Rheumatologic Emergencies

Dr. Rezaeiyazdi

INTRODUCTION

- Rheumatologic emergencies are relatively uncommon; however, some consultations require quick action to prevent serious clinical sequelae.
- Important immediately on the patient's arrival in the office or receipt of inpatient consultation.

INFECTIOUS ARTHRITIS

- The differential diagnosis of acute-onset arthritis always includes infectious arthritis, especially if accompanied by a fever and constitutional symptoms.
- Early synovial fluid analysis.
- Empirically start IV antibiotic treatment if fluid is inflammatory.

- Bedside arthrocentesis can be performed for accessible joints such as the knee, wrist, elbow, and ankle. Otherwise, order radiologically assisted arthrocentesis.
- Joints with orthopedic hard-ware warrant urgent consultation to the orthopedic surgery service.
- The consequences of improperly treated infectious arthritis include rapid joint destruction and permanent disability.

GIANT CELL (TEMPORAL) ARTERITIS

The risk of sudden and permanent vision loss, manage GCA emergently.

The diagnosis is based on presumptive clinical evidence of jaw claudication, headache, scalp tenderness, and sometimes acute, usually unilateral vision loss.

The diagnosis often accompanies polymyalgia rheumatica, which includes symptoms of fever, morning stiffness, and arthralgias in shoulder, hip. neck, and torso.

Lab studies usually show a high FSR (>50) and anemia. Definitive diagnosis involves temporal artery biopsy of an affected segment.

When the diagnosis of GCA is suspected, initiate immediate treatment with oral prednisone, 60 mg/day.

Obtain lab tests followed by arrangement for a temporal artery biopsy within 1 wk of initiating steroids.

Symptoms often rapidly resolve with steroid initiation.

Once symptoms have resolved and ESR returns to normal, taper steroids.

SCLERODERMA RENAL CRISIS

- Renal crisis is a severe, sometimes life-threatening complication that occurs in approximately 10-15% of patients with scleroderma.
- A scleroderma renal crisis is characterized by rapidly progressive azotemia, malignant HTN (although approximately 10% of patients are normotensive), microangiopathic hemolytic anemia, and thrombocytopenia. Risk factors include diffuse, rapidly progressive skin disease, cool ambient temperatures, races with higher prevalence of essential HTN (i.e., African Americans), and use of corticosteroids or cyclosporine.

- The pathogenesis reflects a severe vasculopathy leading to ischemic activation of the renin-angiotensin system.
- Treatment involves BP control with the use of short-acting AC inhihitors. in particular captopril (initiate at 25 mg bid and titrate to maximum of 50 mg PO tid). For patients intolerant to AC inhihitors, angiotensin receptor blockers may be effective.

- Avoid IV antihypertensives, such as nitroprusside and labetalol. and all nephrotoxins.
- Calcium channel blockers can he added to the regimen of AC inhihitor if necessary for BP control
- Dialysis may be necessary for those patients with end-stage renal failure.

CERVICAL SPINE ABNORMALITIES

- Cervical spine involvement is a common manifestation of RA or severe, long-standing OA.
- Neurologic deficits can occur, even in the absence of pain should he addressed urgently.
- C1-C2 instability may result from tenosynovitis of the transverse ligament of C1, erosion of the odontoid process, ligament laxity, ligament rupture, or apophyseal joint erosion.
- The lesions most likely to lead to myelopathy (disease of the spinal cord) are atlantoaxial anterior subluxation or down-ward/upward subluxation of the C1-C2 facet joints.

- Cranial settling (also known as *basilar invigilation)* in which the odontoid process pushes up into the foramen magnum may also occur.
- Radiographic changes of the C1-C2 joints are common but do not always correlate with neurologic deficits.
- Symptoms of myelopathy are usually slowly progressive but may have rapid onset if associated with cord compression.
- Stabilization procedures.
- Additionally, patients with cervical spine arthritis are at increased risk of traumatic injury during intubation and should be appropriately managed perioperatively.

TRANSVERSE MYELITIS

- Transverse myelitis develops as an acute or subacute inflammatory disorder of the spinal cord. The majority of cases occur following an infection or immunization. Additionally, transverse myelitis can be associated with MS. SLE and other collagen vascular diseases, SS. Behcet's disease, antiphospholipid syndrome, and sarcoidosis.

Symptoms of transverse myelitis may include focal neck and back pain, paresthesias, weakness, sensory loss, urinary retention or incontinence, fecal incontinence, and fever.

Symptoms typically start in the lower extremities and ascend. The disease usually progresses over hours to days and varies in severity from mild neurologic involvement to functional transection of the spinal cord.

- Diagnostic workup includes urgent spinal MRI, brain MRI (to evaluate for MS), and CSF analysis. MRI of the spinal cord reveals variable spinal edema and signal enhancement. Obtaining normal CSF is useful to rule out acute infections. Alternatively, pleocytosis, elevated protein, and decreased glucose may be present.
- Treatment of transverse myelitis has not been well researched. However, high-dose IV corticosteroids are usually initiated within 24 his of diagnosis.

CAUDA EQUINA SYNDROME

Cauda equina syndrome is a rare complication of the seronegative spondyloarthropathies (related to arachnoiditis), in particular AS,

In patients with AS, symptoms may be slowly progressive.

Symptoms include severe low back pain, rectal pain, and pain in both legs.

Additionally, with progressive disease patients can develop saddle anesthesia with loss of bladder and bowel control, poor anal sphincter tone, and impotence.

Patients may also develop variable lower extremity areflexia and asymmetric leg weakness or loss of sensation.

Cauda equina syndrome should be distinguished from sciatica or plexopathy, which do not involve symptoms of incontinence or impotence.

MRI can help confirm the diagnosis, and urgent neurosurgical consultation is required to prevent irreversible neurologic changes.

Steroids and localized radiation treatment may be beneficial with lesions caused by malignancies.

PULMONARY HEMORRHAGE

Pulmonary hemorrhage can be a complication of several rheumatologic and non rheumatologic diseases.

Common presenting signs and symptoms include progressive dyspnea with hypoxemia, hemoptysis, radiographic appearance of alveolar or interstitial infiltrates, anemia or a drop in hemoglobin level of 1.5-4 g/dl.

Correction of hypoxemia and appropriate control of airway (possibly requiring intubation and mechanical ventilation) and correction of coagulopathies should be addressed immediately.

Obtain early consult with a pulmonologist to assess the need for urgent bronchoscopy to help refine the diagnosis.

Demonstration of active bleeding or hemosiderin-laden macrophages in bronchoalveolar lavage or sputum helps confirm the diagnosis of pulmonary hemorrhage.

The differential diagnosis of pulmonary hemorrhage includes pulmonary-renal syndromes, including Goodpasture's syndrome or WG, SLE, extraarticular manifestations of RA.

Behcet s disease, and other systemic vasculitides such as HSP, CSS, MPA and cryoglobulinemia.

Other possible etiologies include uremia, congestive heart failure, infection, pulmonary infarction, pulmonary HTN, and coagulopathy.

- Lab evaluation should include routine chemistries, liver function tests, CBC, and coagulation studies as well as antinuclear antibodies, antiglomerular basement membrane antibody. ANCA, and complement levels (C3, C4, and CH50).
- Lung (or other involved tissue) biopsy may be required for a definitive diagnosis.
- In addition to supportive treatment, target specific therapies at the underlying disorder.
- Pharmacologic therapy may involve a combination of corticosteroids (usually in high IV doses), cytotoxic agents, and sometimes plasmapheresis.

INTESTINAL INFARCTION

Intestinal infarction is a rare complication associated with SLE and polyarteritis nodosa. The disease is manifested by diffuse vasculitis of the mesenteric blood vessels

Patients typically present with symptoms of an acute abdomen, which may be masked, however, by corticosteroids or may occur late in the clinical presentation

Emergent surgical exploration and resection is important, but overall prognosis is poor.

KEY POINTS TO REMEMBER

- Acute onset of mono- or oligoarthritis requires immediate workup for septic arthritis, including urgent diagnostic arthrocentesis.
- Clinical suspicion of GCA should prompt immediate treatment with high-dose corticosteroids and arrangement for temporal artery biopsy within 7 days.
- Toxic patients should be sent immediately to the nearest ER.

SUGGESTED READING

- Rraunwald E, Fauci A, Isselbacher K.J, et al.. eds. *Harrison's principles of internal*

- *medicine,* 15th ed. New York: McGraw-Hill, 2001. Halla.I. Rheumatologic emergencies. *Bull Rheum Dis* 1997;4fi:4-6.