Case study: Anaesthesia implications and considerations in a case of pemphigus vulgaris for orthopaedic bipolar prosthesis implant surgery

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Abstract
A 60-year-old patient suffering from pemphigus vulgaris for the past year was admitted to the emergency ward for fracture neck of femur. She also presented with lesions involving oral mucosa, back, inframammary and genital areas which were in partial remission. In hospital she was diagnosed with hypertension and was put on anti-hypertensives. Special attention was paid during positioning for surgery, administration of regional anaesthesia and placement of the intravenous line as well as monitoring devices. General anaesthesia was avoided in the presence of partially active oral lesions. Combined spinal-epidural anaesthesia was administered using bupivacaine-clonidine mixture. No haemodynamic complication was observed with 30 µg of clonidine intrathecally and no skin lesion occurred at the site of injections or Tegaderm application.

Case report

A 60-year-old woman weighing 86 kg had a one-year history of pemphigus vulgaris (PV) lesions involving the arms, back, chest, oral mucosa and abdomen. She was admitted to the emergency ward with chief complaints of pain in the right leg after a fall which was subsequently diagnosed as fracture neck of femur. At the time of surgery, she presented with multiple PV lesions involving the oral mucosa, back, inframammary area as well as vulval region which were in partial remission. She had been on oral and injectable steroids for the past year and complained of relapse following tapering of steroids. She was put on tab. deflazacort 30 mg twice a day which was tapered to 30 mg once daily. The rest of the treatment included cap. Bactoclav (amoxicillin + clavulanate potassium) 625 mg twice daily, Clonate F cream, tab. Azathioprine 50 mg once daily and Hexidine mouth wash. Her ECG showed P-pulmonale, ST and T wave changes in anterior and inferior leads but echocardiography findings were within normal limits with an ejection fraction of 55%, grade 1 diastolic dysfunction and no septal wall motion abnormality. The remaining investigations were within normal limits.

Preanaesthetic assessment revealed an obese woman with a Mallampati Class II airway with buccal lesions in remission. She was diagnosed as suffering from hypertension also in the hospital only and was put on tab. amlodipine 5 mg once daily, tab. ramipril 5 mg and tab. Aquazide 25 mg once daily. In the operation theatre, preloading was done with 750 ml of lactated Ringer's solution. The local area of skin at L2–3 and L3–4 level was infiltrated with 1 ml of 2% lignocaine hydrochloride solution while avoiding infiltration into the superficial subcutaneous tissues. An epidural catheter was secured at L3–4 level with an 18-gauge Tuohy needle by keeping the patient in the sitting position and taking all aseptic precautions. A lumbar puncture was performed at a lesion-free area (L2–3) with a 26-gauge Whitacre needle, and a subarachnoid block was initiated using 12.5 mg of bupivacaine HCl and 30 µg of clonidine with a total diluted volume of 3.2 ml. The catheter was secured to the patient's back with Tegaderm adhesive after consultation with a dermatologist. The surgical procedure commenced after establishing the sensory level at the T6–7 dermatome. The intraoperative surgical and anaesthetic course was uneventful and surgery lasted for two hours.

After surgery the patient was transferred to the recovery room and monitored for two hours. The patient was fully conscious and did not have any nausea, vomiting or haemodynamic complications. Epidural top-ups were given for three days with 8 ml of 0.125% of bupivacaine HCl three times daily. The postoperative

stay in hospital was uneventful. Medication for PV was resumed six hours after surgery. The patient was discharged from hospital after seven days with no complication associated with the site of regional anaesthesia or at the site of Tegaderm application.

Discussion

The term pemphigus refers to a group of autoimmune blistering diseases of the skin and mucous membranes characterised histologically by intraepidermal blister and immunopathologically by the finding of in vivo bound and circulating immunoglobulin G (IgG) antibodies directed against the cell surface of keratinocytes. The three primary subsets of pemphigus are pemphigus vulgaris (PV), pemphigus foliaceus and paraneoplastic pemphigus. PV accounts for approximately 70% of pemphigus cases. Blisters in PV are associated with the binding of IgG autoantibodies to keratinocyte cell surface molecules. These intercellular or PV antibodies bind to keratinocyte desmosomes and to desmosome-free areas of the keratinocyte cell membrane. The binding of autoantibodies results in a loss of cell-cell adhesion, a process termed acantholysis.[1] PV involves mucosa in 50 to 70% of patients. Blistering and erosions secondary to the rupture of blisters may be painful and limit the patient's daily activities. Patients with PV typically heal without scarring unless the disease is complicated by severe secondary infection.[2]

Both regional and intravenous anaesthesia have been used in pemphigus; however, there have always been attempts to avoid airway instrumentation.[3] Skin lesions can also predispose a patient to fluid and electrolyte abnormalities (dehydration and hypokalaemia). Sepsis and skin infection at the site of local anaesthesia injection is possible. Infiltration with local anaesthesia is better avoided because of risk of skin sloughing and bullae formation at the injection site.[4]

Literature and academic evidence recommends avoiding infiltration with local anaesthetic prior to spinal anaesthesia, but in this case the local anaesthetic was injected inside deep tissues while avoiding any infiltration into the skin layers. Management of such patients can be complicated further by drug therapy for pemphigus. Preoperative steroid therapy can produce hypothalamicopituitary-adrenocortical insufficiency. This requires perioperative steroid coverage to avoid Addisonian crisis.[5] Other immunosuppressive drugs and steroids predispose patient to infections, bone marrow suppression and cardio-respiratory side effects.[6]

Intrathecal clonidine has been shown to result in the prolongation of the sensory blockade and a reduction in the amount or concentration of local anaesthetic required to produce perioperative analgesia. The addition of clonidine to intravenous regional anaesthesia resulted in prolongation of the tourniquet time and improvement of postoperative analgesia.[7] The addition of clonidine to intrathecal hyperbaric bupivacaine prolongs the duration of motor block and improves the quality of the block.[8]

The Koebner (isomorphic) phenomenon refers to the appearance of new lesions in traumatised but otherwise normal skin in patients with certain skin diseases such as vitiligo, psoriasis and lichen planus following trauma; these new lesions are identical to those in the diseased skin. The Koebner phenomenon may occur in recent scar or pressure points.[9] This has also been reported in pemphigus vulgaris.[10] Neuraxial opioids have been used in cases of pemphigus and are associated with pruritis, especially morphine.[11] In this case intrathecal clonidine was used because it is free of some of the side effects of opioids, such as pruritis, which may lead to skin trauma due to scratching, thereby provoking new eruptions in patients with pemphigus vulgaris.[12] In conclusion, regional anaesthesia was chosen in order to avoid general anaesthesia, airway manipulation and possible trauma to the soft tissue of pharynx. Intrathecal clonidine with bupivacaine can be used safely to provide satisfactory anaesthesia and postoperative pain control. Epidural top-ups further helped avoiding systemic opioids and other analgesics and consequently their side effects.

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