related auto antibody (antigen centromere, antetopoisomerase 1, anti RNApolymerage 3.	Anticentromere 3	3
	Anti-topoisomerasee 1	
	Anti RNA polymerase 3	

Negative clinical features

- Extensive truncal lesion.
- Scaly lesion.

Dermatitis

Atopic dermatitis

Diagnostic criteria for atopic dermatitis by the Japanese dermatological association.[18]

- Pruritus.
- Typical morphology and distribution:
- Eczematous dermatitis
- Acute lesions: erythema, exudation, papules, vesiculopapules, scales, crusts.
- Chronic lesions: infiltrated erythema, lichenification, prurigo, scales, crusts.
- Distribution
- Symmetrical

Predilection sites: forehead, periorbital area, perioral area, lips, periauricular area, neck, joint areas of limbs, trunk.

- Age-related characteristics.
- Infantile phase: starts on the scalp and face, often spreads to the trunk and extremities.
- Childhood phase: neck, the flexural surfaces of the arms and legs.
- Adolescent and adult phase: tendency to be severe on the upper half of the body (face, neck, anterior chest and back).

Chronic or chronically relapsing course (usual coexistence of old and new lesions):

- More than two months in infancy.
- More than 6 months in childhood, adolescence and adulthood.

A definite diagnosis of atopic dermatitis requires the presence of all three features without consideration of severity. Other cases should be evaluated on the basis of the clinical course with the tentative diagnosis of acute or chronic, non-specific eczema.

Diagnostic standard of Hanifin and Rajika:[19]

Must have three or more basic features described below:

- Pruritus
- Typical morphology and distribution
- Flexural lichenification in adults
- Facial and extensor eruptions in infants and children
- Chronic or chronically relapsing dermatitis
- Personal or family history of atopy (asthma, allergic rhinitis, atopic dermatitis)

Must have three or more following minor features:

- Xerosis.
- Ichthyosis/palmar hyperlinearity, keratosis pilaris.
- Immediate (type I) skin test reaction.
- Elevated serum IgE.
- Early age of onset.
- Tendency towards cutaneous infections (especially *staph. aureus* and *herpes simplex*), impaired cell-mediated immunity.
- Tendency towards non-specific hand or foot dermatitis.
- Nipple eczema.
- Cheilitis.
- Recurrent conjunctivitis.
- Dennie-Morgan infraorbital fold.
- Keratoconus.
- Anterior subcapsular cataracts.
- Orbital darkening.
- Facial pallor, facial erythema.
- Pityriasis alba.
- Anterior neck folds.
- Itch when sweating.
- Intolerance to wool and lipid solvents.
- Peri-follicular accentuation.
- Food intolerance.
- Course influenced by environmental and emotional factors.
- White dermographism, delayed blanch.

UK criteria of atopic dermatitis[20]

Itchy skin condition with three of the following:

- Visible flexural eczema, e.g., antecubital and popliteal fossae (or visible dermatitis of the cheeks and extensor surfaces if under 18 months).
- Personal history of dermatitis as above.
- Personal history of dry skin in the last 12 months.
- Personal history of asthma or allergic rhinitis (or history of eczema in a first-degree relative if <4 years old).
- Onset of signs and symptoms under the age of two years (this criterion should not be used in children <4 years).

Irritant contact dermatitits[21]

Subjective criteria

Major criteria:

- Onset of symptoms within a minute to hours.
- Pain, burning, stiffness or discomfort exceeding itching.

Minor criteria:

- Onset of dermatitis within two weeks of environmental exposure.
- Many people in the environment similarly affected.

Objective criteria:

1. Major criteria:
   - Macular erythema, hyperkeratosis or fissuring predominating over vesicular change.

- Parched or scalded appearance.
- The healing process precedes without plateauupon withdrawal of the offending agent.
- Negative patch test result.

2. Minor criteria:
   - Sharply circumscribed dermatitis.
   - Evidence of gravitational influence, such as a dripping effect.
   - Lack of tendency for the spread of dermatitis.
   - Vesicle, juxtapose closely to patches of erythematous erosion, bullae.

Immunobullous disorders

Paraneoplastic pemphigus[22]

The diagnostic criteria are tabulated in Table 4.

Epidermolysis bullosa acquisita

Diagnostic criteria were updated as follows:[23]

- A bullous disorder within the defined clinical spectrum.
- Histopathology revealing a subepidermal or subepithelial blister.
- A positive DIF microscopy of perilesional skin or mucous membrane with linear IgG, C3, IgA, and/or IgM deposits within the epithelial basement membrane zone.
- Detection of circulating autoantibody against Col7 by immunoblotting, enzyme-linked immunosorbent assay, and/or IDIF microscopy on Col7-expressing human cells.
- Labelling anchoring fibrils by indirect immunoelectron microscopy or negative indirect immunofluorescence microscopy on Col7-deficient skin.
- An u-serration pattern by DIF microscopy.
- Direct immunoelectron microscopy of perilesional skin demonstrating immune deposits within the anchoring fibril zone ± the lower lamina densa.
- *In vivo* bound immune deposits below type IV collagen by fluorescent overlay antigen mapping.
- Alternatively for items 4–8, dermal labelling by direct and/or IDIF microscopy on salt split skin.

Table 4: Anhalt's diagnostic criteria for paraneoplastic pemphigus

Parameter	Criteria
Clinical features	Painful erosions involving mucosa with or without a multiform skin eruption producing blisters and erosions, occurring in association with an occult or evident neoplasm
Histopathology	Suprabasal intraepithelial acantholysis, vacuolar interface changes, necrosis of individual keratinocytes, and/or lichenoid inflammation
Direct immunofluorescence (DIF)	Combined presence of IgG and complement (C3) granular-linear deposition within the epidermal intercellular spaces and along the basement-membrane zone
Indirect immunofluorescence (IDIF)	Presence of circulating antibodies that target the intercellular zone of stratified squamous or transitional epithelia
Immunoprecipitation	Typical complex of proteins, including desmoplakin I (250 kD),bullouspemphigoid antigen (230 kD), envoplakin (210 kD), desmoplakin II (210kD), periplakin (190 kD) and alpha-2-macroglobulin-like-1 (170 kD).

Leprosy

Type I lepra reaction[24]

One major criteria and at least 2 minor criteria are required to establish the diagnosis

Major criteria

- Pre-existing and/or new skin lesion becomes red, inflamed and swollen.

Minor criteria

- One or more nerves become tender or may be swollen.
- Crops of new (painless) lesion appears.
- Sudden oedema of face or extremities.
- Recurrent loss of sensation in hand and feet or sign of recent nerve damage. (Loss of sweating, sensation, muscle strength) in an area supplied by particular nerve.

Type II lepra reaction[25]

One major criteria and at least three minor criteria are required to establish the diagnosis.

Major criteria

- A sudden eruption of tender red, enlarge nodules or plaque which may ulcerate.

Minor criteria

- Mild fever.
- Tender nerve.
- Loss of sensation or muscle power.
- Arthritis.
- Lymphadenitis.
- Epididymo- orchitis.
- Oedema of extremities/face.
- Positive Ryrie or Ellis test.

Psoriatic arthritis

Classification criteria for psoriatic arthritis (CASPAR) criteria[26]

For diagnosis of psoriatic arthritis, patient should have inflammatory articular disease of either joint or spine with three or more of following five points:

- Evidence of current psoriasis, a personal history of psoriasis or a family history of psoriasis.
- Typical psoriatic nail dystrophy including onycholysis, pitting and hyperkeratosis, observed on current physical examination.

- A negative test result for the presence of rheumatoid factor by any method except latex but preferably by **Ellis or Nephelometry.**
- Either current dactylitis or history of dactylitis recorded by rheumatologist.
- Radiographic evidence of juxta-articular new bone formation, appearing as ill-defined ossification near joint margins (excluding osteophyte formation in plain radiograph of hand and feet).

Miscellaneous

Polycystic ovarian syndrome

Modified 1990 national institutes of health/national institute of child health and human disease criteria[27] [Table 5].

ESHRF/ASRM/ROTTERDAM Guidelines[28]

ESHRE- European Society for Human Reproduction and Embryology

ASRM- American Society for Reproductive Medicine

For diagnosis two out of three criteria's should be fulfilled.

- Oligo or chronic anovulation.
- Clinical and/or biochemical sign of hyperandrogenism.
- Polycystic ovaries.
- Twelve or more follicles measuring 2–9 mm in diameter.
- Increase ovarian volume (>10 cm^3).

Relapsing polychondritis[29]

Three out of six criteria's are required for the diagnosis:

- Bilateral auricular chondritis.
- Non-erosive seronegative arthritis.
- Nasal chondritis.
- Ocular inflammation.
- Respiratory chondritis.
- Auriculovestibular damage.

Functional itch disorder[30]

Three compulsory criteria:

- Localised/generalised itching without primary skin lesion.
- Chronic pruritus (>6 week).
- No somatic cause.

Three of seven optional criteria:

- A chronological relationship between the occurrence of pruritus and one or several life events that could have psychological repercussions.
- Variation in intensity with stress.
- Nocturnal variation.
- Predominant during rest/inactivated state.
- Associated psychological disorder.
- Pruritus improved with anti-psychological drugs.
- Pruritus improved with psychotherapy.

Systemic mastocytosis[31]

One major criteria and 1 minor criteria or 3 minor criteria's are required to establish the diagnosis.

Major criteria

- Multifocal infiltrate of mast cell (>15 close to each other observed in bone marrow biopsies and /or mast cell stained for tryptase in biopsies from extracutaneous organ)

Minor criteria

- More than 25% Spindle shaped mast cell in infiltrate in biopsies from bone marrow or other extracutaneous organ presence of >25% atypical mast cell in bone marrow aspirates.
- Demonstration of C-KIT point mutation on codon 816 in bone marrow, blood/extracutaneous organ.
- CD 117 (C-KIT response) positive cell which are also positive for CD2 and/or CD25.
- Serum tryptase >20 microgram/L (not a valid criterion in case of systemic mastocytosis with associated clonal haematological non mast all lineage disease).

Ehler-Danlos syndrome[32]

Bieghton criteria- A score 5/9 or higher defines hypermobility.

- Passive hyperextension of each small finger >90° (1 point each).
- Passive abduction of each thumb to the surface of forearm (1 point each).
- Hyperextension of each knee >10° (1 point each).
- Hyperextension of each elbow >10° (1 point each).
- Forward flexion of trunk with palms on floor and knees fully extended (1 point).

Henoch-Schönlein purpura

American College of Rheumatology, 1990[33]

Presence of two or more following:

- Palpable purpura without thrombocytopenia.
- Patient 20 years or younger at disease onset.
- Bowel angina (diffuse abdominal pain or diagnosis of bowel ischemia).
- Biopsy showing granulocytes in the walls of small arterioles or venules.

Table 5: Diagnostic criteria for polycystic ovarian syndrome

Criterion	Description
Androgen excess	Clinical[a] and/or biochemical hyperandrogenism[b]
Ovarian dysfunction	Oligo-anovulation and/or polycystic ovarian morphology[c]
Exclusion	Other androgen excess or ovulatory disorders[d]

[a] Such as hirsutism, [b] Hyperandrogenemia, such as elevated levels of total or free T, [c] Defined by either the number of intermediate follicles (>8–12 follicles each 2 to 8–9 mm in diameter) and/or an increased ovarian volume (e.g., _10 mL^3), [d] Including, but not limited to, 21-hydroxylase deficient non classic adrenal hyperplasia, thyroid dysfunction, hyperprolactinemia, neoplastic androgen secretion, or drug-induced androgen excess

International consensus conference, 2006[34]

Palpable purpura in the presence of 1 or more following:

- Diffuse abdominal pain.
- Any biopsy showing predominant immunoglobulin A deposition.
- Arthritis (acute, any joint) or arthralgia.
- Renal involvement (any haematuria or proteinuria).

Sezary syndrome[35]

International society for cutaneous lymphomas criteria for diagnosis

- Clinical: erythroderma, generalised lymphadenopathy.
- Histopathologic: acanthosis, dermal fibrosis, striking cerebriform nuclear atypia, minimal epidermotropism and a sparse superficial perivascular infiltrate may be clues. Immunohistochemistry: $CD4^+$ cells.
- Lymph node histology: complete effacement of nodal architecture by infiltrating Sézary cells.
- Immunophenotyping: flow cytometry of peripheral blood shows at least 1000 Sézary cells per mm^3, CD4:CD8 ratio >10, $CD4^+/CD7^-$ >30%, $CD4^+/CD26^-$ >40%, aberrant expression of pan-T-cell antigens.
- Molecular studies: high throughput sequencing of the T-cell receptor beta gene CDR3 region shows clonally related T-cells (Sézary cells) in the skin, peripheral blood and lymph nodes.

Vogt-Koyanagi-Harada disease[36]

A. No history of penetrating ocular trauma or intraocular surgery preceding the initial onset of uveitis
B. Bilateral ocular involvement (time interval between the 2 eyes should be ≤2 wk)
C. No evidence of infectious uveitis or accompanying systemic rheumatic diseases or evidence suggestive of other ocular disease entities[a]
D. Early-phase VKH disease:
    1. Signs of diffuse choroiditis and exudative retinal detachment
    2. Serous retinal detachment on OCT or B-scan ultrasonography
    3. Choroidal thickening on EDI (Enchanced depth imaging)-OCT(optical coherence tomography)[b]
    4. Early punctate staining and late subretinal dye pooling on FFA(Fluoresecence fundus angiography)
    5. Hyperfluorescence of the optic disc on FFA

Definite diagnosis:

Variant 1: In patients presenting with A+B+C+D(1)

Variant 2: In patients without clinically visible exudative retinal detachment, ie, A+B+C+D(2)+D(3) or A+B+C+D(4)

Variant 3: In patients already treated with systemic corticosteroids or combined with other immuno suppressive agents, a history of typical appearances of variant 1 or 2, and A+B+C+D(5)

Late-phase VKH disease

1. Signs of definite sunset glow fundus or retinal pigment epithelium clumping/migration
2. Signs of bilateral recurrent granulomatous anterior uveitis
3. Signs of Dalen-Fuchs nodules or multifocal chorioretinal atrophy
4. Window defects/moth-eaten fluorescence on FFA
5. Previous history of characteristic findings corresponding to diagnosis of early-phase VKH disease

Definite diagnosis:

Variant 1: In patients presenting with A+B+C+E(1)+E(2)

Variant 2: In patients without sunset glow fundus or visible pigment alternations due to early and appropriate treatment, ie, A+B+C+E(2)+E(3) or A+B+C+E(2)+E(4)

Variant 3: In patients with significant media opacity, ie, A+B+C+E(2)+E(5)

[a]This criterion includes (1) no history nor clinical evidence to show ocular tuberculosis, syphilis, or ocular toxoplasmosis; (2) no underlying systemic rheumatic disease that could explain the form of uveitis these patients have; and (3) no history or clinical evidence to suggest the possibility of a specific entity, for instance intraocular tumors, toxic uveitis, Fuchs syndrome, or Posner-Schlossman syndrome.

[b]Ultrasonography can be used to detect the choroidal thickening and therefore may serve as an alternative in the examination where the EDI-OCT is not available, although it is less precise

Drug rash and eosinophilia and systemic symptoms[37]

Bocquet *et al*. proposed criteria for diagnosis of drug rash and eosinophilia with systemic symptoms/drug-induced hypersensitivity.

DRESS is confirmed by the presence of 1 and 2 and 3.

Cutaneous drug eruption.

Adenopathy >2 cm in diameter or hepatitis (liver transaminases >2 times of normal or interstitial nephritis of interstitial pneumonia or carditis).

Hematologic abnormalities eosinophilia > 1.5×10^0/L or atypical lymphocytes.

Dress: Drug rash and eosinophilia with systemic symptoms.

Conclusion

Diagnostic criteria's help to diagnose and differentiate various diseases so that management can be planned accordingly.

Declaration of patient consent

Patient's consent not required as there are no patients in this study.

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Conflict of interest

There are no conflicts of interest.

References

1. Su WPD, Liu HN. Diagnostic criteria for Sweet's syndrome. Cutis 1986;37:167–74.
2. Von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis). J Am Acad Dermatol 1994;31:535–56.
3. Walker DC, Cohen PR. Trimethoprim–sulfamethoxazole–associated acute febrile neutrophilic dermatosis: Case report and review of drug-induced Sweet's syndrome. J Am Acad Dermatol 1996;34:918–23.
4. Su WP, Davis MD, Weenig RH, Powell FC, Perry HO. Pyoderma gangrenosum: Clinicopathologic correlation and proposed diagnostic criteria. Int J Dermatol 2004;43:790–800.
5. Criteria for diagnosis of Behcet's disease. International Study Group for Behcet's Disease. Lancet 1990;335:1078–80.
6. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, *et al.* The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 1990;33:1129–34.
7. Masi AT, Hunder GG, Lie JT, Michel BA, Bloch DA, Arend WP *et al.* The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Rheum 1990;33:1094–100.
8. Han RK, Sinclair B, Newman A, Silverman ED, Taylor GW, Walsh P, *et al*. Recognition and management of Kawasaki disease. CurrPaediatr 1999;8:97–101.
9. Newburger JW, Takahashi M, Gerber MA, Gewitz MH, Tani LY, Burns JC, *et al*. Diagnosis, treatment, and long-term management of Kawasaki disease: A statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Circulation 2004;110:2747–71.
10. Ayusawa M, Sonobe T, Uemura S, Ogawa S, Nakamura Y, Kiyosawa N, *et al*. Revision of diagnostic guidelines for Kawasaki disease (the 5th revised edition). Pediatr Int 2005;47:232–4.
11. Petri M, Orbai AM, Alarcón GS, Gordon C, Merrill JT, Fortin PR, *et al*. Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus. Arthritis Rheum 2012;64:2677–86.
12. Tan EM, Cohen AS, Fries JF, Masi AT, McShane DJ, Rothfield NF, *et al.* The 1982 revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1982;25:1271–7.
13. Aringer M, Costenbader K, Daikh D, Brinks R, Mosca M, Ramsey- Goldman R, *et al.* 2019 European League Against Rheumatism/ American College of Rheumatology classification criteria for systemic lupus erythematosus. Ann Rheum Dis 2019; 78:1151–9.
14. Bohan A, Peter JB. Polymyositis and dermatomyositis (second of two parts). N Engl J Med 1975;292:403–7.
15. van den Hoogen F, Khanna D, Fransen J, Johnson SR, Baron M, Tyndall A, *et al*. 2013 classification criteria for systemic sclerosis: An American College of Rheumatology/European League against Rheumatism collaborative initiative. Arthritis Rheum 2013;65:2737–47.
16. Stokes JH. Scabies among the well-to-do. some principles illustrated by the Élite. JAMA 1938;106:674–8.
17. Chuh AA. Diagnostic criteria for Gianotti-Crosti syndrome: A prospective case-control study for validity assessment. Cutis 2001;68:207–13.
18. Katayama I, Kohno Y, Akiyama K, Ikezawa Z, Kondo N, Tamaki K. Japanese guideline for atopic dermatitis. Allergology International 2011;60:205–20.
19. Hanifin JM, Rajka G. Diagnostic features of atopic dermatitis. Acta Dermatovener (Stockholm) Suppl 1980;92:44–7.
20. Williams HC, Burney PG, Pembroke AC, Hay RJ. The U.K. working party's diagnostic criteria for atopic dermatitis. III. Independent hospital validation. Br J Dermatol 1994;131:406–16.
21. Rietschel RL. Clues to an accurate diagnosis of contacat dermatitis. Dermatol Ther 2004;17:224–30.
22. Mimouni D, Anhalt GJ, Lazarova Z, Aho S, Kazerounian S, Kouba DJ, *et al*. Paraneoplastic pemphigus in children and adolescents. Br J Dermatol 2002;147:725–32.
23. Prost-Squarcioni C, Caux F, Schmidt E, Jonkman MF, Vassileva S, Kim SC, *et al*. International Bullous Diseases Group: Consensus on diagnostic criteria for epidermolysis bullosa acquisita. Br J Dermatol 2018;179:34–41.
24. Naafs B. Treatment and duration of reversal reaction:A reappraisal. Back to the past. Lepr REV 2003;74:328–36.
25. Ankhade VH, Debnath P, Singh RP, Sawatkar G, Bhat DM. A retrospective study of the severe and uncommon variants of erythema nodosum leprosum at a tertiary health center in central India. Int J Mycobacteriol 2019;8:29–34.
26. Taylor W, Gladman DD, Helliwell P, Marchesoni A, Mease P, Mielants H, *et al*. Classification criteria for psoriatic arthritis: Development of new criteria from a large international study. Arthritis Rheum 2006;54:2665–73.
27. Azziz R. Diagnostic criteria for PCOS: A reappraisal. Fertil Steril 2005;83:1343–6.
28. Rotterdam ESHRE/ASRM-Sponsored PCOS consensus workshop group. Revised 2003 consensus on diagnostic criteria and long-term health risks related to polycystic ovary syndrome (PCOS). Fertil Steril 2004;81:41–7.
29. McAdam LP, O'Hanlan MA, Bluestone R, Pearson CM. Relapsing polychondritis: Prospective study of 23 patients and a review of the literature. Medicine (Baltimore) 1976;55:193–215.
30. Misery L, Alexandre S, Dutray S, Chastaing M, Consoli SG, Audra H, *et al*. Functional itch disorder or psychogenic pruritus: Suggested diagnosis criteria from the French psychodermatology group. Acta Derm Venereol 2007;87:341–4.
31. Valent P, Horny H-P, Li CY, et al. Mastocytosis (mast cell disease). In: Jaffe E, Harris N, Stein H, Vardiman J (eds). World Health Organization Classification of Tumours of Haematopoietic and Lymphoid Tissue. IARC Press: Lyon, France, 2001, pp 291–302./.
32. Beighton P, De Paepe A, Steinmann B, Tsipouras P, Wenstrup RJ. Ehlers–Danlos syndromes: Revised nosology, Villefranche, 1997. Ehlers–Danlos National Foundation (USA) and Ehlers–Danlos Support Group (UK). Am J Med Genet 1998;77:31–7. /jrn>.
33. Mills JA, Michel BA, Bloch DA, *et al.* The American College of Rheumatology 1990 criteria for the classification of Henoch-Schönlein purpura. Arthritis Rheum 1990;33:1114–21.
34. Dillon MJ, Ozen S. A new international classification of childhood vasculitis. Pediatr Nephrol 2006;21:1219–22.
35. Olsen E, Vonderheid E, Pimpinelli N, Willemze R, Kim Y, Knobler R, *et al.* Revisions to the staging and classification of mycosis fungoides and Sezary syndrome: A proposal of the International Society for Cutaneous Lymphomas (ISCL) and the cutaneous lymphoma task force of the European Organization of Research and Treatment of Cancer (EORTC). Blood 2007;110:1713–22.
36. Yang P, Ren Y, Li B, Fang W, Meng Q, Kijlstra A. Clinical characteristics of Vogt-Koyanagi-Harada syndrome in Chinese patients. Ophthalmology 2007;114:606–14.
37. Pannu AK, Saroch A. Diagnostic criteria for drug rash and eosinophilia with systemic symptoms. J Family Med Prim Care 2017;6:693–694.