Sarcoidosis

Sarcoidosis is a multisystem granulomatous disorder of unknown etiology that typically affects young adults and is characterized pathologically by the presence of noncaseating granulomas in involved organs. Sarcoidosis can involve all organ systems to a varying extent and degree but has a predilection for intrathoracic lymph nodes and the lungs.

ANESTHETIC CONSIDERATIONS:

- Potential difficult airway: (laryngeal sarcoid, tracheal stenosis)
- Pulmonary sarcoid
  - Restrictive lung disease, pulmonary fibrosis
  - Pulmonary HTN, RV dysfunction
- Extrapulmonary sarcoid
  - Cardiac: CHF (cardiomyopathy), arrhythmia / conduction defects, cor pulmonale
  - Neuropathy: cranial & peripheral
  - Nephropathy: multiple etiologies, renal insufficiency common, ESRD rare
  - Metabolic: hypercalcemia (20%)
  - Arthropathy: polyarticular arthritis
- Treatment
  - Steroids - consider stress dose
- Immunosuppressants - MTX, cyclophosphamide, cyclosporine

ANESTHETIC GOALS:

1. Assess and optimize end organ dysfunction
2. Consider perioperative stress dose steroids

HISTORY

- Identify extent of disease and end organ involvement
- As many as two thirds of patients have no symptoms at the time of presentation
- Assessment of end-organ dysfunction (systemic manifestations vary on the basis of sex, age at presentation, and ethnicity)
  - Functional capacity (cardiopulmonary reserve); however, systemic inflammation may contribute to muscle weakness and exercise intolerance
  - Respiratory disease: restrictive lung disease secondary to scaring from noncaseating granulomas, dyspnea on exertion, cough, O_2 requirements
  - Cardiovascular disease: CHF (restrictive or dilated cardiomyopathy), arrhythmias (SVT, VT, heart block), valvular dysfunction (MR)
    - Granulomatous involvement of the ventricular septum and conduction system can lead to a variety of arrhythmias, including complete heart block and sudden death; such involvement may be heralded by palpitations, syncope, dizziness, or chest pain
    - In addition, chronic pulmonary hypertension and cor pulmonale can result from severe scarring of the pulmonary parenchyma and vascular obliteration; death from sarcoidosis usually results from right ventricular failure
  - Nephropathy (multiple etiologies)
  - Neuropathy
    - Bulbar dysfunction
    - Peripheral nerve palsies
  - CNS: space occupying lesions, seizures, psychiatric examination
  - Generalized symptoms of fatigue, malaise, fever, and weight loss

PHYSICAL

- HEENT:
  - Airway assessment (laryngeal sarcoidosis, epiglottis, and arytenoid involvement)
    - Involvement of nares, polyps w/ distorted anatomy
    - Nasal stuffiness, wheezing, hoarseness, stridor
    - Assess patency of airway (hoarseness, stridor etc)
    - Tracheal stenosis uncommon but may be severe
  - Lymphadenopathy
  - Sarcoidosis can cause diffuse goiter or, rarely, a solitary thyroid nodule
- CVS:
  - Symptoms secondary to arrhythmias, heart failure (RV heave with cor pulmonale)
- RESP:
  - Airway obstruction with endobronchial disease -wheeze
  - Pulmonary fibrosis (dry rales, crepitations)
  - Pleural involvement is unusual but can result in lymphocytic exudative effusion, chylothorax, hemothorax, and pneumothorax
- GI:
  - Can involve any portion of the GI tract from the esophagus to the rectum
  - Hepatomegaly
  - Splenomegaly - Hypersplenism can lead to anemia, leukopenia, and thrombocytopenia
  - Pancreatitis occurs in some cases
- Renal:
  - Symptoms of hypercalcemia and kidney stones
  - Abnormalities related to calcium metabolism are the most common renal and electrolyte abnormalities due to extrarenal production of calcitriol by activated macrophages
  - If untreated, renal calcium deposition can lead to chronic renal failure and end-stage renal disease
  - Other renal complications of sarcoidosis include membranous nephropathy, a proliferative or crescentic glomerulonephritis, focal segmental glomerulosclerosis, polyuria (due to nephrogenic and/or central diabetes insipidus), hypertension, and a variety of tubular defects
- CNS:

- Granulomatous basal meningitis with infiltration or compression of adjacent structures is responsible for most of the CNS manifestations, including:
    - Hypothalamic hypopituitarism
    - Central diabetes insipidus (focal nerve deficits)
    - Hydrocephalus
    - Lymphocytic meningitis
    - Cranial nerve palsies, particularly facial palsy
  - The most common form of neurologic involvement in sarcoidosis is unilateral facial nerve palsy
- MSK:
  - Acute polyarthritis (especially the ankle joints), usually in association with erythema nodosum and occasionally with acute uveitis
  - Chronic arthritis with periosteal bone resorption
  - Diffuse granulomatous myositis -uncommon complication

INVESTIGATIONS

- Degree of investigation determined by complexity of surgery
- **Labs**
  - CBC (anemia, leukopenia, thrombocytopenia), Lytes (hypercalcemia, other electrolyte abnormalities), BUN, Cr, iCa (renal insufficiency), ALP (elevated with diffuse granulomatous hepatic involvement)
  - ABG (may reveal hypoxia and hypocapnia)
  - PFT (may show restrictive pattern with a reduced DLCO; endobronchial involvement may show obstructive pattern)
- **Imaging**
  - CXR (perihilar adenopathy, reticular opacities (upper>lower lung zones), multiple bilateral lung nodules, pulmonary fibrosis)
  - CT (more detailed evaluation of interstitial lung disease – fibrosis, ground glass and traction bronchiectasis, tracheal stenosis)
  - PET scan to differentiate from malignancy
  - ECG (evidence of conduction defects / arrhythmias)
  - ECHO (if history of sarcoid heart or history or physical suggestive of CHF or cor pulmonale)
  - Mediastinoscopy may be necessary to provide lymph node tissue for the diagnosis of sarcoidosis

OPTIMIZATION

- Identify, assess and optimize organ dysfunction as indicated by severity of disease and complexity of surgery / anesthetic
- Stress dose steroids if indicated - Corticosteroids are administered to suppress the manifestation of sarcoidosis and to treat the hypercalcemia

ANESTHETIC OPTIONS

- Local, regional and GA all acceptable
- Decision should be based on patient / surgery / co-morbidities

ANESTHETIC SETUP

- **Drugs**
  - Standard emergency drugs
- **Equipment**
  - CAS monitors
  - Arterial line (if CHF)

MANAGEMENT OF ANESTHESIA

- **Induction**
  - Airway
    - Distortion or obstruction secondary to laryngeal/epiglottis/arytenoid granulomas may interfere with passage of a normal-sized ETT
    - Hypoxia secondary to lung disease
    - See management of anesthesia and considerations for restrictive lung disease, pulmonary HTN
- **Maintenance**
  - Dependent on procedure & underlying condition
- **Emergence**
  - Awake & reversed as always

DISPOSITION & MONITORING

- Depends on extend of disease and end-organ (especially pulmonary) involvement

COMPLICATIONS

- Airway problems secondary to distorted anatomy
- Pulmonary problems secondary to lung involvement

PATHOPHYSIOLOGY

- Sarcoidosis is a multisystem granulomatous disorder of unknown etiology that affects individuals worldwide and is characterized pathologically by the presence of noncaseating granulomas in involved organs
- Despite an extensive research effort, the exact etiology and pathogenesis of sarcoidosis remain unknown
- The estimated prevalence of sarcoidosis is 10 to 20 per 100,000 population
- Sarcoidosis is more common among blacks than whites
- Sarcoidosis typically affects young adults
- In approximately one-half of cases, it is detected in asymptomatic individuals due to incidental radiographic abnormalities (e.g. bilateral hilar adenopathy, reticular opacities)
- Common presenting symptoms include cough, dyspnea, chest pain, eye lesions, and / or skin lesions
- The stage of pulmonary sarcoidosis is based upon the chest radiograph

    - Stage I is defined by the presence of bilateral hilar adenopathy
    - Stage II consists of bilateral hilar adenopathy and reticular opacities
    - Stage III consists of reticular opacities with shrinking hilar nodes
    - Stage IV is characterized by extensive parenchymal damage including reticular opacities, volume loss, conglomerated masses, traction bronchiectasis, calcification, cavitation, and / or cyst formation
- Sarcoidosis can involve all organ systems
    - The most prominent sites of extrapulmonary disease include the skin, eyes, reticuloendothelial system, musculoskeletal system, exocrine glands, heart, kidney, and central nervous system
- Patients with sarcoidosis commonly have an elevated angiotensin converting enzyme (ACE) level and hypercalciuria
    - 20% of patients have hypercalcemia
    - Hypercalcemia results from unregulated production of 1,25-diOH-vit D by granuloma macrophages
- Pulmonary function tests characteristically demonstrate a restrictive defect with impaired gas exchange
- A definitive diagnostic test for sarcoidosis does not exist
    - Diagnosis requires compatible clinical and radiographic manifestations, exclusion of other diseases that may present similarly, and histopathologic detection of noncaseating granulomas

CONSIDERATIONS IN PREGNANCY:

- Systemic sarcoidosis, in the absence of significant cardiopulmonary compromise, does not affect fertility and does not increase the incidence of fetal or obstetrical complications
- It will often improve during pregnancy, possibly due to increases of maternal free cortisol

REFERENCES

- Coexisting Chapter 9
- Barash Chapter 11
- UpToDate