



Case Report

Still's disease and anaesthetic concerns: A case report

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ABSTRACT

Adult-onset Still's disease (AOSD), is a chronic systemic inflammatory¹ disorder rarely encountered in clinical practice, described by Sir George Frederick Still in 1897. AOSD is of unknown aetiology with the incidence estimated to be 0.16 per 100,000 persons with articular and extra-articular or systemic manifestations. AOSD has a more acute course than compared to rheumatoid arthritis in adults, often affecting many parts of the body before settling in the various joints. Its diagnosis is made by exclusion. Securing airway mainly tracheal intubation may be difficult due to involvement of cervical spine, temporomandibular joint and laryngeal involvement (crico-arytenoid arthritis). In addition, intermittent disease flare-ups with laryngeal involvement may cause delayed extubation. This case highlights the anaesthetic concerns involved with the Still's disease.

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1. Case Report

A 33-year-old averagely built female patient presented to emergency room of obstetrics and gynaecology department of a tertiary care centre for pain in abdomen and bleeding per vaginum for last 5-6 hours. A quick sonography of abdomen showed ruptured ectopic pregnancy with mild free fluid in the abdomen. Seeing the condition of patient, an emergency laparotomy was planned. Preoperatively on history, she had amenorrhoea since 8 weeks with pain abdomen of increasing intensity for last few hours and bleeding per vaginum. She also gave history of joint pain along with swollen painful joints with limited movement in both hands since many years. She also had a history of treatment for pott's spine 3-4 years back. While screening her past medical records, we found that she was diagnosed with AOSD at 26 years of age and was on methotrexate, steroid therapy and NSAIDs for one and half years after which patient herself defaulted the treatment and presently not on any medication for AOSD since last 5 years.

General physical examination revealed hyper-pigmented rash over the both malar prominences. Her vitals recorded

as pulse 110/minute, regular with BP of 90/54 mm- Hg and BMI of 24.5 kg/m². Auscultation of heart and lungs was unremarkable. ECG was normal. Airway examination revealed mouth opening of 3.0 cm, Mallampatti class 3 and restricted neck extension and flexion. Haemoglobin-7.2gm%, and Platelet count-1.2 lakh/mm³ were only blood reports available. LFT, K FT and coagulation profile could not be done because of urgency of surgery. Instruction to arrange adequate blood products was given.

2. Anaesthesia Conduct

General anaesthesia was planned taking into consideration the urgency of operation, severe anaemia and history of pott's spine. Well-informed written high risk consent was taken in view of difficult airway and associated AOSD. After shifting the patient in the operation theatre, all standard ASA monitors were attached (ECG, NIBP, pulse oximeter) and baseline parameters noted (HR=110beats/min, BP=90/54mm of Hg, MAP=63 mm of Hg). Two large bore 18G cannulas were secured over both forearms and ringer lactate infusion via fluid warmer was started. Difficult intubation cart including conventional laryngoscopic blades, McGrath video laryngoscope, bougie, ETT of various sizes,

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Table 1:

Fautrel's Diagnostic Criteria	Yamaguchi's Diagnostic Criteria
Major criteria	Major criteria
Spiking fever >39°C	Fever ≥ 39°C intermittent for > 1 week
Arthralgia	Arthralgias > 2 weeks
Transient erythema	Typical rash
Pharyngitis	Leukocytosis >10k (>80% PMN)
Glycosylated ferritin <20%	
PMN ≥ 80%	
Minor criteria	Minor criteria
Maculopapular rash	Sore throat
Leukocytes ≥10x10 ⁹ /l	Significant lymphadenopathy
	Hepatomegaly or splenomegaly
	Abnormal liver function
	Negative ANA and RF
	Exclusion criteria • Infections • Malignancies
	Other rheumatic diseases
Diagnosis requires 4 major criteria or 3 major and 2 minor	Diagnosis requires 5 or more criteria, 2 of which must be major

LMA proseal (#3,4), surgical cricothyroidotomy tray and tracheostomy tubes of different sizes were kept ready. Fiberoptic was not available with us in emergency. ENT surgeon was also called for stand by in OT during induction of general anaesthesia. Inotropic infusions along with compatible blood and blood products were kept ready.

Patient was given ranitidine 50mg and ondansetron 4mg intravenously. After preoxygenation with 100% O₂ for 3 minutes, rapid sequence induction was done with thiopentone 5mg/kg i.v and succinylcholine 1.5mg/kg i.v. Laryngoscopy was done with the McGrath video laryngoscope in view of restricted neck movement but the view was very hazy, so direct laryngoscopy with Macintosh blade #3 was attempted and the Cormack Lehane grade 3b laryngeal view confirmed. Intubating bougie was inserted across the vocal cords and size 7.0 mm ID cuffed oral PVC endotracheal tube was rail roaded over the bougie, thereafter bougie removed and bilateral air entry confirmed. Airway management was completely atraumatic. Patient attached to mechanical ventilator on volume control mode with tidal volume of 8 ml/kg. Anaesthesia was maintained with O₂:N₂ O mixture 50:50 with Isoflurane up to 1 MAC with boluses of Atracurium. Dexamethasone IV 4 mg was given for suspected airway oedema and Fentanyl 2µg/kg body weight IV was given to the patient for analgesia. Intraoperative blood loss was 900 ml, which was replaced with 2 units of packed cell volume and 1.0 litre crystalloid. Urine output intraoperatively was 150 ml. Paracetamol infusion 1 gm IV given for multimodal analgesia 30 minutes before the expected extubation and bilateral TAP block with 20ml 0.25% bupivacaine was given on each side at the end of surgery. Intraoperative course remained uneventful. At the end of surgery, pulse rate was 92/min and BP was 94/56 (MAP 64 mm Hg). ETT cuff deflation demonstrated

peritubal air leak signifying that no laryngeal oedema was present. Patient was extubated when fully awake, breathing spontaneously and shifted to post-anaesthesia care unit. Postoperative course remained stable.

3. Discussion

AOSD is a rare chronic systemic inflammatory disorder, affecting mainly patients between 16 and 35 years of age. Etiopathogenesis remained unknown; however, a genetic component with HLA antigens involvement has been proposed.

Adult Onset Still's Disease Diagnostic Criteria^{2,3} helps in reaching a diagnosis for further management.

Abnormalities predisposing to a difficult airway include TMJ ankylosis, cervical spine or atlantoaxial joint involvement, and cricoarytenoid arthritis. Acute cricoarytenoiditis flares resulting in marked arytenoids swelling, narrowed glottic aperture, and upper airway obstruction⁴ causing symptoms of sore throat, hoarseness, odynophagia or occasional stridor, have been well described. The disease's clinical course has three patterns: (1) self-limiting with remission within a year, (2) intermittent with recurrent disease flare-ups and complete remission, and (3) chronic articular pattern with persistent active disease. AOSD is a diagnosis of exclusion with nonspecific lab results such as significant Leucocytosis, markedly elevated ESR & CRP, with the absence of positive ANA and rheumatoid factor. Treatment with NSAIDs, corticosteroids, and/or methotrexate is the mainstay therapy.⁵ Disease-modifying anti-rheumatic drugs (DMARDs) have been used with mixed results. Lately, use of TNF-inhibitor and IL receptor antagonist has proved to be effective. Symptomatic patients with laryngeal edema should initially be treated with racemic epinephrine nebulizations, humidified O₂,

and systemic corticosteroids. If regional anaesthesia is not a reasonable option, then fiberoptic intubation with a small ETT is recommended provided there is no urgency of surgery. Surgical intervention with tracheostomy, arytenoidectomy or, arytenoidopexy may be necessary if the problem persists despite medical treatment.

4. Conclusion

Airway management in the presence of an arthritic triad involving cervical spine, temporomandibular joints, and larynx may challenge the expertise of even the most experienced anaesthesiologist. Varying degrees of laryngeal obstruction due to cricoarytenoid arthritis is a well-known but uncommon complication of rheumatologic disorders, and anaesthesiologists should be fully aware of this problem. A thorough preoperative airway assessment, preparedness for potential problems and alternative plans are essential for the successful and safe management of these patients. In our case, anaesthesia team was well prepared for a difficult airway; but patient was intubated orally without any significant problem and had an uneventful anaesthetic course. Postoperatively, laryngeal reflexes must return before extubation, and oversedation must be avoided in order that the poor airway found in these patients is at best preserved.

5. Conflict of interest

None.

6. Source of funding

None.

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