Case Report

COEXISTENCE OF SYSTEMIC LUPUS ERYTHEMATOSUS AND ADDISON'S DISEASE

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ABSTRACT:

An 18 years olds girl, presented with complaints of continued low grade fever, jet black skin rash and photosensitivity, generalized hyperpigmentation, marked anorexia with extreme lethargy, dull aching abdominal pain with frequent loose stools. Painless oral ulcers, moderate loss of hair, remarkable weight loss, with joint pain, morning stiffing, colour changes of fingers on exposure to cold with digital ulceration. She was amenorrhoic during the period of illness.

She was extremely ill with moderate anaemia, raised temperature, postural hypotension, pigmented butterfly rashes on the face and discoid lesions all over the body. Marked pigmentation on the palmer creases, base of the nails, knuckles of the hands, buccal mucosa, gums, tongue, aerola of breasts and valvae, with oral ulcers were present. There were symmetrical polyarthritis with restriction of movement.

Laboratory investigations showed an elevated ESR, low haemoglobin, normal platelets, total and differential count of WBC, prothrombin time and APTT were normal, with positive ANA and anti-ds DNA antibody, normal IgG anticardiolipin antibody and a positive direct Coombs' test, with moderate proteinuria. X-ray chest and KUB regions were normal. Abdominal CT scan showed bilateral adrenal cortical atrophy. Tuberculin test was negative. There was hyponatraemia and relative hyperkalaemia. Low serum cortisol level at 8:00 am and positive short synecthine test.

Treatment started with prednisolone 1mg/kg in three divided doses which resulted in dramatic improvement, then prednisolone was gradually tappered to 7.5 mg daily. She was in remission during four years follow-up.

Keywords: Systemic Lupus Erythematosus, Addison's disease.

Pak J Med Sci January-March 2006 Vol. 22 No. 1 74-77

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* Received for Publication: May 04, 2005

Accepted: October 10, 2005

INTRODUCTION

Coincidence of primary adrenal insufficiency and systemic lupus erythematosus (SLE) is a rare occurrence.[1,2] Several pathological processes have been suggested to explain the association but variability of the reported cases suggest a multifactorial aetiology, in which tissues, cells are damaged by pathogenetic autoantibodies and immune complexes,[3] Association of anticardiolipin antibodies with thrombosis is well-established.[4,5] In clinical settings, the symptoms of Addison's disease are masked by multisystemic nature of SLE and manifestations vary according to tissues affected.[6] We are reporting a case of SLE with Addison's disease to share our experiences with other professional colleagues.

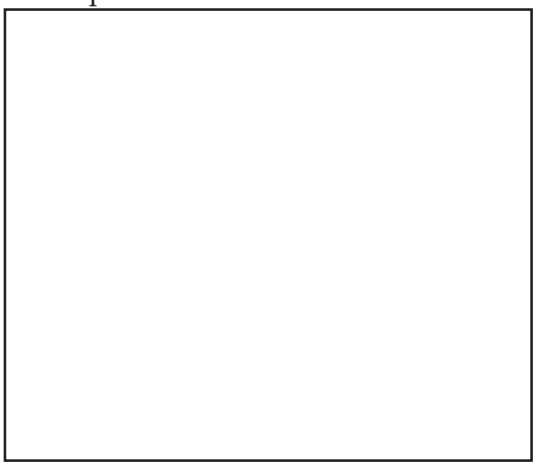
CASE REPORT

An 18 years old unmarried college girl, presented to us with complaints of continued low grade fever, skin rash and photosensitivity of 5 months duration. She noticed generalized hyperpigmentation and the rashes, which were initially pink in colour turned to jet-black areas within few days. She had marked anorexia with extreme lethargy, dull aching abdominal pain with frequent loose stools. She also noticed painless oral ulcers, moderate loss of hair along with weight loss of 12 kg within previous one month. She had joint pain, morning stiffing, colour changes of fingers on exposure to cold and had ulceration on the right index finger tip and was amenorrhoic during her illness. She took only paracetamol and amoxicillin during the period of illness from a local registered practioner.

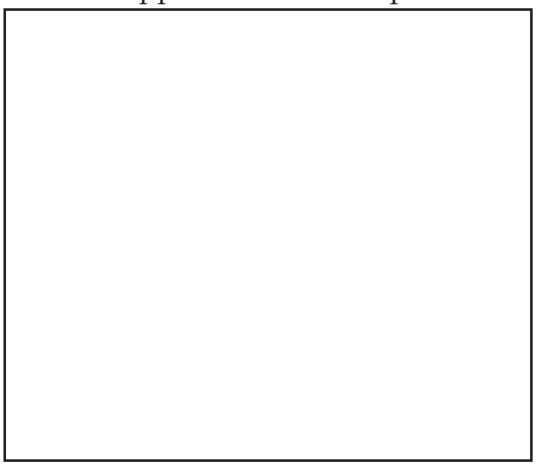
She was extremely ill with moderate anaemia, temperature was 99.6°F, postural hypotension (BP 100/70 mm Hg in supine and 80/60mm Hg on standing), the rashes had a butterfly distribution on the face and discoid lesions all over the body with central scarring and jet black hyperpigmentation. Marked pigmentation was present on the palmer creases, base of the nails, knuckles of the hands, buccal mucosa, gums, tongue, aerola of breasts and valvae. There was multiple, symmetric, large and small joint involvement with restriction of movement of ACR functional grade II. Multiple mouth ulcers were found especially over the hard palate.

Laboratory investigations showed an elevation of erythrocyte sedimentation rate (105 mm in first hour), haemoglobin (86gm\L), platelet ($240x10^9$/L), WBC ($4x10^9$/L), polymorphs ($2.8x10^9$/), lymphocytes ($1.18x10^9$/L), prothrombin time and activated partial thromboplastin time were normal, with positive ANA and anti-ds DNA antibody. Other investigations include normal level of IgG anticardiolipin antibody and a positive direct Coombs' test. Urine examination revealed moderate proteinuria (total urinary protein 2.4 gm/24 hours). Renal biopsy was not possible at that time. X-ray chest and KUB regions were normal. Abdominal CT scan showed bilateral adrenal cortical atrophy. Tuberculin test was negative. Serum electrolytes showed hyponatraemia (121mmol/L) and relative hyperkalaemia (4.1 mmol/L). Serum cortisol level measured at 8:00 am was 122 nmol/L (normal 140-550 nmol/L), short synecthin test at 8:00 am resulted in rise of cortisol level to only 138 nmol/L after one hour.

She was diagnosed as a case of SLE with Addison's disease. Treatment started with prednisolone 1mg/kg weight in three divided doses. Improvement occurred with prompt correction of postural hypotension, relief of abdominal pain and diarrhoea, healing of oral ulcers and skin rashes. There were gradual reduction of pigmentation, increase in appetite with weight gain, correction of anaemia, restoration of menstruation to normal and proteinuria disappeared. Dose of prednisolone



Before treatment, A. frontal view; B. lateral view



After treatment, C. at six months, D. at one year

was then gradually tapered to 7.5 mg daily, (5 mg at morning and 2.5 mg at night). The patient sustained in a remission during four years follow-up.

DISCUSSION

Several reports describe association between acute adrenal insufficiency (adrenocortical haemorrhage or haemorrhagic infarction) and antiphospholipid antibody syndrome.[7] Our case presented with features of SLE (fulfilling the ARC criteria) manifested by malar rash, discoid rash, photosensitivity, painless oral ulcers, arthritis, haemolytic anaemia, renal involvement and features of Addison's disease e.g., hyperpigmentation, marked postural hypotension, anorexia abdominal pain with diarrhoea extreme fatigue low serum cortisol level (122 n mol/l) which fail to rise one hour after intravenous injection to 0.25mg co-syntropin.

Table-I: Comparison of clinical features of adrenal insufficiency and SLE

	Adrenal insufficiency	*SLE*
Constitutional		
Fatigue	+	+
Malaise	+	+
Anorexia	+ +	+
Weight loss	+	+
Fever	–	+
Mucocuteneous		
Pigmentations	+ +	±
Ulceration	–	+
Gastrointestinal		
Abdominal pain	+ +	+
Nausea	+ +	+
Vomiting	+ +	+
Diarrhoea	+ +	+
Endocrine		
Amenorrhoea	+	+
Scanty axillary/ pubic hair	+	–
Neurological		
Apathy	+	+
Psychosis	–	+ +
Vascular		
Postural hypotention	+ +	–
Miscellaneous		
Anaemia	+	–
Hair loss	+	+ +

As SLE is a multi system disorder, many of the clinical features are common with adrenal insufficiency (Table -I), several features in our patient's presentation raise the suspision of adrenal insufficiency in her initial visit.

Among 20-reported cases[1] with positive anticardiolipin antibodies with adrenal failure only 4 cases developed features of SLE. According to Rao, et al.[8] the symptoms and signs of bilateral massive adrenal haemorrhage are not always dramatic in the early stases. Pain of varying severity localized to the abdomen, flank, lower chest, or back is the only consistent feature; gastrointestinal and neuropsychiatric symptoms occur much less frequently. The only reliable physical sign is fever. Laboratory finding are fall of haemoglobin level and haematocrit, hyponatraemia, hyperkalaemia and volume contraction. The aetiology of hypoadrenalism in SLE is unknown, but proposed mechanisms my be adrenal vascular thrombosis and infarction, haemorrhage due to abnormal coagulation, vasculitis and a direct organ specific autoimmune insult.[6]

In our case the cause of Addison's disease is not clear and she did not take any form of steroid preparation earlier. In some cases, of adrenal damage due to haemorrhage, incomplete destruction of adrenal cortex may leave enough residual function to prevent acute adrenal crisis, with later development of chronic adrenal insufficiency.

This case suggests the need for increased suspicion of adrenal insufficiency in patients with SLE with systemic complaints. Adrenal insufficiency is potentially a fatal disease and notoriously variable in its presentation. Clinical suspicion should be high when SLE patients present with features such a abdominal pain, fever, falling haemoglobin, electrolyte changes and marked postural hypotension.[6] To confirm hypoadrenalism, evidence of low cortisol with a short ACTH stimulation test plus anatomical evidence by CT scanning is necessary.[8]

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