Case Report

Anaesthetic Considerations in a Patient with Dermatitis Herpetiformis

Ashwini.H.R¹, Jyothi B², Deepsen Gupta³

¹Senior resident, ²Associate Professor, ³Post Graduate Student;
Department of Anaesthesia, Karnataka Institute of Medical Sciences, Hubli, Karnataka, India.

Corresponding Author: Ashwini.H.R

Received: 27/03//2014 Revised: 17/04/2014 Accepted: 21/04/2014

ABSTRACT

Dermatitis herpetiformis (DH), a blistering skin disease is frequently associated with autoimmune conditions like insulin dependent diabetes mellitus, thyroid abnormalities, neurological dysfunction and intestinal malabsorption. We are presenting a case report on anaesthetic management of 14 year old girl with DH on treatment with dapsone for a minor surgery under subarachnoid block.

Keywords: Dermatitis herpetiformis, dapsone, methaemoglobinemia.

INTRODUCTION

Dermatitis herpetiformis (DH) is an autoimmune skin disorder frequently associated with gastrointestinal disease and characterised by chronic, intensely itchy, papulovesicles distributed symmetrically on extensor surfaces. [1] Patients with dermatitis herpetiformis do not carry an increased operative risk during surgical procedures but association with other conditions like neurological dysfunction, thyroid abnormalities, insulin dependent diabetes mellitus and intestinal malabsorption leading to deficiency syndromes demands attention of anaesthesiologists and proper work up of the case to manage it uneventfully. We report here the anaesthetic management of a patient with dermatitis herpetiformis with cellulitis of lower limb undergoing debridement. To the best of our knowledge there are very few reports of dermatitis herpetiformis in the literature with relevant to anaesthesia.

CASE REPORT

A 14 year old female who was a known case of dermatitis herpetiformis diagnosed at the age of 9 years, was posted for debridement of lower limb cellulitis. Past history revealed recurrent blisters waning off spontaneously. She was treated with tab dapsone 50 mg daily and local application of steroids with which she responded. On examination she looked pale with a bald tongue. Her general examination showed tense blisters, ruptured vesicles and scabs over sacral area, extremities and around neck whereas healed areas were hypopigmented. Systemic examination was within normal limits with adequate mouth opening and normal neck movements while spine examination revealed blisters and scabs over upper back. Preoperative
investigation revealed Hb-8gm/dl, platelets-3,50,000 cells/ dl, normal coagulation profile, random blood sugar- 98 mg/dl, serum creatinine- 1.2 mg/dl, serum urea 38mg/dl with serum electrolytes in the normal range. Her previous histopathological report of skin lesions revealed dermatitis herpetiformis.

Preinduction, she was advised nil by mouth for 8 hours and tab diazepam 10 mg orally. She was wheeled into operation theatre and monitoring included electrocardiography (ECG), pulse oximetry (spo2), non invasive blood pressure (NIBP). Regional anaesthesia was planned with sub arachnid block being the technique of choice. In the operating room, her preoperative oxygen saturation ranged from 89- 91% with other vitals being normal. She was preloaded with ringer lactate solution of 500 ml followed by subarachnoid block using 3 ml of 0.5% heavy bupivacaine under asepsis. Intraoperatively there was no incidence of hypotension or hypoxia. Patient was given 100 % oxygen for some time but, it did not improve the pulse oximetry readings. Duration of surgery was 45 minutes and intraoperative course was uneventful. She was shifted to recovery room. Patient’s arterial blood sample was sent for acid base gas analysis later and found to be normal.

DISCUSSION

Dermatitis herpetiformis, characterised by IgA deposition in the papillary dermis, is a blistering skin condition that exemplifies an extra intestinal manifestation of gluten sensitivity. Although only 10% of DH patients have gastrointestinal symptoms, they are all said to have gluten sensitive enteropathy. The age of onset is usually about 15-40, but DH can also affect children and the elderly. Men and women are equally affected. The incidence of DH varies from 1 in 400 to 1 in 10000 and is associated with the HLA-DQ2 haplotype along with celiac disease and gluten sensitivity. It is most common in patients of northern European ancestry but 22 cases have been reported in China with similar presentation as in Caucasians. [3]

Anaesthetic management of dermatitis herpetiformis varies depending on the clinical manifestation and associated other conditions. Anaesthetic consideration includes, presence of other autoimmune disorders like thyroid (5-11%), pernicious anaemia (1-3%), type I diabetes (1-2%), sjogren’s, SLE, celiac disease of gut and associated malabsorption syndromes. [4] These patients are treated with Dapsone which may cause methemoglobinemia interfering with interpretation of patient’s oxygen status. Preoperative thyroid function tests or other serum immunological studies should be done whenever indicated as anaesthetic implications are based mainly on presence of associated disorders.

Though DH is rare in paediatric population, Per Helsing reported a case of child presenting with ataxia which improved after treatment with dapsone and gluten free diet. [5]

Although association of neurological dysfunction is still debated, Zsolt Barta reported a case of hypokalemic myopathy in a patient of DH with neurological dysfunction which resolved with gluten free diet. [6] The association of celiac disease (CD) and epilepsy has been demonstrated in a number of studies, but the nature of association remains obscure. [7,8] Three groups have shown an increased incidence of CD in patients with idiopathic cerebellar ataxia. The name gluten ataxia was proposed for it by Hadjivassiliou et al. a noteworthy finding that merits further attention is association of common HLA haplotype in them. [9-11]

A case of 15 year old girl was reported by M.P.S. Sahney, who had
primary hypothyroidism, pituitary mass mimicking macroadenoma and DH. She improved with thyroxin, dapsone and steroid therapy thus negating the need for pituitary surgery.\textsuperscript{[1]} Heinlin reported a case of 15 year old girl of DH presenting with palmar and digital petichiae and she was successfully treated with Dapsone.\textsuperscript{[12]}

Pulse oximetry readings in our patient showed 89-91% throughout the surgery in spite of all known and common causes of hypoxia being ruled out. Acid base gas analysis was done in curious to know the fall in spo2 readings but, it was normal. Co oximetry or methemoglobin levels should have been done to rule out dapsone associated methemoglobinemia. She was taking dapsone 50 mg regularly for the past 6 years. Dapsone is a synthetic sulfone antimicrobial used in the treatment of DH however it causes methemoglobinemia, which occurs when hemoglobin iron becomes trapped in the ferric (Fe3+) state. Decreased oxygen carrying capacity leads to severe hypoxia and cyanosis if not treated. Confirmed symptomatic methaemoglobinemia is defined as otherwise unexplained cyanosis or hypoxia (O2 saturation under $\leq 95\%$) together with an elevated methemoglobin level $\geq 3\%$ based on co-oximetry measurement on venous blood. Suspected symptomatic methemoglobinemia included the above symptoms without laboratory-confirmed methemoglobin level. The prevalence and associated risk factors for dapsone-associated methemoglobinemia in pediatric patients has not been well studied. Cytochrome b5 reductase is an enzyme that reduces the toxic metabolite of dapsone (dapsone hydroxylamine) in the liver. Complete absence of cytochrome b5 reductase enzyme activity (CYB5RA) is associated with congenital methemoglobinemia, and its role in dapsone-associated methemoglobinemia is unknown.\textsuperscript{[13]}

**CONCLUSION**

This case indicates that management of a case DH requires an insight into its associated conditions, preoperative medications and its adverse effects as they interfere with the anaesthetic management and interpretation of pulse oximetry respectively.

**REFERENCES**


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