Emergencies in Dermatology : Acute Skin Failure

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Abstract

Rational understanding of etiopathogenesis of the systemic complications arising out of sudden, severe alterations in structure and function of the skin consequent to the syndrome of acute skin failure clearly establishes the necessity of a dedicated ICU in a skin department.

Immune suppression due to increased age, organ transplantation, malignancy, prolonged intake of steroids as also the indiscriminate use of drugs have lead to a spurt in the incidence of widespread, recalcitrant dermatoses with significant potential to eventuate into reaction patterns terminating into acute skin failure, viz. universal erythema and scaling of erythroderma and widespread denudations of bullous dermatoses. Prompt intensive management of all such cases in the ICU on the lines of 100% burns is mandatory.

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Key Words: Acute skin failure; Erythroderma; Bullous dermatoses; Recalcitrant dermatoses.

Introduction

Prof Rene Touraine first established an ICU in a skin department in 1974. It has now been recognized as a necessity due to a large number of extensive skin diseases eventuating into the potentially fatal syndrome of 'acute skin failure' which fulfills the definition of an emergency, i.e. 'a risk perceived by a doctor or a patient to life, limb or the structure/ function of an important organ of the body' [1].

Besides the physiological roles of skin, alterations in the colour and texture of this 'interface that stares in the face' are no less significant. Disfiguring dermatoses can generate emergency situations due to their disproportionate and spectral psychocutaneous morbidity consequent to disruption of the psychosocial function of self worth and emotional expression subserved by this largest body organ.

Various dermatological emergencies can disrupt the interconnected anatomy and physiology of skin resulting in a number of complications known collectively as acute skin failure. Understanding of the etiopathogenesis of this entity can save many lives by prompt institution of appropriate treatment particularly in cases of those at increased risk, viz. extremes of age, extensive involvement, taking high dose of glucocorticosteroids/ immunosuppressants, neutropenia, renal transplant recipients and having recalcitrant underlying/ concomitant diseases etc.

Emergencies in STD, Leprosy & Dermatology

Due to the prevalent misunderstanding and discrimination, the mere suspicion of acquiring STD especially HIV evokes extreme psychological anxiety that can lead to suicidal ideation and worsening illness by delay in logical course of action. Paraphimosis, phimosis, phagedenic ulceration, bubo formation, rupture of dorsal artery of penis etc. are common emergencies. Penicillin therapy in syphilis can cause Jarisch-Herxheimer reaction whose consequences can be fatal in late syphilis.

Leprosy evokes extreme fear not only because of associated stigma but also due to various permanent deformities due to acute inflammatory episodes (lepra reactions) leading to acute neuritis, eye and testicular involvement etc. These emergencies especially acute neuritis of ulnar, common peroneal and facial nerves should be actively looked for and treated promptly to obviate permanent damage. Dapsone syndrome, acute abdomen due to clofazimine and 'flu' like syndrome due to rifampicin are serious adverse effects of commonly used antileprosy drugs.

Anaphylactic, exanthematous, acute eczematous, urticarial / angioedematous, purpuric and photosensitive reactions can all be spectral as can be the vasculitic conditions of Henoch-Schonlein purpura, purpura fulminans and pyoderma gangrenosum. Stevens-Johnson syndrome-Toxic Epidermal Necrolysis (SJS-TEN)

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complex, secondary to drugs in over 95% of cases, can occur with dramatic ferocity as can be the extensive skin denudations due to immunobullous eruptions and staphylococcal scalded skin syndrome (SSSS). Universal reddening and scaling of exfoliative dermatitis can be the end result of drug reactions, malignancies as well as many skin disorders, of keratinisation, like pustular psoriasis, erythrodermic psoriasis, ichthyosiform erythroderma, etc. Diseases of the new born like purpura fulminans, lamellar desquamation, infantile acute hemorrhagic oedema, sclerema neonatorum, congenital absence of skin and metabolic disorders like porphyrias can be serious emergencies.

Etiopathogenesis of acute skin failure [2,3,4] involves failure of skin to perform its multiple functions can lead to acute failure of heart, lung, kidney and death consequent to structural and functional alterations in various components of the skin.

Destruction of stratum corneum, the layer mainly responsible for the barrier function of the skin, can cause up to 40 times increase in fluid loss than the normal rate of 01 ml/cm²/hour. 50% body surface area (BSA) involvement leads to daily fluid loss of up to 4-5 liters. Loss of proteins (40 gm/L), Na (120-150 mmol/L), Cl (10-90 mmol/L) and K (5-10 mmol/L) in the bullous fluid leads to decrease in intravascular volume. The resultant decrease in urinary output and increased blood nitrogen can lead to renal failure unless treated energetically [5].

Damaged skin and its exudates support growth of a wide spectrum of endogenous and exogenous organisms leading to systemic infection, severe sepsis and death. Altered immunological function due to damage to the skin structure promotes development of sepsis.

Impaired thermoregulation can cause either hyper- or hypothermia depending on the surrounding environment. Shivering due to increased interleukin-1 production, reflects need to maintain higher central core temperature. Hypothermia, a bad prognostic marker indicates severe sepsis and shock.

Hypercatabolic state increases energy expenditure by 2-4 times. A lower environmental temperature increases this expenditure further. Loss of proteins in the exudates leads to hypoalbuminaemia. Inhibition of insulin secretion and insulin resistance lead to hyperglycemia and glycosuria, which cause amino acid breakdown leading to further worsening of hypercatabolic state and condition of the patient.

Increased cutaneous blood flow nearby doubles the cardiac output and may prove fatal, particularly in the elderly and in those with previous cardiac disease.

Management of acute skin failure

Prompt initiation of appropriate treatment on the lines

of a 100% burns patient and excellent double barrier nursing care are the twin principles of management that can salvage many lives.

Observation should be meticulous with heart rate, pulse rate, urinary volume (50-100ml/hr) monitored hourly and urinary osmolarity (less than 1020), glycosuria, temperature, gastric contents monitored 3-4 hourly. Any change in extent of skin lesions and body weight should be noted daily along with calculation of fluid loss.

Daily arterial blood gas analysis, complete blood count, blood urea, creatinine, glucose, electrolytes, albumin, LFT, complete urine examination and chest radiograph are essential. Culture from skin lesions and venous line alternate day is desirable for antibiotic therapy.

Correction and maintenance of haemodynamic and electrolyte equilibrium by fluid and electrolyte administration is of prime importance. Fluid requirement during first 24 hours is isotonic saline 0.7ml/kg/% of body surface area (BSA) affected and human albumin 1ml/kg/% BSA. Potassium phosphate is added to I/V fluids to prevent insulin resistance. About 1500ml of nasogastric feed can be given in addition on first day. Subsequently depending on the progress oral feeds are increased and I/V fluids are reduced gradually.

Aggressive nutritional support is required to compensate the hypercatabolic state and to promote tissue healing. Energy requirement in adults is 1500-2000 Kcal in first 24 hrs; with an increment of 500 Kcal daily up to 3500-4000 Kcal/day. Protein intake of 2-3 gms/kg/day (3-4 gms/kg in children) should help in faster healing.

Judicious use of antibiotics is a must to avoid strain selection, fungal infection and drug reactions. Sudden rise or fall of temperature, deterioration of consciousness, oliguria, accelerated pulse, tachypnoea, increase in insulin requirement and gastric residual volume indicate need for antibiotics in absence of pus/ blood culture results.

Topical antiseptics like silver sulphadiazine (contraindicated in patients sensitive to sulpha drugs) should be applied after proper bath/soaks with potassium permanganate solution.

Nursing in a room at temperature of 30-32°C, in an air-fluidized bed is helpful. Care of mucous membranes like eyes, nose, mouth, genitals, etc., is essential to prevent morbidity and infections.

Specific therapy depends on the underlying cause.

Common dermatoses leading to acute skin failure

The incidence of acute skin failure, the end result of large number of dermatoses and adverse severe

reactions to drugs, is on the rise. Some common antecedent dermatoses that can eventuate into this syndrome are described below.

Erythroderma (Exfoliative dermatitis): Erythema and scaling involving most of the body surface area, develops either *de novo* (primary or idiopathic) or as a progression of a preexisting skin disease (secondary). Psoriasis is the commonest antecedent illness in approximately 40% of cases and 20% are secondary to eczemas. Miscellaneous causes like ichthyosis, pemphigus, TEN, SSSS, crusted scabies and lymphomas accounts for 10% of cases [6,7]. Intakes of drugs like sulphonamides, dapsone, NSAIDs, antiepileptics, penicillins etc. is the cause in about 10-15 % of cases. A small proportion of cases are idiopathic [8].

Management consists of treatment of the underlying cause (if diagnosed), and that of acute skin failure induced systemic problems [9].

Bullous diseases: Immunobullous diseases like pemphigus, pemphigoid, etc. and hereditary mechanobullous disorders like epidermolysis bullosa can be disabling and even life-threatening in some cases.

Pemphigus vulgaris: There are three main types of pemphigus- P foliaceus, (the blister is in the superficial granular layers), P vulgaris, (the blisters form just above the basal layer) and paraneoplastic pemphigus that occurs in association with malignancy.

Though flaccid blisters are the primary lesions of pemphigus, erosions are common and usually painful. Nikolsky's sign is positive. Oral mucosal involvement is universal and may be the only involvement for an average of 5 months. Painful and difficult swallowing helps to serve as a mode of presentation.

The usual mortal outcome of pemphigus during the pre-steroid era has now been reversed to less than 10%, patients, succumbing mostly to the side effects of high dose steroids. The patients who survive the disease for five years are considered cured. Dexamethasone, cyclophosphamide pulse therapy (DCP) introduced by Pasricha is increasingly being accepted as a standard steroid sparing regimen [10].

Bullous pemphigoid: This subepidermal blistering skin disease of the elderly is characterized by large, tense bullae arising on normal or erythematous skin. Eroded skin tends to reepithelise. Early inflammatory disease presents with 'urticated' lesions. Oral mucosal erosions in 10-15% cases also heal without scarring. Drugs like penicillin, frusemide, sulfasalazine, penicillamine, captopril, etc. local trauma, burn wounds, grafts, irritants, UV radiation & malignancies like carcinoma stomach can precipitate this immunobullous disease. Biopsy of a fresh small blister shows diagnostic changes of a

subepidermal blister and a dermal infiltrate containing eosinophils, lymphocytes and histiocytes. The disease presentation being spectral, treatment has to be individualized. In extensive disease, oral prednisolone is given in starting dose of 60 mg per day and is tapered at the earliest. Steroid sparing immunosuppressive agents include DCP therapy, plasmapheresis, IVIg and high dose IV methylprednisolone pulse.

Erythema multiforme (EM): is an acute, self-limiting, mucocutaneous reaction pattern to many viral, bacterial, protozoal and fungal infections, tumors, drugs, autoimmune states and miscellaneous conditions. HSV infection is the most frequent cause followed by mycoplasma pneumoniae.

Clinical spectrum of EM ranges from mild (erythema multiforme minor) to severe form (Steven-Johnson's syndrome-TEN complex and TEN) consists of variable prodromal symptoms and a symmetrically distributed polymorphic rash classically with iris or target lesions seen on hands with a central vesicle, or erythema surrounded by a pale and then a red ring. Mucosal lesions occur in 25-65% cases. The eruption in Stevens-Johnson syndrome (Severe EM) occurs preferentially periorificially or on mucocutaneous locations as painful erosions with thick adherent crusts.

Toxic Epidermal Necrolysis (TEN) - Extensive denudation of skin is hallmark of TEN, which has a very high mortality rate, ranging from 15 to 40%. Malaise, pruritus, fever, prostration and myalgia are common. Severe erosions of oropharynx may be painful enough to prevent eating or talking. Ocular involvement may lead to keratitis, corneal ulceration and its resultant complications.

Besides management of acute skin failure as mentioned above multiple therapeutic modalities have been tried. Role of systemic corticosteroids continues to be debated and most agree that they have no role after initial period of management. Immunosuppressive drugs including cyclosporin have been tried with variable results. IVIG is being used in many centers with good response but some have reported increased mortality [11,12].

Conclusion

Sudden severe alterations in the anatomy and physiology of skin consequent to generalized dermatoses can lead to disabling complications eventuating in the potentially fatal syndrome of acute skin failure. Understanding the etiopathogenesis of various systemic complications of acute skin failure and their prompt management in ICU on lines similar to that of burns can salvage many lives.

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Answer to ECG Quiz

Discussion

This ECG shows wide complex tachycardia (WCT) which refers to cardiac rhythm of >100 bpm with a QRS duration of 120 msec or more on surface ECG [1]. The QRS duration in this ECG is 300 msec and RR duration is 344 msec corresponding to a heart rate (HR) of 174 bpm. There is right bundle branch block (RBBB) with frontal plane QRS axis of +135, q in VI with uniform widening of initial and terminal QRS complex and QS complexes in V2-V6. The morphology of QRS in chest leads is that of a sine wave pattern.

The differential diagnosis of WCT includes ventricular tachycardia (VT), supraventricular tachycardia (SVT) with aberrancy or an underlying bundle branch block and antidromic pre-excited tachycardia.

In an old MI, any WCT is likely to be a VT until proved otherwise and should be managed as such. However, in this case, the sine wave pattern of QRS complex with uniform widening suggests hyperkalaemia [2]. Patient was haemodynamically stable and hence serum potassium was done to corroborate the ECG diagnosis. Serum potassium was 9 meq/L, blood urea 235 mg/dl and serum creatinine 8.6 mg/dl. Emergency therapy for hyperkalaemia was started with intravenous calcium gluconate and glucose-insulin infusion and patient was dialysed.

ECG done a few hours later is shown in Fig. 2 which shows underlying RBBB, Q waves in V1-V5, absence of sinus P waves and presence of retrograde P waves suggestive of junctional rhythm. Serum potassium at that

Fig. 2 :

time was 7.8 mcq/L. These patterns of ECG should always be recognized and remembered by the treating physician, as it has therapeutic significance. The therapeutic modality in this case was treatment for hyperkalaemia and not a rush for electrical or pharmacological cardioversion. The patient had bilaterally contracted kidneys on subsequent evaluation which was most likely due to long standing systemic hypertension resulting in CRF and hyperkalaemia.

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