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ADULT ONSET STILL'S DISEASE

Definitions:

Adult onset Still's Disease or systemic onset juvenile rheumatoid arthritis – an inflammatory disorder with sudden onset, daily, high spiking fevers, arthritis or arthralgia, and an evanescent rash.

Pathology:

Etiology unknown. Infectious sources have been suggested as triggers including rubella, echovirus, EBV, CMV and parvovirus. HLA linkages have been implicated however nothing conclusive.

Epidemiology:

- 0.16 cases / 100,000 persons / year
- Equal sex distribution
- Age – bimodal w/ peaks between 15-25 and 36-46

Clinical presentation and physical exam:

- Sudden onset of high spiking fevers – usually once daily, often in evening; returns to normal w/o antipyretics in 80% of cases.
- Arthralgias/myalgias – universal
- Arthritis – almost universal, often starts oligo- then polyarticular. Often occurs *after* the onset of the fevers.
  - Knees > wrists > ankles > elbows > PIPs > shoulders.
  - May become destructive.
- Rash – present in 85-88%. Salmon-pink, macular or maculopapular, frequently evanescent and often associated w/ the fever spike. Called pathognomonic.
  - Located on trunk and proximal extremities.
  - Koebner's phenomenon – rash may be precipitated by mechanical irritation from rubbing of clothing or hot water.

Diagnosis:

At least 6 criteria systems have been proposed:

Major criteria:

- Fever >39°C lasting >1 wk
- Arthralgia or arthritis lasting >2 wks
- Rash – macular/maculopapular, nonpruritic, salmon-pink eruption, evanescent.
- Leukocytosis >10000/mm^3 with > 80% PMNs

Minor criteria:

- Sore throat
- Recent development of significant lymphadenopathy
- Hepato- or splenomegally
- Elevated aminotransferases (73%) and LDH
- Negative ANA (92%) and RF (93%)

Diagnosis (cont):

Japanese criteria are any 5 of above w/ at least two from the major category (93% sensitive).

Must exclude – infection, malignancy, PAN, SLE, vasculitis or other rheumatic diseases.

Other labs: Elevated ESR (99%)
Anemia (68%)
Elevated ferritin (70%) – elevations correlate w/ disease activity; suggested as marker to monitor disease response to treatment.

Radiologic findings:

Nonspecific at presentation

With time there is cartilage narrowing, particularly MCP and intercarpal joint spaces of the wrist.

Disease course:

1/5 experience long-term remission within 1 year

1/3 have complete remission followed by 1 or more relapses; relapse is less severe and shorter.

Remainder have chronic disease often w/ destructive arthritis

Markers of chronic course include early polyarthritis or root joint involvement (i.e. shoulders or hips)

Marker of poor prognosis includes need for > 2 years of systemic steroid therapy

Patients have higher level of pain, physical and psychological disability but less than other chronic rheumatic diseases. Functional status is very good.

Incidence of amyloidosis may be as high as 30% within 10 yrs of onset.

Treatment:

NSAIDs – ¼ respond – have good prognosis. There is concern for hepatotoxicity therefore monitor liver enzymes.

High dose ECASA +/- indomethacin.

Systemic corticosteroids in pt w/ severe disease (pts w/ pericardial tamponade, myocarditis, severe pneumonitis, DIC, or increasing liver enzymes for pts treated w/ NSAIDs).

Hydroxychloroquine, sulfasalazine, penicillamine, methotrexate have no controlled studies however are used to treat arthritis w/ anecdotal reports of benefit. Increased toxicity w/ sulfasalazine

Immunosuppressants like azathioprine, cyclophosphamide, cyclosporine used in resistant cases.

References:
1. Primer on the Rheumatic Diseases, Chapter 25, 12th Edition, 2001.
2. Ruddy: Kelley's Textbook of Rheumatology, pp 970-71, 6th Edition, 2001
3. Up to Date. "Adult Still's disease."