REVIEW

The emergency room in systemic rheumatic diseases

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Complications of systemic rheumatic diseases frequently have protean manifestations and may present a diagnostic problem. Patients with connective tissue diseases and vasculitides may have dangerous or life threatening conditions, which must be recognised and treated promptly to prevent rapidly evolving morbidity and mortality. Knowledge of possible emergencies in the context of a defined rheumatic disease may aid in promoting a high index of suspicion and contribute significantly to the timely diagnosis of many potentially dangerous conditions. This review is written for the emergency room physician and discusses the early recognition of selected emergencies in the context of a defined rheumatic disease.

Patients with rheumatic complaints may account for up to 8% of all emergency room (ER) visits.[1] Those patients with connective tissue diseases and vasculitides may present with dangerous or life threatening conditions, which must be recognised and treated promptly to prevent rapidly evolving morbidity and mortality. These emergencies may present protean manifestations, at times mimicking other conditions or misleading with an illusory innocent clinical picture. A missed presentation of some of these threatening conditions may erroneously lead ER personnel to direct the patient back to their rheumatologist for further outpatient work up, which may result in significant delays in diagnosis. Therefore, knowledge of possible emergencies in the context of a defined rheumatic disease may aid in promoting a high index of suspicion and contribute significantly to the timely diagnosis of many potentially dangerous conditions.

The literature usually reviews rheumatic emergencies using the problem oriented approach. This approach is preferable in patients with defined organ or system involvement, such as vasculopathy of the central nervous system (CNS) in systemic lupus erythematosus or Behcet's syndrome, or abdominal emergencies in vasculitides. However, in this review we use the disease oriented approach, describing selected life threatening emergencies of the most common defined rheumatic disorders which are more likely to be missed. Other rheumatic emergencies, which have been reviewed recently or elaborated in standard emergency medicine textbooks, are not discussed.

The review is written for the ER physician in an attempt to facilitate early diagnosis, which may be arrived at or at least suspected in the ER. It should be added that almost all of the described emergencies may also be the initial manifestations of the systemic diseases discussed.

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RHEUMATOID ARTHRITIS (RA)

Septic arthritis in the RA patient

While a suspicion of septic arthritis is always high in any patient with acute monoarthritis, an infected joint in an RA patient is frequently overlooked, particularly during the earlier stages of the infection, due to the presumption that this merely represents a flare up of the underlying disease. Delay in the diagnosis of septic arthritis in these patients averages 1–3 weeks and frequently leads to irreversible joint damage with poor functional outcome.[2] The mortality rate of 20–33% in this setting underlines the significance of prompt diagnosis and treatment.

Any monoarticular or oligoarticular flare up in these patients should be promptly evaluated for the possibility of septic arthritis. Redness over a swollen joint, rather unusual in RA, may raise the suspicion of an infected joint.

Factors inhibiting the correct diagnosis of septic joint in RA include the frequently reported insidious onset of the complication, the absence of fever in 50% of patients, and the polyarticular pattern of joint involvement in up to 20–30% of patients.[2] The prevalence of septic arthritis is increased particularly in RA patients with prolonged erosive disease and/or treated with glucocorticosteroids. Treatment with TNFα blockers may be also associated with a higher rate of septic arthritis as well as with infection by opportunistic or rare pathogens.[3]

Prompt joint aspiration of the septic joint will typically reveal nucleated cell counts above 50 000/mm^3 with neutrophilic predominance, while lower synovial white blood cell counts may be seen rarely, especially in the early stages. The presence of low synovial fluid glucose levels may suggest the diagnosis. Direct smear with Gram stain should be performed and the drained fluid cultured.

The differential diagnosis of septic joint usually includes crystalline induced arthritis.

Abbreviations: AH, alveolar haemorrhage; APLA, antiphospholipid antibodies; AS, ankylosing spondylitis; CA, cricoarytenoid arthritis; CAPS, catastrophic antiphospholipid syndrome; CNS, central nervous system; CT, computed tomography; ER, emergency room; ESR, erythrocyte sedimentation rate; GCA, giant cell (temporal) arteritis; MRI, magnetic resonance imaging; RA, rheumatoid arthritis; RTA, renal tubular acidosis; SGS, subglottic stenosis; SjS, Sjogren's syndrome; SLE, systemic lupus erythematosus; SSc, systemic sclerosis; TM, transverse myelitis; VBI, vertebro-basilar insufficiency; WG, Wegener's granulomatosis

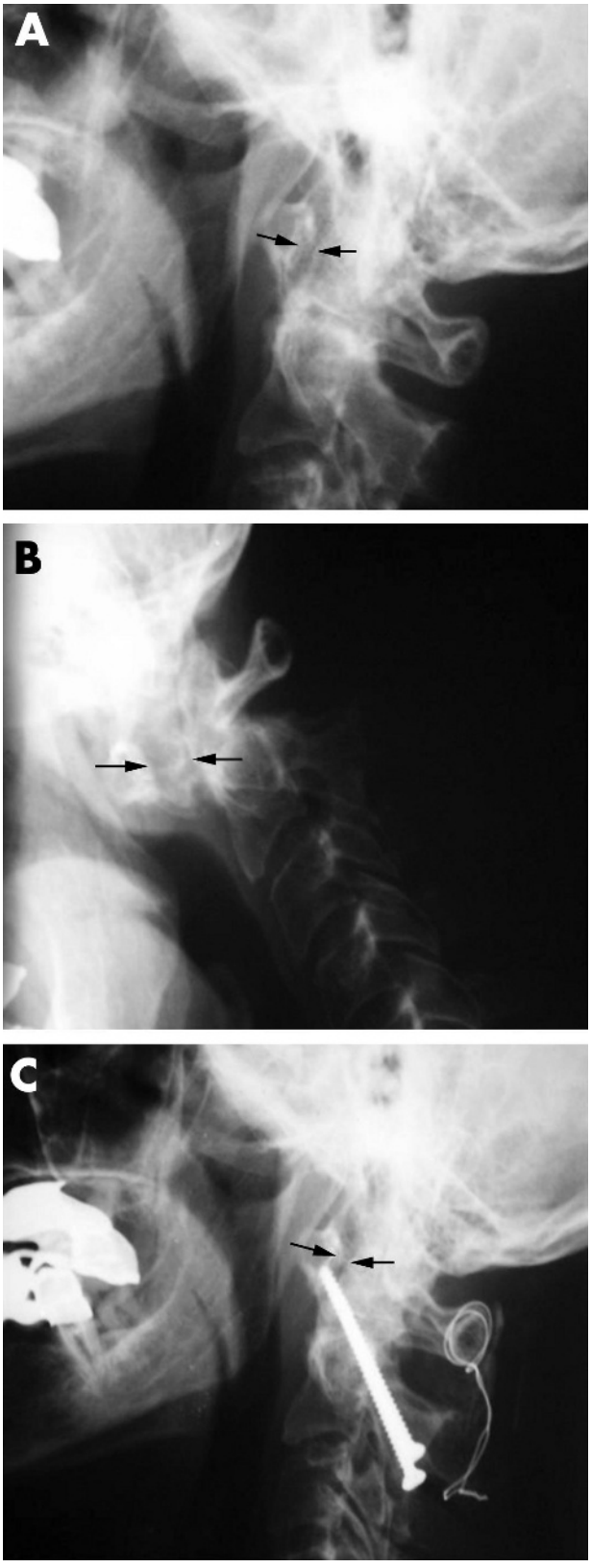

Figure 1 Lateral radiographs of the cervical spine during extension (A) and flexion (B) of the patient's neck. Severe atlantoaxial subluxation becomes evident in the flexed position where the distance (arrows) between the posterior surface of the anterior arch of the atlas and the anterior surface of the odontoid process widens to more than the normal ⩽3 mm. (C) X ray of the cervical spine of the same patient after surgical stabilisation.

Though infrequently reported in the literature, both gout and pseudogout are regularly seen in clinical practice in RA patients, so examination of the synovial fluid for crystals is essential. ''Pseudoseptic arthritis'' of RA is another condition mimicking an infected joint. The clinical presentation of pseudoseptic arthritis is indistinguishable from septic arthritis; however, negative cultures and the short (24–48 h) course of the flare up which resolves without antibiotics differentiate between the two entities.

Any RA patient with suspected infectious arthritis should be hospitalised for appropriate investigations, follow up, and treatment.

Instability of the cervical spine (fig 1A–C)

Cervical spine radiological involvement is a frequent finding in RA patients,[4] with atlantoaxial subluxations particularly dangerous because of the risk of cervical myelopathy. Atlantoaxial subluxations appear typically in patients with erosive seropositive long-standing RA, with a forward dislocation being the most common (occurring in 43% of patients with RA of mean duration of 12 years[5]) and a vertical dislocation being the most ominous.

The first clinical signs raising suspicion of an unstable cervical spine may include new occipital pain and tingling of the fingers. The neurological examination may be difficult and sometimes unrewarding due to severe joint deformities, muscle wasting, and entrapment neuropathies secondary to RA. Sensory disturbances in the extremities are major pointers to a spinal cord lesion. The diagnosis is easy in most cases if the index of suspicion is high. Both forward and vertical dislocations of the atlantoaxial joint can be seen on a lateral radiograph of the upper cervical spine in maximal active anterior flexion.[6] As a word of caution, when examining RA patients in general, it may be unwise to passively flex their cervical spine maximally, as C1–C2 subluxation may be initially clinically silent. This may be especially important to remember after trauma such as falls or vehicular accidents. Sudden death has been reported in patients with RA and atlantoaxial subluxation.[7]

Computed tomography and, especially, magnetic resonance imaging (MRI) are useful whenever plain radiographs leave any doubt about the diagnosis. Most patients with atlantoaxial dislocation associated with myelopathy will benefit from surgical treatment stabilising the cervical spine.[8]

Cricoarytenoid arthritis (CA)

Involvement of the cricoarytenoid joint has been reported in up to 30% of RA patients,[9] but in most this involvement is probably asymptomatic. Clinical signs of CA involvement frequently appear secondary to laryngeal manipulation or infection and include hoarseness, sensation of a foreign body, fullness, or tension in the throat, and inspiratory stridor.[10] Concomitant wheezing may lead to a mistaken diagnosis of asthma or bronchitis. Laryngoscopy, showing oedema, reduced vocal cord motility, and arytenoid cartilage asymmetry, and computed tomography (CT) imaging have both been used for the diagnosis of CA. In the non-emergency setting, treatment with systemic or locally injected glucocorticosteroids is usually effective. In patients with acute airway compromise intubation may be difficult and traumatic, requiring tracheostomy.[10]

ANKYLOSING SPONDYLITIS (AS)

Spinal fractures

The pathologically rigid spine of AS patients gradually becomes osteoporotic and is thus increasingly vulnerable to fracture. It has been calculated that up to 14% of AS patients will experience a vertebral fractures during their lifetime.[11] Neurological complications are frequent and about two thirds

of these patients may not completely recover neurologically.[12] The most common manifestation of spinal fracture is pain, usually localised, which is aggravated on movement and thus different from the inflammatory pain of AS. The localised bleeding and oedema associated with the fracture may create a mass effect with presentations of acute radiculopathy and myelopathy. Early diagnosis, confirmed by appropriate imaging studies, should lead to urgent immobilisation and surgical fixation in patients with vertebral instability or bracing in patients with stable fractures.

Factors associated with a higher likelihood of missed diagnosis include the absence of major spinal trauma in more than 50% of AS patients with vertebral fracture[12] and failure of standard imaging to detect a fracture, usually of the low cervical spine, due to non-displacement of these fractures and their small size when only syndesmophytes are involved.[13] The differential diagnosis includes aseptic spondylodiscitis of AS.[14]

Atlantoaxial subluxation

Patients with longstanding AS may develop instability of the cervical spine due to atlantoaxial subluxation with resulting myelopathy, similar to that of RA.[15]

SYSTEMIC SCLEROSIS (SSc)

Renal crisis

Scleroderma renal crisis is associated with approximately 20% mortality with an additional 20% of patients remaining on chronic dialysis treatment after the crisis has resolved.[16] This complication usually develops in patients with early (during the first 4 years after diagnosis) diffuse scleroderma and has a vascular non-inflammatory nature. Glucocorticosteroids have been implicated in precipitating renal crisis.[17] In its classic form, when an SSc patient presents with headache, malignant hypertension, high serum creatinine, and evidence of microangiopathic anaemia with thrombocytopenia, the diagnosis of renal crisis is straightforward. It should be remembered, however, that approximately 10% of these patients do not develop major hypertension. In these cases progressive fatigue may be a leading complaint and the diagnosis suspected with the finding of raised serum creatinine and/or microangiopathic haemolytic anaemia on peripheral blood smear.[18] Angiotensin converting enzyme inhibitors, even in the presence of progressive renal dysfunction and dialysis, are the cornerstones of the treatment of scleroderma renal crisis.

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

Alveolar haemorrhage (AH)

AH is a rare but grave complication of SLE with mortality of up to 70–90%.[19] The cause of AH in SLE is capillaritis, which is pathogenetically similar to lupus microangiopathy of the kidney and related to the deposition of immune complexes.[20] The most frequent clinical features on presentation are dyspnea with pulmonary infiltrates (100%) and high fever (82%), while haemoptysis may be seen only in half of these critically ill patients.[21] The differential diagnosis includes infectious pneumonia and acute lupus pneumonitis. In the absence of haemoptysis, dropping red cell indices in the presence of a dense pulmonary infiltrate and concomitant progressive glomerulonephritis (in approximately 75% of patients) are frequent pointers to the sometimes difficult appreciation of AH in SLE.[22 23] The diagnosis in equivocal cases may have to be confirmed by bronchoscopy with broncho-alveolar lavage, revealing haemosiderin laden or pigment laden macrophages and negative bacterial and fungal cultures.[22] Early treatment with high dose glucocorticosteroids, cyclophosphamide, and/or plasmapheresis may be critical to patient survival.

Transverse myelitis (TM)

TM is a devastating neuroinflammatory disorder affecting approximately 2% of patients with SLE.[23] SLE associated TM can be caused by CNS vasculitis or result from arterial thrombosis, related in some cases to antiphospholipid antibodies (APLA).[24] If not recognised and treated promptly, TM may lead to irreversible paraplegia. This rapidly progressive disorder usually presents with weakness, paraesthesias, or neuropathic pain in the lower extremities, often associated with back pain and bladder dysfunction. Sensory loss, usually at the mid-thoracic level, may be found in most but not all SLE patients with TM.[25] A high level of suspicion is needed to correctly interpret the diffuse TM related complaints of SLE patients, particularly those with concurrent fibromyalgia syndrome or back/radicular pain.[26] Examination by an experienced neurologist may be critical to the diagnosis of TM in these patients. When TM is suspected, MRI should be carried as soon as possible for the characteristic finding in TM of increased signal intensity and atrophy of the spinal cord. On the other hand, a normal MRI picture may be seen occasionally in patients with the characteristic clinical picture of TM.[25] Cerebrospinal fluid showing pleocytosis or increased IgG index may be used to confirm the inflammatory pathogenesis of TM in patients with SLE related vasculitis of the CNS. The presence of APLA may point to a thrombotic pathogenesis of TM in some cases.

Catastrophic antiphospholipid syndrome (CAPS)

SLE patients as well as patients with primary antiphospholipid syndrome may develop CAPS, which is an extreme variant of the antiphospholipid syndrome with predominant occlusion of small vessels, mainly affecting parenchymal organs.[27] The most common known trigger of CAPS is infection, with trauma, surgery, oral contraceptives, neoplasia, and warfarin withdrawal also reported. In almost half of the cases no obvious precipitating factors have been identified.[27] Mortality is approximately 50%, and diagnosis may be difficult because of the wide spectrum of potential manifestations of CAPS. Seventy per cent of patients present with renal dysfunction, 66% with pulmonary complications (ARDS and pulmonary emboli being the most frequent), and 60% with cerebral symptoms (infarcts, seizures, venous occlusions). Other frequent thrombotic manifestations include myocardial infarction and skin necrosis.[28] Livedo reticularis is an important finding on physical examination, pointing to the potential diagnosis. Thrombocytopenia persists in more than 60% of patients and is another significant diagnostic feature of CAPS. The differential diagnosis includes thrombotic thrombocytopenic purpura, marantic endocarditis with multiple embolic events, SLE vasculitis, and heparin induced thrombocytopenia-thrombosis syndrome.[27] High serum levels of APLA are characteristic of CAPS. Immediate treatment should include intravenous heparin and high dose glucocorticosteroids, with intravenous immunoglobulins and plasma exchange being the second line therapies.[27]

Pericardial tamponade

Signs and symptoms of pericarditis in SLE patients are typical of those of pericarditis in general.[29] Cardiac tamponade is a rare event and usually presents with progressive dyspnea. Pericardial involvement occurs predominantly in patients with widespread active SLE and manifests as fatigue, Raynaud's phenomenon, joint pains, and low serum levels of complement factors C3 and C4.[30] A low threshold for the referral of SLE patients with chest pain and/or dyspnea already in the ER to echocardiography may contribute to early diagnosis and successful treatment. The association of large pericardial effusions with active nephritis, Libman-Sacks endocarditis, and myocardial dysfunction was recently reported in SLE patients.[30]

SJOGREN'S SYNDROME (SjS)

Hypokalaemic paralysis

Distal renal tubular acidosis (RTA) occurs in approximately 30% of patients with primary SjS. Asymptomatic in most patients, RTA may lead to symptomatic hypokalaemia in others. Hypokalaemic paralysis leading to quadriparesis and, rarely, to respiratory arrest has been repeatedly reported in SjS and should always be considered in a patient with SjS presenting to the ER with unusual weakness.[31 32] Low serum potassium levels along with hyperchloraemic metabolic acidosis and abnormally acidified urine may be diagnostic. Immediate treatment consists of vigorous potassium replacement and intensive monitoring and support.

GIANT CELL (TEMPORAL) ARTERITIS (GCA)

Loss of vision

Visual loss is the most feared complication of GCA. If it occurs, it is irreversible in most patients and may even worsen during the first days following initiation of treatment with high dose glucocorticosteroids.[33] The presence of other ischaemic complications related to GCA, elevated thrombocyte count, and visual hallucinations have been thought to predict the development of irreversible blindness in these patients. Of interest, a lower erythrocyte sedimentation rate (ESR) independently predicted a higher risk of visual loss in GCA patients.[34–37] Possibly, an ESR that was only mildly elevated failed to alert to the probability of GCA in these patients and dissuaded the attending physicians from initiation of steroids. Immediate administration of glucocorticosteroids in a patient with a clinical picture of GCA is best to prevent permanent visual loss. Optimally, these patients should be hospitalised for a biopsy of the temporal artery, which may be safely performed after a few days of treatment with glucocorticosteroids.

Vertebro-basilar insufficiency (VBI)

GCA is known to affect the extracranial part of the vertebral arteries in 75–100% of patients and frequently causes neurological deficits in the vertebro-basilar circulation territory.[38] When a part of the classic presentation of GCA with headache, jaw claudication, scalp hypersensitivity, polymyalgia syndrome, and elevated ESR, the signs of VBI are easily attributable to the disease. However, the diagnosis of GCA may be significantly delayed in the elderly patient presenting with gait disturbance, dizziness or vertigo, vomiting, and sometimes slurred speech, if the classic symptoms of the disease are not apparent. Treated GCA patients may also relapse during tapering of glucocorticosteroids with only progressive neurological deficit, even if the first presentation of GCA had had a full blown systemic character (unpublished personal data). Untreated GCA in these patients may progress to bilateral vertebral artery occlusion, a condition with 75% mortality.[38] Thus, ESR as a screening test for active GCA should be promptly performed in all known GCA patients presenting with a new neurological symptom or any elderly patient with possible VBI, particularly when accompanied by headache or fever. Conventional or MR angiography may differentiate GCA involvement of the vertebral arteries from an atherosclerotic process via the different localisation of narrowings.[38] As in classic GCA with temporal arteritis, immediate high dose glucocorticosteroid treatment should be administered to patients with symptomatic GCA involvement of the vertebral arteries.

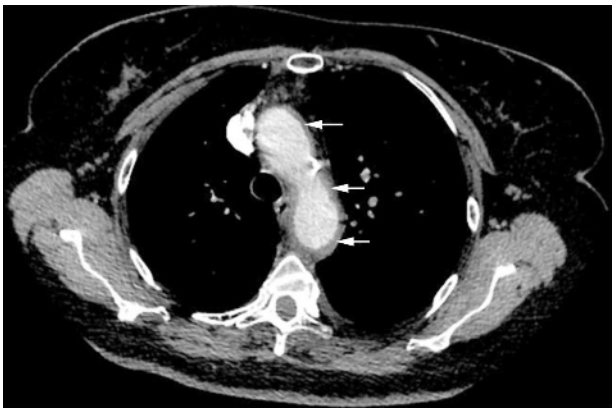

Figure 2 CT angiograph showing aortic wall thickening (arrows) of the aortic arch, compatible with aortitis, in a 67 year old patient complaining of recurrent bouts of upper chest and back pain, in whom ESR was elevated at 68 mm/h.

Aortitis (fig 2)

GCA may be associated with a large artery complication in up to 27% of patients, with aortic aneurysms, ruptures, and dissections, aortic valve incompetence, and aortic arch syndrome reported.[39] Aneurysms of the ascending thoracic aorta occurred 17 times more often in patients with GCA than in a control group, while abdominal aortic aneurysms were 2.5 times more frequent.[40] Aortic pain may be an early sign of the catastrophic event and its timely recognition is of primary importance. Best known is the pain of acute thoracic aorta dissection, which is classically described as of abrupt onset, anterior chest or posterior suprascapular in location with downwards radiation, searing or tearing in character, and pulsating, sometimes with an odd sensation in the legs. Sometimes partial aortic tears or an evolving aortic dissection may masquerade as angina pectoris or atypical chest or upper back pain, with only a high level of suspicion leading to timely diagnosis in these patients. Echocardiography may be an effective means of evaluation when disease of an ascending aorta is suspected. CT or MRI is usually required for imaging of the descendent thoracic and abdominal aorta when the clinical presentation suggests such involvement.[41]

WEGENER'S GRANULOMATOSIS (WG)

Subglottic stenosis (SGS)

SGS occurs in approximately 20% of patients with WG.[42] It can manifest with hoarseness, cough, dyspnea, and/or stridor. Airway obstruction results not only from the subglottic lesion itself but also from tracheobronchial secretions, trapped at the level of stenosis. Initial diagnosis may be easily confused with respiratory symptoms from coexistent pulmonary disease, and the stridor may be misdiagnosed as the wheeze of bronchial asthma.[43] All patients in whom SGS is being considered should be immediately evaluated by laryngoscopy. Close monitoring for signs of acute airway compromise is essential, with tracheostomy performed promptly when indicated. It should be mentioned that the SGS in WG is not necessarily reflective of overall disease activity and is frequently resistant to systemic immunosuppressive therapy, with fibrosis reported despite aggressive treatment.[42 43] Intratracheal dilatation with intralesional injections of glucocorticosteroids may be an effective means of treating SGS.[42]

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