Adult Onset Still's Disease

Sir,

Adult Onset Still’s Disease (AOSD) was reported first by Baywaters in 1976 with clinical features identical to those of Juvenile Rheumatoid Arthritis (JRA).

Studies from France, Japan and India comparing and analyzing clinical features, course and prognosis of JRA and AOSD are basically identical. The pathogenesis and treatment are also the same. It seems therefore reasonable to consider them as a single distinct entity irrespective of age of onset and use a single nosological term viz. Still’s disease.

AOSD is uncommon and even less frequently reported. About 30 cases have been reported from Northern India. There are no reports from the south and over 300 cases have been reported in world literature.

Here is a fairly typical case of Still’s disease. A 19 year old boy presented with 12 days history of sore throat, high fever with rigors and spikes, bodyache, pain in joints and erythematous rash all over the trunk which was blanching on pressure. Rash was prominent with spikes of fever and almost disappeared with fall in temperature. On examination he was found to be febrile with non-itching macular erythematous rash on the trunk. He had arthralgia of left hip, left knee and right elbow. Inguinal glands were minimally enlarged and non-tender.

Serial laboratory tests revealed rise in WBC count from 8700 to 13000 /cmm, neutrophils from 69% to 84%, ESR from 15 to 108 normal platelet count and hemoglobin level. Malaria, dengue, infectious mononucleosis, typhoid fever, HIV infection and syphilis were ruled out on the basis of laboratory tests. CXR and abdominal ultrasound were normal. CT abdomen showed mild splenomegaly. Blood cultures were sterile. Rheumatoid factor, ANA, C-ANCA and P-ANCA tests were negative. LE cells were not found in blood. Aspiration and trephine biopsy examination of bone marrow showed mild myeloid hyperplasia and increased iron stores. ASO titer was negative CRP was 96 mg/L (1:16 positive) LDH was 477 IU/L (N=109-193). SGOT was raised. Bilateral lymphnode biopsies showed reactive hyperplasia with no evidence of malignancy. Serum ferritin level was 16887.82 mg/L (N=18.7 to 323 mg/L). Hyperferritinaemia is a powerful marker of AOSD.

This costly and time consuming work up to exclude infections, seropositive autoimmune disorders and certain malignancies may be obviated if a proposed new set of (AOSD) criteria suggested by Fautrel B et al1 (Table 1) and the earlier preliminary criteria proposed by Yamaguchi M et al2 (Table 2) are considered first.

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<th>Table 1</th>
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<td>Major Criteria</td>
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<td>Spiking fever ≥ 39°C</td>
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<tr>
<td>Arthralgia</td>
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<tr>
<td>Transient erythema</td>
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<tr>
<td>Pharyngitis</td>
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<td>Polymorphs ≥ 80%</td>
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<td>Glycosylated ferritin ≤ 20% (Of serum ferritin)</td>
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Presence of 4 or more major criteria or 3 major + 2 minor criteria have 80.6% sensitivity and 98.5% specificity.

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<th>Table 2</th>
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<td>Major Criteria</td>
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<tr>
<td>1. Fever &gt; 39° or higher lasting 1 week or more</td>
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<td>2. Arthralgia</td>
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<td>3. Typical macular evanescent rash</td>
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<td>4. Leucocytosis (10,000/cmm) with 80% or more granulocytes</td>
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Presence of total of 5 criteria with 2 or more major criteria have 96.2% sensitivity and 92.1% specificity.

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REFERENCES

Renal Failure and Neuromuscular Weakness in Cleistanthus collinus Poisoning

Sir,

The case report of Cleistanthus collinus poisoning by Benjamin SPE et al in September, 2006 issue of JAPI was very informative.1 One of our patients, who ingested the extract of Cleistanthus collinus leaves, had renal failure and neuromuscular weakness at presentation.2 In animal models, it has been demonstrated that the leaf extract markedly inhibited muscle contractions by reducing excitability of the nerve and muscle membranes and also blocked neuromuscular transmission.3 But these are without the hypokalemic milieu seen in
actual poisoning and the concentrations of the leaf extract used may not be comparable to the actual concentration seen in human cases. Benjamin SPE et al have identified distal renal tubular acidosis (dRTA) as one of the manifestations. Hypokalemic paralysis can occur in dRTA. Hypokalemia, a well known cause of rhabdomyolysis, can cause respiratory paralysis (often irreversible), myoglobinuric renal failure and not uncommonly, death. The dRTA is probably one of the explanations for the renal failure and neuromuscular weakness seen in our patient in addition to the direct effect of the poison on the nerve and muscle membranes and neuromuscular transmission. These possibilities need to be explored further.

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REFERENCES

Reply from Author
Sir,

It is possible for distal renal tubular acidosis (dRTA) to cause hypokalemia induced rhabdomyolysis and myoglobinuric renal failure. Such a sequence of events has been reported in dRTA of diverse etiologies viz., Sjögren’s syndrome, Chinese herbal nephropathy, chronic glue sniffing, congenital dRTA and idiopathic dRTA. The combination of neuromuscular weakness, renal failure and raised total creatinine kinase (CK) levels in the setting of dRTA certainly points to the presence of rhabdomyolysis and myoglobinuria in Cleistanthus collinus poisoning. However, probably due to the high mortality associated with ARDS and shock, it has so far not been possible to identify rhabdomyolysis.

Rhabdomyolysis should be confirmed by doing serial creatinine kinase levels (will be above 10,000 U/L), analysing urine for myoglobin and by muscle biopsy. Before obtaining these data it may be too early to extrapolate animal experiments on neuromuscular transmission or nerve conduction to human cases.

Our patient had moderate elevations (883 U/L) in total CK (he did not have neuromuscular weakness or renal failure) probably implying that rhabdomyolysis and myoglobinuria were averted by correction of hypokalemia and acidosis.

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6. Subrahmanyam DK, Mooney T, Raveendran R, Zachariah B.

Erratum
Medical Philately published in October 2006 issue of JAPI on page 811 (J Assoc Physicians Ind 2006; 54 : 811) Para 3, Line 1 & 2 should be read as ‘Larry’s imagination in planning for the health of the soldiers, and practical skill in the care of the wounded in the field hospital, equaled, the genius and strategies of Napolean. Emperor utilized his special talents in planning, for several campaigns’.

Editor, JAPI