EARLY PRESENTATION OF LESS COMMON ASSOCIATION- SYSTEMIC SCLEROSIS WITH APLA

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PRESENTATION OF CASE

A 25-year-old female presented to outpatient department with complaints of pain in small joints of hand and on exposure to cold water since 2 months. Patient was nonsmoker and nonalcoholic. Patient was previously treated with analgesics with minimum improvement in symptoms. She noticed small ulcers in hand and foot since 1 month.

Examination revealed painful sclerodactyly involving all fingers solitary ulcer was noted in foot and there was restricted mouth opening, rest of systemic examination was within normal limits.

CLINICAL DIAGNOSIS

Clinical diagnosis of systemic sclerosis was made and patient was evaluated with routine and specific investigations.

Figure 1. Sclerodactyly/Raynaud’s Phenomenon

CLINICAL DISCUSSION

Raynaud’s phenomenon is one of the earliest manifestation of systemic sclerosis in patients who present early.1 It is seen that the incidence of systemic sclerosis is less in Asian group when compared to European population.2 The incidence is more common among females and is seen around 4th to 5th decade. However, certain associations are found with systemic sclerosis, which make the disease manifest early and behave more aggressively. One such association is between systemic sclerosis and Antiphospholipid Antibody (APLA).

It is seen in several studies that APLA is positive in about 10% of patients with systemic sclerosis.3 These patients have a tendency to develop ulcers in fingers early in the course of their illness as compared to APLA negative patients.4 Further higher titre of APLA correlate with the severity of pulmonary complication in systemic scleroris,5 though this is not consistent in all studies. Thus, screening for APLA for all patients is necessary is systemic sclerosis.6

Figure 2. Ulcers

DISCUSSION OF MANAGEMENT

Battery of investigations was carried out and patient was diagnosed with systemic sclerosis with APLA positive. Barium swallow was done to look for oesophageal dysmotility and was found to be normal.

Table 1. Investigations

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
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<tbody>
<tr>
<td>ANA</td>
<td>Positive for Ro-52 recombinant, Scl-70, Ribosomal-p-protein</td>
</tr>
<tr>
<td>APLA</td>
<td>IgG positive</td>
</tr>
<tr>
<td>2D-echo</td>
<td>Normal</td>
</tr>
<tr>
<td>Complement levels C3, C4</td>
<td>Normal</td>
</tr>
<tr>
<td>Pulmonary function test</td>
<td>Early small airway obstruction</td>
</tr>
<tr>
<td>Barium swallow</td>
<td>Normal</td>
</tr>
</tbody>
</table>

Rheumatologist opinion was taken and patient was started on medications, which included T. Mycophenolate Mofetil 360 mg twice a day, T. HCQ 200 mg daily, T. Aspirin 75 mg daily, T. Pentoxifylline 200 mg twice a day and T. Acitrom 1 mg daily. Patient showed significant improvement in her symptoms.

**FINAL DIAGNOSIS**

A final diagnosis of systemic sclerosis with APLA was made for this patient. Positive antiphospholipid antibody is detected in about 75% of patients with SLE and about 10-15% of patients with systemic sclerosis. A high proportion of patients showing association of systemic scleroderma and APLA suggest the presence of a morbid correlation between these pathologies. It would be useful to follow a cohort of patient affected by systemic sclerosis in order to monitor vascular complications following confirmation of the presence of antiphospholipid antibody. A strong relationship exists between APLA and systemic sclerosis. Patients positive for these antibodies are more likely to suffer from pulmonary artery hypertension, thrombosis and digital infarction.

**REFERENCES**


