Adult Rash: Bullous Pemphigoid

ICD-9 Codes
694.5 – Pemphigoid

Diagnosis Synopsis
Bullous pemphigoid (BP) is a chronic autoimmune subepidermal blistering disease most frequently seen in the elderly.

The result is the formation of local or generalized, tense bullae. The disease can occur on any body surface, but mucous membrane involvement is rarely seen.

BP has been associated with other autoimmune diseases such as diabetes mellitus, thyroiditis, dermatomyositis, lupus erythematosus, rheumatoid arthritis, ulcerative colitis, myasthenia gravis, and multiple sclerosis. Therapeutic radiation or drugs (furosemide, NSAIDs, captopril, penicillamine, and some antibiotics) have also been associated with BP. It may also follow certain nonbullous inflammatory skin diseases, such as psoriasis and lichen planus, or vaccination (most often in children). In whites, there has been a significant association with the DQB1*0301 allele, whereas Japanese patients have a higher frequency of alleles DRB1*04, DRB1*1101, and DQB1*0302.

Look For
Bullae are tense compared to the flaccid bullae of pemphigus vulgaris. Bullae are most often seen on the lower abdomen, thighs, and forearms. They can be extensive or localized, and there is a flexural predilection. They may appear on normal-looking skin or have an erythematous base. Vesicles or bullae can either be filled with serous or blood-tinged fluid. The lesions typically heal without scarring or milia formation.

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Adult Rash: Henoch-Schönlein Purpura

ICD-9 Codes
287.0 – Allergic purpura

Diagnosis Synopsis
Henoch-Schönlein purpura (HSP) is a small vessel vasculitis of uncertain etiology characterized by IgA-immune complex and C3 deposition in venules, capillaries, and arterioles. Approximately half of patients report a preceding upper respiratory tract infection 1–2 weeks prior to presentation. The classic tetrad of clinical manifestations includes hematuria, colicky abdominal pain, arthritis, and palpable purpura. HSP occurs predominantly in children, but it may be seen in adults and is more common in males. HSP is also seasonal, with most cases occurring during winter.

Additional symptoms include fever, malaise, headache, vomiting, hematemesis, diarrhea, hematochezia, melena, and scrotal pain. An individual episode may persist for 3–6 weeks, and recurrences are frequent. Occasionally, inflammation of the bowel may lead to appendicitis, ileus, and intussusception. Arthritic complaints most commonly involve the ankles and knees.

Renal involvement is common but is usually self-limited, with only a small fraction of patients progressing to chronic renal failure. The risk of renal failure is higher in adult patients and in those presenting with nephrotic or nephritic syndrome.

Look For
Palpable purpura (violaceous erythematous nonblanchable papules) most commonly distributed on the legs and buttocks but occasionally involving the upper extremities, face, and trunk. Urticarial wheals may occur early in the course. Lesions may develop necrotic centers. Localized soft tissue edema of the hands, feet, scalp, ears, or scrotum may also be present.

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Adult Rash: Pemphigus Vulgaris

ICD-9 Codes
694.4 – Pemphigus

Diagnosis Synopsis
Pemphigus vulgaris is a chronic autoimmune intraepidermal blistering disease that can cause significant morbidity and serious complications. IgG autoantibodies are formed to the keratinocyte cell surface molecule desmoglein, creating problems with cell-cell adhesion (acantholysis). Bullae and erosions develop on skin and mucous membranes. Severe cases can be life threatening. Complications are related to the use of high dose steroids, secondary infection, loss of the skin barrier, and poor oral intake. The disease appears equally in men and women, often in their fifties and sixties. Pemphigus vulgaris occurs in all races but may be more common among those of Jewish ancestry. It is also associated with other autoimmune diseases, especially myasthenia gravis.

Look For
Flaccid bullae, erosions, and crusts, which can be quite thick. The vesicles and erosions can affect mucosal surfaces, including conjunctiva and oral mucosa. Large areas of epidermis can denude, creating risk for bacterial infection and sepsis. Look for active vesicles at the margins of plaques.

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Adult Rash: Stevens-Johnson Syndrome

ICD-9 Codes
695.1 – Erythema multiforme

Diagnosis Synopsis
Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are two rare severe drug reactions that are characterized by high fevers, skin tenderness, mucosal erosions, and skin detachment about 1–3 weeks after the inciting medication is started. SJS and TEN should be considered the same disease on opposite ends of the spectrum, with the level of skin detachment differentiating these two entities. SJS is characterized by less than 10% of body surface area (BSA), while TEN involves greater than 30% BSA. SJS-TEN overlap occurs when 10–30% BSA is involved. Note that SJS can rapidly evolve into TEN, and both can have an unpredictable clinical course. Even expert groups may identify patients in the overlap area, and this can complicate decision making for therapy.

SJS/TEN can affect all ages and races, with a slight preponderance seen in women (1.5:1) and an increasing incidence with age. Risk factors that confer a worse prognosis include extent of body surface area involved at time of diagnosis, older age, cancer or hematological malignancy, AIDS, number of medications, and elevated serum urea, glucose, and creatinine levels.

SJS and TEN run an unpredictable clinical course. The primary lesions include dusky red macules of irregular size and shape that start on the trunk and spread to the proximal extremities, neck, and face. The onset of disease occurs 1–3 weeks after the ingestion of an antibiotic or within the first 2 months of anticonvulsant treatment. Within hours to days, the epidermis can detach from the dermis, with serosanguineous fluid filling the space and subsequent flaccid blister formation. Ocular, oral, and genital mucosa will be affected in more than 90% of cases. Mucosal symptoms that should be screened for include painful eyes, painful swallowing, dysuria, and diarrhea.

Look For
Specifically for SJS:

- Less than 10% BSA.
- Isolated, irregularly shaped, dusky red macules on the trunk, face, and palms/soles.
- Atypical target lesions may also be seen; they are atypical in the sense that they do not have the characteristic 3 concentric rings seen in erythema multiforme.
- "Scalded" skin – Epidermal detachment with underlying bleeding dermis exposed.
- Flaccid bullae due to epidermal-dermal detachment.
- Nikolsky sign – Tangential mechanical pressure of an erythematous macule leads to epidermal-dermal detachment.
- Mucosal involvement including the eyes, lips, mouth, and genitalia.
- Look for hemorrhagic crust, bullae, and denudation in these areas.
- Systemic symptoms are commonly present but not invariably (as seen with TEN).

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http://www.visualdx.com/visualdx/visualdx6/getDiagnosisTextPrint.do?moduleId=7&diag...
Adult Rash: Staphylococcal Scalded Skin Syndrome

ICD-9 Codes
695.81 – Ritter's disease

Diagnosis Synopsis
Staphylococcal scalded skin syndrome (SSSS), or Ritter's disease, is a toxin-mediated infection that is characterized by skin tenderness, flaccid bullae, and skin detachment. A prodrome of fever, sore throat, malaise, and irritability accompanied by purulent rhinorrhea and/or conjunctivitis often occurs prior to the onset of bullae and desquamation. SSSS is mainly a disease of infants and in children younger than 6 years, with a higher incidence seen in males. Among adults, risk factors for SSSS include immunosuppression and chronic renal insufficiency.

The natural history of SSSS is characterized by the following:

- Prodromal symptoms and/or purulent rhinorrhea and/or conjunctivitis
- Facial erythema that generalizes to the body in less than 48 hours
- Bullae development, positive Nikolsky's sign, very tender skin
- Skin wrinkling and epidermal sloughing within 48 hours after bullae develop
- Desquamation continues for up to 5 days
- Re-epithelialization, without scarring, completed over following 2 weeks

Look For
The skin findings begin as a scarlatiniform eruption. Flaccid blisters may develop within 24–48 hours. Nikolsky's sign (disrupting the epidermal barrier with firm rubbing) is positive, and large sheets of epidermis are shed. Circumoral erythema becomes crusted after a few days. Mild facial edema, lip fissuring, and purulent conjunctivitis can occur. The erythema is accentuated in periorificial and flexural areas.

Involvement is generally widespread.

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Adult Rash: Toxic Epidermal Necrolysis

ICD-9 Codes
695.1 – Erythema multiforme

Diagnosis Synopsis
Toxic epidermal necrolysis (TEN) and Stevens-Johnson syndrome (SJS) are 2 rare severe drug reactions that are characterized by high fevers, skin tenderness, mucosal erosions, and skin detachment about 1–3 weeks after the inciting medication is started. SJS and TEN should be considered the same disease on opposite ends of the spectrum, with the level of skin detachment differentiating these 2 entities. SJS is characterized by less than 10% of body surface area (BSA), while TEN involves greater than 30% BSA. SJS–TEN overlap occurs when 10–30% BSA is involved. Note that SJS can rapidly evolve into TEN, and both may follow an unpredictable clinical course. Even expert groups may identify patients in the overlap area, and this can complicate decision making for therapy.

SJS/TEN can affect all ages and races, with a slight preponderance seen in women (1.5:1) and an increasing incidence with age. Risk factors that confer a worse prognosis include extent of BSA involved at time of diagnosis, older age, cancer or hematological malignancy, AIDS, number of medications, and elevated serum urea, glucose, and creatinine levels.

Primary lesions include dusky red macules of irregular size and shape that start on the trunk and spread to the proximal extremities, neck, and face. The onset of disease occurs 1–3 weeks after the ingestion of an antibiotic or within the first 2 months of anticonvulsant treatment. Within hours to days, the epidermis can detach from the dermis, with serosanguineous fluid filling the space and subsequent flaccid blister formation. Ocular, oral, and genital mucosa will be affected in more than 90% of cases. Mucosal symptoms that should be screened for include painful eyes, painful swallowing, dysuria, and diarrhea.

Rapid identification and withdrawal of the offending drug and transfer to a burn unit with aggressive supportive care are the most critical steps in the management. The fatality rate of TEN approaches 35%. Death is usually due to sepsis, adult respiratory distress syndrome, gastrointestinal bleeding, or pulmonary embolism. A few days after the epidermis is sloughed, re-epithelialization begins. Most of the epidermis is re-epithelialized after 2–3 weeks with no scarring over most areas. Mucous membrane erosions may persist for months, however.

Look For
- Greater than 30% BSA.
- Irregularly shaped, dusky red macules on the trunk and face and palms/soles largely coalescing.
- Atypical target lesions may also be seen (ie, atypical in the sense that they do not have the characteristic 3 concentric rings seen in erythema multiforme).
- "Scalded" skin – epidermal detachment with underlying bleeding dermis exposed.
- Flaccid bullae due to epidermal–dermal detachment.
- Nikolsky’s sign – tangential mechanical pressure of an erythematous macule leads to epidermal–dermal detachment.
- Mucosal involvement including the eyes, lips, mouth, and genitalia – Look for hemorrhagic crust, bullae, and denudation. Mucous membrane changes occur at these sites in descending order of frequency: the oropharynx, eyes,
genitalia, and anus. Greater than 90% of patients experience mucous membrane involvement with painful erosions. In one-third of cases, mucous membrane changes precede cutaneous changes by 1–3 days. Pseudomembranous conjunctival erosions and hemorrhagic crusting of the lips are common findings.

- Systemic symptoms are invariably present.

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