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CLINICAL VIGNETTE

Anesthetic Considerations for a Patient with Felty Syndrome

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Introduction

Felty syndrome is a rare complication of rheumatoid arthritis (RA) that was first described in 1924.[1] The syndrome is described as a triad: seropositive RA, neutropenia, and splenomegaly. It primarily presents in the context of long-standing RA with severe joint involvement, including, but not limited to, cervical spine instability. While the complete triad is not necessary for the diagnosis of Felty syndrome, neutropenia is a hallmark finding and must be present.[2] The prevalence is estimated to be 1-3% in patients with RA although this seems to be decreasing due to advancements in RA treatment such as methotrexate and biologics.[3,4]

The exact pathophysiology of Felty syndrome is unknown, however, the combination of a systemic pro-inflammatory state and splenic sequestration of neutrophils often puts patients at high risk for respiratory and skin infections. Splenectomy can result in the remission of neutropenia; however, it is often only indicated in the setting of recurrent infections and severe, persistent neutropenia.[5] Given the high risk for infection, the likely long-term immunosuppressant use, and, possibly the most concerning for anesthesiologists, cervical spine instability, patients with Felty syndrome present unique anesthetic considerations. We present a case of a patient with Felty syndrome who underwent monitored anesthesia care for hysteroscopy.

Case Presentation

A 61-year-old female with Felty syndrome presented for hysteroscopy for evaluation of endometrial hyperplasia. The patient had a medical history of long-standing, seropositive RA diagnosed approximately 11 years ago. She was recently hospitalized for complications due to Felty syndrome at which time she was started on Prednisone 10 mg daily. Prior to her hysteroscopy, the patient was optimized from a rheumatology standpoint.

Her other medical history included chronic obstructive pulmonary, systemic lupus erythematosus, remote COVID hospitalization, former tobacco use, and obesity. Past surgical history included C-section and appendectomy. The patient was on the following prescription medications: etancercept, methotrexate, prednisone, fluconazole, gabapentin, folic acid, omeprazole, and ibuprofen. Her pre-operative electrocardiogram and chest x-ray were within normal limits and her CT abdomen scan was notable for borderline, enlarged spleen. More importantly, the patient reported having pain with neck movement and her cervical spine flexion and extension films showed the following: generalized osteoporosis with reversed normal cervical spine lordosis, narrowing of intervertebral discs from C4-C7, spondylolisthesis of C2 on C3, C3 on C4, C4 on C5, and C5 on C6; there is spondylolisthesis of C3 on C4 and C4 on C5 on flexion; and spondylolisthesis of C3 on C4 and C5 on C6 on extension of the neck.

On the day of the procedure, the patient's vital signs were within normal limits and her BMI was 31.5. Her pre-procedure airway examination was notable for a Mallampati classification of 3, decreased submental space at 2 fingerbreadths, and poor dentition. Otherwise, the patient had adequate mouth opening and full neck range of motion. Cardiac and respiratory systems were within normal limits.

She was premedicated with midazolam as an anxiolytic after all consents were obtained and verified. Upon arrival to the operating room, the patient was pre-oxygenated and standard ASA monitors were placed. Extra care was taken to ensure the patient's cervical spine stayed in the neutral position throughout the case. Glidescope was prepared for the patient in case of an airway emergency as it provides visualization of the airway while maintaining cervical spinal neutrality. Stress dose hydrocortisone 100 mg was administered intravenously. Her anesthetic included lidocaine, fentanyl, and propofol and she was maintained on a propofol infusion that was titrated to desired effect of spontaneous breathing.

Intraoperatively, the patient was placed in the dorsal lithotomy position with cervical spine neutrality and all her extremities were kept less than 90 degrees. She was kept warm and comfortable for the uneventful 2-hour hysteroscopy under monitored anesthesia care. At the end of the case, she was awake, responsive, following commands, moving all extremities, and was transported to the post-anesthesia care unit for further monitoring and recovery. The patient was later discharged home on the same day.

Discussion

As described earlier, the Felty syndrome triad of RA, neutropenia, and splenomegaly can present unique anesthetic challenges. Because Felty syndrome is often a consequence of long-standing RA, these patients should be managed similarly.

One of the most important considerations for anesthesiologists is atlantoaxial instability and subluxation as they can be long-term consequences of Felty syndrome. Patients should receive a lateral flexion-extension radiograph or an MRI if they show symptoms of spinal cord involvement. Great care should also be taken with the airway evaluation as to not exacerbate the condition. If a difficult airway is anticipated, an algorithmic approach should be employed to assess whether fiberoptic or video-laryngoscope assisted intubation is indicated.

Furthermore, balancing the risk of infection versus glucocorticoid replacement therapy to prevent adrenal insufficiency is also important. Studies suggest that there is a dose-dependent increase of post-surgical infection in RA patients when receiving peri-operative glucocorticoids.[6] Other potential adverse effects of glucocorticoid use include hypertension, hyperglycemia, and fluid retention. On the other hand, adrenal crisis can also be life threatening.

While there are no clear guidelines, stress dose steroids should only be given based on the patient's disease progression and history of glucocorticoid use. Patients with only minor disease progression and taking therapeutic steroid doses do not need perioperative glucocorticoid replacement. Generally, this includes patients taking less than 10 mg of prednisone or any dose of glucocorticoids for less than 3 weeks.[7-9] These patients often do not have significant hypothalamic-pituitary-adrenal (HPA) axis suppression. However, those with severe disease often have some element of HPA axis suppression and should be considered for intraoperative stress dose steroid supplementation and avoidance of certain medications such as etomidate, as it can further suppress the HPA axis, especially if given repeatedly.[10] Our patient received a stress dose of 100 mg hydrocortisone IV given the patient's 11-year history of having seropositive RA, diagnosis of Felty syndrome, disease progression, and long-term steroid use.

Additional anesthesia pre-operative considerations for those with Felty syndrome include proper cardiac risk stratification as the incidence of coronary artery disease is increased in patients with RA. Patients should receive a pre-operative electrocardiogram and undergo atherosclerotic cardiovascular disease (ASCVD) risk scoring. Lower metabolic equivalents (METs) is expected in patients with long-standing RA as joint involvement can limit everyday mobility. These parameters should be used in the context of patient's overall history to determine the safety of surgery.

For more complicated surgeries where significant blood loss is expected, blood transfusions may be indicated as patients with Felty syndrome present with baseline hypochromic anemia due to chronic disease.

Conclusion

Felty syndrome is often associated with long-term progression of RA which itself is a complex, multi-system disease. The presence of atlantoaxial instability and subluxation is important for the anesthesia care as it could significantly change airway management and the type of anesthesia being provided.

Not all patients with RA will require stress dose steroids. However, as Felty syndrome is often a complication of long-standing disease, proper rheumatological evaluation should inform the provider whether HPA axis suppression is present and if stress dose steroids would be indicated.

Within the Felty symptom triad, neutropenia is an important anesthetic consideration. Combined with the likely use of immunosuppressants, patients are at increased risk of serious infections. RA patients already have a higher risk of prosthetic joint infection which theoretically can be exacerbated by perioperative glucocorticoid use.[11] Post-operative monitoring for infection is important.

There is no definitive, curative treatment for Felty syndrome. If neutropenia is severe and persistent (ANC <500/mm^3) splenectomy can be an option; although, due to the advent of disease-modifying anti-rheumatic drugs (DMARDs), this is rarely performed.[12]

Our patient had an uneventful hysteroscopy under monitored anesthesia care.

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