

Rheumatological Emergencies in Clinical Practice

R Handa*, P Aggarwal*, J P Wali*

Rheumatological emergencies are fairly common in clinical practice. However, the average physician with little or no formal training in rheumatology often feels unsure about how best to tackle these acute problems. Such patients are routinely referred to orthopaedic surgeons, while in most instances the physician can easily manage the problem. The present write up is meant to acquaint clinicians with rheumatological emergencies likely to be encountered in day-to-day practice.

Classification of Rheumatological Emergencies

For purposes of convenience, rheumatological emergencies can be divided into 2 broad categories :

A. True Rheumatological Emergencies

1. Acute low backache
2. Acute gout
3. Acute arthritis
 - (a) Arthritis arising *de novo*
 - (b) Acute exacerbation in a patient with chronic arthritis, e.g., rheumatoid flare
4. Lupus flare
5. Systemic necrotizing vasculitides
6. Scleroderma renal crisis
7. Catastrophic antiphospholipid syndrome
8. Erythema nodosum

B. Medical Emergencies in Patients with Systemic Rheumatic Disease

e.g.,

- NSAID induced gastrointestinal bleed
- Acute left ventricular failure (LVF) in lupus

nephritis with hypertension (HT)

- Intracranial bleed in lupus nephritis with HT
- Tuberculous meningitis in SLE
- Acute adrenal insufficiency due to sudden steroid withdrawal
- Seizures in SLE
- Cyclophosphamide induced haemorrhagic cystitis
- Drug (immunosuppressive) induced bone marrow suppression, etc.

The present article discusses only the true rheumatological emergencies. Medical emergencies like LVF, GI bleed, seizures, etc., in a setting of systemic rheumatic disease are handled like in any other setting and are not discussed in this article.

Acute Low Back Ache

Low backache (LBA) is an extremely common problem in medical practice. Individuals especially at risk of LBA include manual labourers, truck drivers, material handlers, medical care providers e.g., who shift trolleys and patients, persons caring for infants (repeated lifting from cribs, play pens etc.), weight lifters, dancers, divers, and gymnasts. The notable thing about acute LBA is its favourable natural history (Table I).

Table I : Acute Low Back Ache (Natural History)

- 40% cases recover within 1 week
- 80% cases recover within 3 weeks
- 90% cases recover within 6 weeks
- Only 7-10% experience symptoms for > 6 months
- Only 1% require surgical intervention

The important causes of LBA are tabulated in Table II. The ones that can present acutely are marked

* Rheumatology Division,
Department of Medicine,
All India Institute of Medical Sciences,
New Delhi-110029

with an asterisk (*). A good history and thorough physical examination are mandatory.

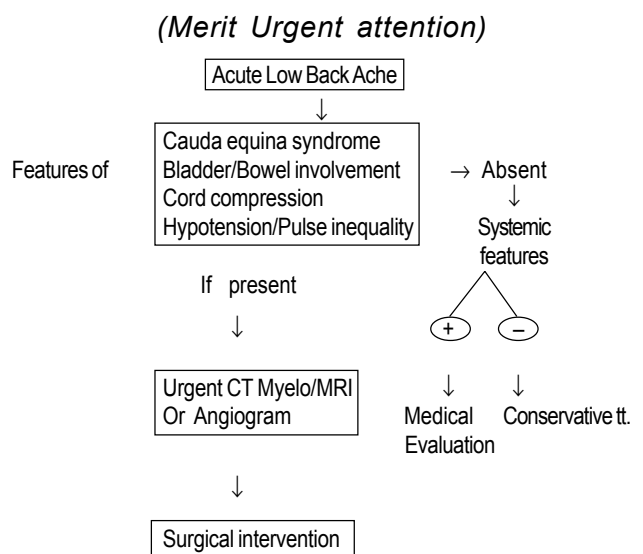
Table II : Causes of LBA (Acute causes marked*)

1. Seronegative spondyloarthropathies (SpA)
 - Ankylosing Spondylitis
 - Psoriatic SpA
 - Inflammatory bowel disease
 - Reiter's syndrome*
 - Reactive Arthritis*
 - Undifferentiated SpA
2. Neoplastic disease
 - Multiple myeloma
 - Metastatic deposits
3. Infections
 - Pott's spine
 - Epidural abscess*
4. Metabolic
 - Osteoporotic fracture*
 - Paget's disease
5. Mechanical causes
 - Prolapse intervertebral disc*
 - Lumbar canal stenosis
 - Spondylolysis/spondylolisthesis
6. Referred pain
 - Pelvic inflammatory disease
 - Cystitis
 - Prostatitis
 - Pancreatitis
 - Pancreatic tumour
7. Miscellaneous
 - Rupture of abdominal aortic aneurysm*
 - Aortic dissection*
 - Psychosomatic
 - Malingering

The first task of the clinician should be to rule out/rule in medical emergencies, viz., cauda equina

syndrome, epidural abscess, rupture of aortic aneurysm or dissection of aorta. The clinician should look for saddle anaesthesia, bladder/bowel involvement, asymmetric loss of reflexes, pulse inequality, hypotension, or circulatory instability. A neurological compromise or features of cord compression warrants urgent CT myelogram or MRI and neurosurgical intervention. Cardiovascular catastrophes warrant angiography and surgery (Figure 1).

Figure 1 : Red Flag Signs in Acute Low Back Ache



After ruling out the emergent medical conditions, the clinician should ascertain if the LBA is mechanical or inflammatory (Table III). Most of the cases of LBA are mechanical in origin (90% episodes)^{1,2}. Similarly malignancy as a cause of LBA is very uncommon (only 1 in 1,000 patients). Clinical evaluation helps one to arrive at a diagnosis in most cases. Investigations in acute LBA should be kept to a minimum³. Over-reliance on x-rays should be avoided. Most often the x-rays are normal. Several arguments³ have been advanced against over-reliance on radiographs, namely:

- Patients with back pain of mechanical origin often have normal x-rays.
- In patients < 50 years of age the yield of abnormal findings is 1:2,500⁴.

- Patients with radiologic abnormalities are often asymptomatic.
- Congenital abnormalities frequently seen on radiographs like spina bifida, Schmorl's nodes are unlikely causes of back pain.
- After the age of 50, nearly 67% of normal individuals show degenerative changes.
- 2/3rd patients with radiographic evidence of lumbar disc degeneration are asymptomatic.
- Significant radiation.

Table III : Differentiating Inflammatory from Mechanical LBA

	Inflammatory	Mechanical
Classical example	SSpA	PVD
Age of onset	Typically < 40 years	Any age
Onset	Insidious	May be acute
Precipitating factor	Usually none	Strenuous task, e.g., Weight lifting
Morning stiffness	+ +	-
Improves with	Exercise	Rest
Acute phase reactants	↑↑	Normal
Peripheral joint involvement	May be present	Absent

SSpA = Seronegative spondyloarthritis

PVD = Prolapse intervertebral disc

Patients who need laboratory investigations/radiological work up are listed in Table IV⁵. All patients where the basic history and physical examination are normal should be subjected to six weeks of conservative therapy. This is because the natural history of LBA is favourable and 90-95% patients improve within 2 months. The catchword should be "restricted physical activity" rather than absolute bed rest, which is often counter productive. No more than 2 days of bed rest is advisable. Progressive mobilization and exercise should follow this as the patient's pain improves. Analgesics (like paracetamol, tramadol, etc.) in combination with muscle relaxants can be used. A lot of clinicians prefer to use NSAIDs for their analgesic effect. Heat/cold application depending upon personal preference is permitted. After the acute stage, physiotherapy should be

started. Surgical intervention is needed in only 1 in 100 patients. The indications include fractures/dislocations, epidural abscess, Pott's spine, tumours, aortic dissection/aneurysm rupture.

Table IV : Identifying patients with Low Back Ache who need laboratory testing/x-ray⁵

- Trauma
- Neurologic dysfunction
- Sphincter involvement
- Constitutional/Systemic features
- Previous malignancy
- Older age
- Drug/alcohol abuse

Acute Gout

Acute gout is one the commonest rheumatological emergencies. The patient, usually a male, presents with acute pain in one of the lower limb joints. The presentation is usually with monoarthritis (single joint) or oligoarthritis (2-4 joints). Polyarticular presentation, though known, is very rare in gout. Gout is uncommon in menstruating females because estrogen is uricosuric. One should be extremely reluctant to make a diagnosis of gout in females of childbearing age with polyarticular joint disease.

The diagnosis of gout is made on clinical grounds. Confirmation is by crystal identification after synoviocentesis. Serum uric acid levels may be normal during an acute attack because ACTH released in response to stress is uricosuric. Joint aspiration is mandatory in patients with acute monoarthritis.

Conventionally colchicine has been used to treat acute gout. However, NSAIDs give equal results and are better tolerated⁶. Many rheumatologists prefer indomethacin though other NSAIDs like diclofenac, naproxen, etc., may give equal results. Colchicine is used as 1 Tablet (0.5 mg) every hour till one of the 3 endpoints is reached:

- (a) Clinical relief
- (b) Maximum dose of 6 mg reached

(c) Gastrointestinal toxicity

The use of small, repeated doses is intended to minimize G.I. toxicity and offers no pharmacotherapeutic advantage. Despite this, 80% of the patients have G.I. toxicity which may be severe. This has resulted in most rheumatologists preferring NSAIDs over time honoured colchicine. Intravenous colchicine is not available in India.

In patients with renal failure or previous G.I. bleed where NSAIDs may be contraindicated, corticosteroids may be used. For acute gout 20-40 mg of prednisolone is used for 5-6 days and then tapered over 1-2 weeks⁷. Joint aspiration and intra-articular steroids may also be employed in patients where systemic corticosteroids have to be avoided.

Allopurinol should never be used in patients with acute gout unless the patient's pain has been brought under control by use of NSAIDs/colchicine and all signs of inflammation have subsided. Patients already on allopurinol should continue it.

Acute Arthritis

Acute arthritis may start afresh (*de novo*) or represent acute exacerbation in a patient with pre-existing arthritic illness like rheumatoid arthritis. The causes of *de novo* arthritis can be conveniently divided into :

A Acute Monoarthritis

- Septic arthritis
- Gout
- Trauma

B Acute Oligo/Poly Arthritis

- Reactive arthritis/Reiter's syndrome
- Viral arthritis
- Rheumatic fever
- HIV
- Disseminated gonococcal infection

Rarely, rheumatoid arthritis may have an acute onset. The clinician would do well to recognize that several arthritic illnesses resemble each other

at the onset and proper categorization into a specific disease entity may not be possible at the first patient encounter. Repeated clinical observation is mandatory.

Acute monoarthritis should be considered a medical emergency. The condition warrants immediate joint aspiration. Synovial fluid should be aspirated to rule out pus in the joint. The synovial fluid should also be subjected to crystal studies preferably using polarized microscopy. Table V lists the important practical points about synovial fluid analysis⁸.

Table V : Synovial Fluid Examination : Key Points⁸

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- Sample should be anticoagulated with heparin or liquid EDTA. Powder EDTA causes confusion in crystal identification.
 - Viscosity, mucin clot, protein, glucose, ANA and RF in synovial fluid do not confer any meaningful information. These need not be done.
 - Total and differential WBC counts, culture, Gram's stain, ZN (Ziehl Neelsen's) stain and crystal identification should be performed on all fluids.
 - WBC count should be done normally using saline as diluent. Acid diluents cause mucin to clot.
 - High viscosity of synovial fluid interferes with counts in automated blood counters. Manual counts are preferable.
 - Polarized light microscopy is ideal for crystal identification. However, useful information can also be obtained on light microscopy.
 - The ordinary light microscope can be adapted to perform polarizing light microscopy using economical polarizing plates.
 - Crystal identification requires experience.
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Septic Arthritis

The clinical profile of acute bacterial arthritis includes the rapid onset of severe joint pain,

warmth and tenderness either in a normal individual, immunocompromised individual like diabetics, transplant recipients or in patients with prosthetic joints. Usually a single joint is involved. However, in 10-15% patients septic arthritis may be polyarticular⁹. Large joints like knee and hip are commonly involved. The elderly patients may not manifest fever or other signs of inflammation. *Staph. aureus* is the commonest organism followed by streptococcal species, Gram negative organisms, and anaerobes. Gonococcal infections should be suspected in sexually active patients. The classical triad of disseminated gonococcal infection (DGI) comprises acute tenosynovitis, dermatitis and arthritis. Tenosynovitis is most common over the dorsum of hands and wrists, whereas the septic arthritis in DGI commonly affects the knee, ankle, wrist and the elbow in descending order of frequency. Maculopapular skin lesions are typical but may be pustular, vesicular or bullous. Local genitourinary lesions are unusual in DGI.

Joint aspiration is mandatory in patients with suspected septic arthritis. Joint drainage is a must to drain the intra-articular pus. This can be achieved by large bore needle aspiration or lavage, which may be arthroscopic. Surgical arthrotomy is usually required for septic hips/shoulders, if osteomyelitis co-exists with septic arthritis and if joint infection is not controlled by 5-7 days of needle/arthroscopic drainage. Antibiotics need to be given for 2 weeks parenterally followed by 2-6 weeks of oral therapy. The duration of therapy is governed by clinical response.

Drugs used to treat DGI include parenteral ceftriaxone till clinical response, and then followed by cefuroxime axetil or co-amoxyclav. *Staph. aureus* is best treated with cloxacillin. Gram negative organisms require addition of aminoglycosides. The management of gouty arthritis has been outlined *vide supra*. Radiology has a limited role in acute monoarthritis. X-rays help only to exclude fracture in patients with history of trauma.

Reactive arthritis may be post dysenteric or follow non-gonococcal urethritis/cervicitis with chlamydia. The classical triad of Reiter's syndrome, which includes urethritis, conjunctivitis and arthritis, may not be present in all the patients. In the acute stage, reactive arthritis and Reiter's syndrome are managed by NSAIDs. Antibiotics have virtually no role, although long-term antibiotics (lemecycline) have been tried in sexually acquired reactive arthritis. Patients with chronic joint involvement may be candidates for methotrexate or sulfasalazine. Viral arthritides are most often self-limited and managed symptomatically. The clinical profile and treatment of rheumatic fever are well known and will not be discussed here. HIV is also known to have a variety of rheumatic manifestations including Reiter's syndrome, psoriatic arthritis, arthralgias as part of acute HIV syndrome, HIV associated arthropathy and the "painful articular syndrome". The latter presents as acute, severe, sharp pain in the joint especially the knees, elbows and shoulders. It is often self-limited and may require narcotic analgesics. It is thought to be due to the direct effect of HIV on the joint.

Acute flares of pre-existing arthritis like rheumatoid arthritis are best treated with a short course of corticosteroids with escalation of DMARD therapy. Differentiation from disease "slip out" is mandatory. The latter situation is analogous to secondary failure to sulfonylureas in diabetes mellitus and needs change of DMARD. Another common presentation is sudden worsening of knee pain in osteoarthritis. This may be due to osteoarthritis flare, septic arthritis, rupture of Baker's cyst, pseudogout (CPDD disease), osteonecrosis or Anserine bursitis. Synovial fluid analysis should be carried out and, if infection is excluded, intra-articular steroid injection can be very useful in providing relief.

Lupus flare

A lupus flare may be precipitated by stress, exposure to sunlight, steroid reduction/withdrawal, pregnancy, infection, etc. Infection needs to be

ruled out in febrile lupus patients before attributing fever to SLE. A low TLC and normal CRP favour lupus activity while leukocytosis and raised CRP suggest infection. Lupus flares are best treated by withdrawal of a precipitating factor, if any, and by increasing corticosteroid dosage.

Systemic vasculitides

In general, flares of systemic vasculitides require escalation of immunosuppressive treatment, viz., corticosteroids, azathioprine, cyclophosphamide etc., depending on the extent of organ involvement and the specific diagnosis. Life threatening situations are tackled by intravenous methyl prednisolone pulses (usually 1 Gm daily X 3days).

Scleroderma renal crisis

Approximately 80% of the renal crises occur in patients with diffuse cutaneous systemic sclerosis within the first 4-5 years of disease. The problem is much less frequent in patients with long-standing disease. Less than 1% of the patients with limited scleroderma develop renal crisis. Blood pressure is usually abnormal but normotensive renal crisis is also known (11% patients). Risk factors include diffuse skin disease, new unexplained anaemia, new cardiac events (like pericardial effusion or congestive cardiac failure), high dose steroids, and the presence of anti-RNA polymerase III antibody. The syndrome is characterized by accelerated hypertension, rapidly progressive renal failure, increased plasma renin activity, microangiopathic haemolytic anemia and thrombocytopenia. Congestive heart failure and pericardial effusions are common, as are dyspnoea, headache, seizures, fundoscopic changes and urinary abnormalities¹⁰.

The treatment of choice for scleroderma renal crisis is therapy with ACE inhibitors, which reverse underlying hyper reninaemia and control HT. Other anti hypertensives can also be added if needed. ACE inhibitors are of value even in normotensive renal crisis. In some patients renal failure ensues despite early and vigorous

treatment. ACE inhibitors should be continued in these patients. Even in patients on dialytic support continuation of ACE inhibitors often leads to enough renal recovery to permit dialysis discontinuation after 6-18 months. No studies on prophylactic role of ACE inhibitors are available¹¹.

Plasmapheresis, corticosteroids and immunosuppressives have no role in the management of scleroderma renal crisis.

Catastrophic Antiphospholipid Syndrome (CAPS)

The presentation in CAPS is essentially that of multi-organ failure in a patient with APS. Occasionally patients may not give history of APS and CAPS may be the first presentation making diagnosis difficult. Pulmonary, gastrointestinal, cardiac, renal and neurologic involvement is common as in systemic inflammatory response syndrome. Table VI outlines the important points that need to be remembered while diagnosing APS. The common precipitating factors include surgery, infections, oral contraceptives and anticoagulant withdrawal. These are present in more than one-third patients. Disseminated intravascular coagulation is common. Death occurs in as many as 60% patients from a variety of causes (myocardial failure, ARDS, renal failure, CNS causes, or a combination). Plasmapheresis is useful in patients unresponsive to conventional therapy with heparin, aspirin, steroids, etc. It needs to be stressed that the term "CAPS" does not apply to a variant of APS but rather to a time limited episode¹².

Table VI : Caveats in diagnosis of APS

- The mere presence of aPL is not sufficient to diagnose APS.
- aPL antibodies should be demonstrable in moderate to high titres.
- aPL should be positive on at least 2 occasions 6 weeks apart. Transient aPL are not uncommon in infections.
- Both LAC and aCL should be ordered since

30% patients have one without the other.

- IgG aCL is more important than IgM and IgA. It is preferable to obtain all three.
- While interpreting LAC remember that mere prolongation of screening tests is not important. Failure of coagulation test to normalise after addition of plasma is important since it rules out clotting factor deficiency. In APS addition of phospholipid normalises the test.

APS = antiphospholipid syndrome, aPL = antiphospholipid antibodies, aCL = anticardiolipin antibodies, LAC = lupus anticoagulant

Erythema nodosum

Erythema nodosum (EN) is the commonest panniculitis seen in clinical practice. Panniculitides are a rare group of disorders characterized by inflammation of subcutaneous fat. The patient presents with painful, erythematous nodules mainly on the shin and lower limbs. Joint pains are seen in nearly 50% of the patients with E. nodosum. Ankles are the commonest joints affected. Other joints, which may be affected, include knees, hips, wrists, fingers, shoulders, and elbows. The condition may be idiopathic. Known triggers include infections (streptococcal pharyngitis, TB, etc.); drugs (sulfonamides, penicillin, oral pills, etc.); sarcoidosis; lymphoma; inflammatory bowel disease; and Behcet's Syndrome. Usually EN is a self-limiting disease. NSAIDs may be used to give pain relief. If acute pain persists or is unresponsive to NSAIDs, a short course of steroids may be used. For recurrent attacks of unknown cause potassium iodide, colchicine, or dapsone may be tried.

Conclusions

Rheumatological emergencies are common in clinical practice. In most instances, the emergency can be handled by the physician himself/herself. Rheumatology referral or orthopedic intervention is needed only in a fraction of the patients.

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