Challenge of Difficult Airway and Anesthetic Management in a Patient with Still's Disease

Ranita R. Donald, M.D., Emi Taylor, AA-C, Thomas Gallen, M.D.
Department of Anesthesiology & Perioperative Medicine, Medical College of Georgia, Augusta , Georgia

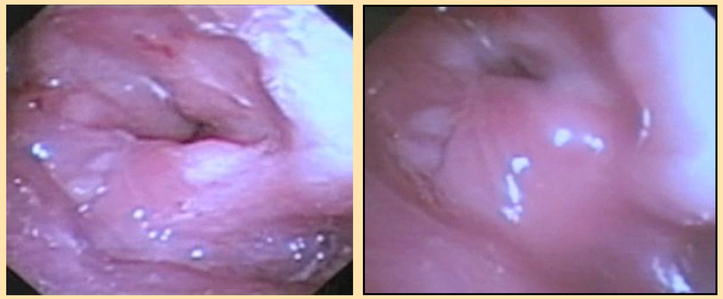
Introduction

Adult-onset Still's disease (AOSD) is a rare systemic inflammatory disorder of unknown etiology with articular and extra-articular (systemic) manifestations. The disorder owes its name to Sir George Frederick Still, who in 1897 described 22 children with symptoms consistent with what is currently known as systemic onset juvenile idiopathic arthritis. AOSD was established almost a century later in 1971, when Eric Bywaters encountered and described adult patients presenting with pediatric Still's disease symptoms. Compared to rheumatoid arthritis in adults, AOSD runs a much more acute course, quite often affecting many parts of the body before settling in the various joints. Diagnosis of AOSD is difficult to establish due to the nonspecific clinical and laboratory findings. Tracheal intubation may become difficult due to impairment of cervical spine, temporomandibular joint and laryngeal involvement (crico-arytenoid arthritis). Patients with chronic articular disease have more disability and worse prognosis than patients with only systemic symptoms.

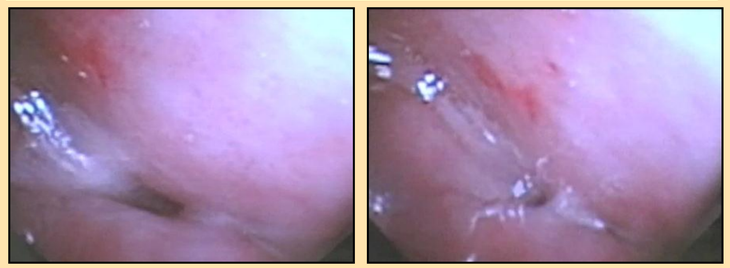
Case Report

A 50-year-old African American female with a 25-year history of polyarthritis and deformities was initially diagnosed with rheumatoid arthritis but was later diagnosed by her rheumatologist to have severe juvenile rheumatoid arthritis (Still's disease). She had a history of hypertension; anemia; seizures (remote); GERD; osteoporosis with bilateral hip and knee replacements, deformed swollen painful joints, and limited movement; and contracted extremities and fingers. She was scheduled for cervical LEEP for high-grade dysplasia. Airway evaluation revealed a difficult airway with limited mouth opening, very limited neck extension, and minimal neck flexion due to cervical spine arthritis. Spinal/epidural option was excluded because of patient's reluctance and a healing herpes zoster rash in her lower back. Patient's airway was fully topicalized for fiberoptic intubation with local anesthetic nebulized inhalation, spray to oropharynx, and vasoconstrictor spray twice to each nostril. After patient was sedated, a trial of lithotomy position was done in the operating room while patient was still awake to assess her joint flexibilities. Later, patient was given small incremental boluses of propofol for anxiety so that she lost consciousness but maintained spontaneous ventilation. Regular laryngoscope blade and Glide-scope blade wouldn't go far enough in mouth due to limited mouth opening and rigid neck. Nasal flexible fiberoptic scope demonstrated swollen arytenoids and aryepiglottic folds obscuring visualization of vocal cords. Patient was intubated after the difficult search for glottic aperture, using 6.5 mm endotracheal tube (ETT), following the path of escaping air bubbles and stream through the narrowed glottic opening surrounded by swollen laryngeal tissues. Airway management was completely atraumatic. Intraoperative course remained uneventful. Patient was given intravenous steroid for laryngeal edema. Patient was extubated when fully awake, breathing spontaneously. ETT cuff deflation demonstrated air leak. Patient was closely observed in the postanesthesia care unit and remained stable. Patient returned two months later for robotic hysterectomy. Anesthesia team was well prepared for a difficult airway; but patient was intubated orally without problems and had an uneventful anesthetic course. This case highlights the literature-described intermittent disease flare-ups with laryngeal involvement.

Figures



Swollen arytenoids and aryepiglottic folds



Escaping air stream through narrowed glottis

Adult Onset Still's Disease Diagnostic Criteria

Yamaguchi's Diagnostic Criteria

Major criteria
- Fever ≥ 39° intermittent for > 1 week
- Arthralgias > 2 weeks
- Typical rash
- Leukocytosis >10k (>80% PMN)

Minor criteria
- Sore throat
- Significant lymphadenopathy
- Hepatomegaly or spenomegaly
- Abnormal liver function
- Negative ANA and RF

Exclusion criteria
- Infections
- Malignancies
- Other rheumatic diseases

Diagnosis requires 5 or more criteria, 2 of which must be major

Fautrel's Diagnostic Criteria

Major criteria
- Spiking fever >39°
- Arthralgia
- Transient erythema
- Pharyngitis
- Glycosylated ferritin <20%
- PMN ≥ 80%

Minor criteria
- Maculopapular rash
- Leukocytes ≥10x10^9/l

Diagnosis requires 4 major criteria or 3 major and 2 minor

Discussion

AOSD is a rare, not readily diagnosed, variable chronic systemic inflammatory disorder. It usually affects people between 16 and 35 years of age. In a French retrospective study, the incidence was estimated to be 0.16 per 100,000 persons, while a Japanese epidemiological analysis reported an incidence of 0.22 and 0.34 per 100,000 amongst men and women. Etiopathogenesis is still unknown, however some progress has been made. Proposed etiologies include a genetic component, linking disease with number of HLA antigens, viral and microbial infections, and more recently alterations in cytokine production such as increased production of IL-2, -6, -8 & -18, interferon γ and tumor necrosis factor α promoting cell-mediated immunity.

Disease is characterized by spiking fevers exceeding 39°C, an evanescent salmon pink rash, arthritis, sore throat, myalgia, lymphadenopathy, hepatomegaly, splenomegaly, pleuritis, pericarditis, neutrophil leucocytosis, and high serum ferritin level. Abnormalities predisposing to a difficult airway include TMJ ankylosis, cervical spine or atlantoaxial joint involvement, and cricoarytenoid arthritis. Acute cricoarytenoiditis flares resulting in marked arytenoid swelling, narrowed glottic aperture, and upper airway obstruction causing symptoms of sore throat, hoarseness, odynophagia or occasional stridor, have been well described.

AOSD is a diagnosis of exclusion with nonspecific lab results such as significant leukocytosis, markedly elevated ESR & CRP, with the absence of positive ANA and rheumatoid factor. Various AOSD diagnostic criteria have been proposed as shown in the chart. 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.

Treatment with NSAIDs, corticosteroids, and/or methotrexate is the mainstay therapy. DMARDs have been used with mixed results. Lately, the use of TNF-inhibitor and IL receptor antagonist have proved to be effective. Symptomatic patients with laryngeal edema should initially be treated with racemic epinephrine nebulizations, humidified O_2, and systemic corticosteroids. If regional anesthesia is not a reasonable option, then fiberoptic intubation with a small ETT is recommended. Surgical intervention with tracheostomy, arytenoidectomy or, arytenoidopexy may be necessary if the problem persists despite medical treatment.

Summary

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 anesthesiologist. Varying degrees of laryngeal obstruction due to cricoarytenoid arthritis is a well known but uncommon complication of rheumatologic disorders, and anesthesiologists 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.

References

1. Bagnari V, et al. *Rheumatol Int* 2010; 30:855-862
2. Efthimiou P, Paik PK, Bielory L. *Ann Rheum Dis* 2006;65:564-572
3. Kolman J, et al. *Can J Anesth* 2002;49;729-732
4. Bandi V, Mannur U, Braman S. *Critical Care Clinics* 2002;18:749-764
5. Phelps JA. *Anesthesiology* 1966; 27: 518
6. Yamaguchi M, et al. *J Rheumatol* 1992;19:424