Case Report

Surgical repair of umbilical hernia in a patient with Churg–Strauss syndrome under regional anaesthesia

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Abstract

Churg–Strauss syndrome (CSS), also referred to as eosinophilic granulomatosis with polyangiitis, is characterised by asthma, peripheral and tissue eosinophilia, extravascular granuloma formation and vasculitis of multiple organ systems. Patients with CSS have higher risk for hyper-reactive airway disease. Administering general anaesthesia in such patients may be deleterious as it requires airway manipulation, use of muscle relaxants some of which are associated with release of histamine and use of anti-cholinesterases which can cause bradycardia, increased secretions and bronchial hyper-reactivity. We report a case of a 36-year-old male with known CSS presented with umbilical hernia for mesh repair which was managed with regional anaesthesia.

Keywords: Anti-neutrophil cytoplasmic antibody-associated vasculitis, asthma, Churg–Strauss syndrome, eosinophilic granulomatosis with polyangiitis, hyper-eosinophilic syndromes

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INTRODUCTION

Churg–Strauss syndrome (CSS), also referred to as eosinophilic granulomatosis with polyangiitis (EGPA), is a rare systemic necrotising vasculitis that affects small-to-medium-sized vessels and is associated with severe asthma and blood and tissue eosinophilia, extravascular granuloma formation and vasculitis of multiple organ systems.[1,2] It usually starts at a mean age of 48 years, with a male to female ratio of 1.2:1. EGPA is an anti-neutrophil cytoplasmic antibody-associated vasculitis.[3,4] It can involve any organ of the body such as lungs, skin, cardiovascular system, kidney, peripheral nervous system and gastrointestinal tract. Patients mostly present with severe asthmatic attacks, allergic rhinitis and sinusitis, and the second most common manifestation is mononeuritis multiplex.

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Churg Strauss is a primary systemic vasculitis. CSS was first described in 1951 by Dr Jacob Churg and Dr Lotte Strauss as a syndrome consisting of asthma, eosinophilia and additional vasculitis of multiple organ systems. CSS results from inflammation that occurs in certain types of cells in blood or in tissues. This inflammation causes injury to organ systems – the most commonly involved are the lungs, kidneys, gastrointestinal tract, heart, central nervous system, skin, peripheral nerves, lymph nodes and joints.[3]

Immune complexes are probably involved as the cause of CSS.[5] Patients with CSS display cholinesterase deficiency, hyper-sensitivity of the airway and multiple organ dysfunction.[6] For this reason, anaesthetic management of patients with CSS can be difficult. English literature search

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showed scant reports over anaesthetic management of these cases. We report here one the anaesthetic management of a patient with CSS presented with umbilical hernia scheduled for mesh repair. We preferred regional anaesthesia over general anaesthesia, thereby avoiding airway manipulation, muscle relaxants and the use of anti-cholinesterases.

CASE REPORT

A 36-year-old male patient with body mass index of 35 kg/m^2 presented with umbilical hernia and was scheduled for mesh repair. He had a history of asthma for the past 15 years and was on bronchodilators since then. He had prior hospital admission for polyarthritis and mononeuritis multiplex. During the course of hospital admission, his nerve biopsy showed vasculitis neuropathy, skin biopsy was suggestive of non-specific inflammation and his anti-proteinase 3 of neutrophils was positive. He received 6 cycles of cyclophosphamide therapy which alleviated his symptoms. Later, after 3 years, he was hospitalised for recurrent episodes of bronchial asthma, bilateral pedal oedema, facial puffiness which was increasing in the early morning hours, shortness of breath, cough, wheeze and temporo-frontal throbbing type of headache during evening hours. There was no history of any new onset of rash, sensory symptoms, proximal distal muscle weakness, oro-genital ulcers, malar rash, discoid rash, arthritis, photosensitivity and alopecia. Further investigations showed nephrotic range proteinuria, peripheral eosinophil count >20%, with pulmonary infiltrates in the chest X-ray. Kidney biopsy showed membranous nephropathy. Immunologically, he was positive for anti-nuclear antibody, anti-ribonucleoprotein proteinase 3 of neutrophils and anti-myeloperoxidase antibodies. His nephropathy was treated with cyclosporine therapy. During this course of hospital admission, he was labelled to be a case of CSS.

Pre-operative evaluation was done and was found that he had recurrent attacks of bronchial asthma, so his respiratory status was optimised using bronchodilators and incentive spirometry for over a week. Pulmonary function testing (PFT) showed mild obstructive pattern after the treatment, his chest X-ray improved and electrocardiogram (ECG) and echocardiogram were normal. He was found to have umblical hernia measuring 5 cm × 7 cm which was reducible with wide neck not associated with pain or obstruction. Ultrasonography revealed omentum and small bowel as contents. His routine preoperative laboratory values were within normal limits. He was on tablet azathioprine 50 mg bd, tablet methyl prednisolone 5 mg, tablet losartan 50 mg once-a-day and on salbutamol rotacaps twice-daily.

He was pre-medicated with tablet alprazolam 0.25 mg and salbutamol rotacaps on the night before and morning of surgery; tablet pantoprazole 40 mg and tablet prednisolone 5 mg on the morning of surgery were continued; tablet azathioprine and tablet losartan were discontinued on the day of surgery. In the operating room, standard monitoring including arterial oxygen saturation measured by pulse oximetry (SpO_2), capnography, electrocardiogram (ECG), non-invasive blood pressure and clinical monitoring was done and the plan of anaesthesia was combined spinal and epidural anaesthesia/analgesia. Intravenous cannula was secured, and the patient was administered epidural anaesthesia at the level of thoracic 9–10 intervertebral space with 18-gauze Tuohy needle and sub-arachnoid block at the level of L2–3 intervertebral space using a 25-gauze spinal needle with 3 mL 0.5% bupivacaine heavy along with 25 μg fentanyl and the block achieved was up to T8 level. Intraoperatively, he was supplemented with oxygen at 6 lpm via a simple facemask. The entire procedure went uneventful and was haemodynamically stable. The patient was shifted to the post-operative ward and pain was mitigated with epidural analgesia using 0.125% bupivacaine infusion for 24 h. He was monitored in post-operative period for 24 h and it was uneventful.

DISCUSSION

The American College of Rheumatology (ACR)[7] defined criteria to confirm the diagnosis of CSS. A patient should have at least 4 of the 6 ACR criteria for CSS to be diagnosed. These criteria are asthma, eosinophilia (>10% on the differential white blood cell count), mononeuropathy, transient pulmonary infiltrates on chest X-rays, paranasal sinus abnormalities and a biopsy containing a blood vessel with extravascular eosinophils. Specific symptoms depend on the organs or diseases involved. Patients may have dyspnoea from asthma, chest pain from disease affecting the lungs or heart, rashes on the skin, muscle and/or joint pain, features of sinusitis, abdominal pain or blood in the stools occurring as a result of intestinal tract involvement and paraesthesia as a result of nerve involvement. It is characterised by marked leucocytosis (60,000) and eosinophilia (84%). Eosinophil counts fluctuate widely.

Therefore, there is a need for repeated differential count. Typically, the patient presents with allergic rhinitis, which will usually be complicated by nasal polyposis and sinusitis. Asthma and peripheral blood eosinophilia are essential features associated with pulmonary infiltrates.[8] It resembles several other granulomatous, vasculitis and eosinophilic disorders. CSS has all these three features during various phases of disease. Major long-term problems in the

post-vasculitis phase are due to hypertension and persistent peripheral neuropathy. Allergic upper and lower respiratory tract diseases contribute to morbidity.[7] CSS shows good response to oral prednisone 40–60 mg/day, which can later be tapered down. Other immunosuppressive drugs such as azathioprine, mycophenolate mofetil, cyclophosphamide or methotrexate may be used in addition to prednisone. The prognosis of patients with CSS is not clearly known, but 1-year survival rate of 90% and 5-year survival rate of 62% have been reported after treatment with corticosteroid. A significant reduction in mortality has been reported after treatment.[9] Relapse of asthma and allergic manifestations is independent of vasculitis. Relapse of vasculitis is also seen in 30%–50% of patients.[3]

Pre-operative evaluation should include detailed history regarding severity of disease, medications and precipitating factors. Clinical examination should include detailed physical examination with a focus on respiratory system. Bedside PFTs such as breath holding time and forced expiratory time can be used to assess the pulmonary reserve. Wheeze on auscultation needs special consideration. Pre-operative optimisation using incentive spirometry is essential in improving the surgical outcome. Special attention should be paid towards motor and sensory examination in patients with mononeuritis from medico-legal point of view. Pre-operative investigations should include PFTs to assess patient's respiratory reserve and to predict risk of post-operative pulmonary complications, as well as cardiac evaluation with echocardiography to assess myocardial function. It is necessary to assess the involvement of other systems such as renal system, joints and central nervous system.

Patients on long-term corticosteroids could develop adrenal insufficiency and may require peri-operative supplemental corticosteroids. The patient in this case was on 5 mg prednisolone and no additional dose of steroid was given peri-operatively as per the guidelines. Anxiety can trigger bronchospasm. Pre-medication should aim to relieve anxiety. Over-sedation which can lead to respiratory depression should be avoided. The airway in these patients being hyper-reactive may pose problems during general anaesthesia. Pain, fluid shifts and delayed mobilisation can contribute to an increased risk of post-operative pulmonary complications in these patients. Regional anaesthesia is not totally free of complications. Anxiety during surgery in awake patients can precipitate bronchospasm. Ester local anaesthetics carry a low but appreciable incidence of allergic reactions. The choice of anaesthesia depends on patient's disease status, location of surgery and duration of surgery. We chose regional anaesthesia for this patient taking all measures to decrease anxiety such as pre-medication and communication with the patient. Regional anaesthesia remains the anaesthetic management of choice in asthmatic patients whenever feasible as it avoids tracheal intubation, which itself may induce bronchospasm. In cases where general anaesthesia needs to be given adequate depth of anaesthesia should be achieved before intubation. Other concern is decreased cholinesterase activity or abnormal sensitivity to suxamethonium.[10] Care should be taken while positioning the patient especially if joint involvement is present. Post-operative management should focus on adequate analgesia and prevention of post-operative pulmonary infections. Good post-operative analgesia, incentive spirometry, chest physiotherapy and bronchodilators help in optimising post-operative pulmonary function.

The anaesthetic management should focus on the organs involved during the disease process, optimising them before surgery and careful execution of the anaesthetic plan chosen, thereby avoiding the potential risks involved in patients with CSS. The present case was successfully managed using regional anaesthesia with focus on adequate analgesia and prevention of pulmonary infections.

Declaration of patient consent

We the authors certify that we have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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Korambayil PM, Alappatt K, Ambookan PV. Microtia deformity correction with three-dimensional and two-dimensional templates: A pathway for effective ear reconstruction. J Clin Sci Res 2021;10:252-5.

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B. Vengamma
Hon. Editor-in-Chief