Multiple/Overlap Autoimmune Syndrome: An Overlap of Systemic Lupus Erythematosus, Dermato-polymyositis and Non-insulin Dependent/Adult Onset Diabetes Mellitus Concomitant Herpes (Varicella) Zoster/Shingles, an Innovation

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Abstract

Multiple/overlap autoimmune syndrome, comprising systemic lupus erythematosus, dermato-polymyositis, and non-insulin dependent/adult onset diabetes mellitus is being reported in a 53-year-old woman. The diagnosis of each of them was based on cardinal clinical criteria supplemented by laboratory undertones. The drug delivery (management) and approaches (strategies), in particular takes cognizance of stability and acceptable improvement, but for abrupt multiple group vesicular eruption over the erythematous base accompanied by numbness, tingling and pain essentially afflicting the right forehead and the face for which acyclovir was administered in recommended dosages for herpes zoster varicella (shingles).

Introduction

Multiple autoimmune/overlap [1-5] syndromes are well-known phenomenon, and have been gathering momentum time and again. The credit of its initial perception [1,2] was visualize in early 1988 where concomitant occurrence of vitiligo, alopecia areata, Crohn’s disease, form the subject matter for the future attention, thus setting the ball rolling that one autoimmune disease made predispose to yet another [5], a conceptual hypothesis, which needs astute indulgence to diversify and enrich the literature and through current case report.

Case Report

A 53-year-old woman, weighing 54 kg, a well-established case of systemic lupus erythematous (SLE), based on the criteria for its classification [6] as well as clinical practice guidelines [7] that included fever, feeling tired, red rash, painful and swollen joints, occasional chest pain, hair loss, in particular, alopecia totalis, recurrent mouth ulcers and swollen lymph nodes were the features of systemic LE whereas, the diagnosis of polymyositis was made on the basis of dysphagia, rapid onset (<4 weeks) of myositis, cutaneous necrosis and vasculitis, conforming to the criteria laid down for dermatomyositis and polymyositis. The concomitance of systemic lupus erythematosus and dermatomyositis/polymyositis has been a fascinating overture, and now stands well-recognized as an overlap syndrome [5,8] therefore, warranted elaborate laboratory background comprising several interrelated parameters, the salient details of which are being portrayed in the following (Table 1) for ready reference. In addition, the diagnosis of polymyositis was undertaken on the muscle biopsy of affected gastrocnemius muscle of the thigh, which depicted marked in fibre size variation along with fat infiltration, and minimal perimysial fibrosis. There was no evidence of peri-fascicular atrophy or significant inflammation. Fibre typing and myofibrillar architecture was maintained. Immuno-histochemistry of spectrum, alpha (α), beta (β), gamma (γ), delta (δ), sarcoglycan, dysferelin. Lamerin and emerin showed reservation in muscle fibres. Human leukocyte antigen (HLA) 1 and 2 were not overexpressed, and were within normal limits, features suggestive of that of a myopathy.

| Test name                              | Results | Units      | Bio. Ref. interval |
|----------------------------------------|---------|------------|-------------------|
| Complete Haemogram Test or Complete Blood Count (CBC) |         |            |                   |
| Haemoglobin (Hb)                       | 11.6    | g/dl       | 12.5-16.0 g/dl    |
| Total Leucocyte Count (TLC)            | 6900    | /cumm      | 4000-11000/cumm   |
| Differential Leucocytic Count (D.L.C)  |         |            |                   |
| Neutrophil                             | 73      | %          | 40-75             |
| Lymphocyte                             | 18      | %          | 20-40             |
Table 1: Laboratory investigation; depicting name of the test; Results; units and Bio. Reference interval.

| Test                                      | Result       | Reference Interval         |
|-------------------------------------------|--------------|----------------------------|
| Eosinophil                                | 5%           | 6-Jan                      |
| Monocyte                                  | 4%           | 10-Feb                     |
| Erythrocyte Sedimentation Rate (E.S.R) (westergren) | 18 mm/hr     | 20-Jan                     |
| Packed cell volume (P.C.V)                | 36.7%        | 35.0-6.50                  |
| Red Blood Cells (RBCs)                    | 4.26 Million/mmcm | 3.50-5.50                |
| Platelet Count                            | 3.08 Lakh/mmcm | 1.50-4.50                |
| Mean Corpuscular Vol (MCV)                | 86.2 fl      | 80.0-99.0                  |
| Mean Corpuscular Haemoglobin (MCH)        | 27.2 Picogram | 27.0-31.0                  |
| Mean Corpuscular Haemoglobin Concentration (MCHC) | 31.6 gm/dl   | 33.0-37.0                  |
| Red Cell Distribution Width (R.D.W)       | 15.6%        | 11.5-14.5                  |

Drug delivery (management) and approaches (strategies)

Consequent upon aforementioned revelations, treatment comprising 50 mg of prednisolone (Orapred, Prelone) once a day after breakfast adequate to stabilize the condition, supplemented by oral administration of pantoprazole (Protonix, pantocid) 40 mg, telmisartran (Twynsta, telma) 20 mg, calcium carbonate (Bio max 3D) 500 mg, methylcobalamin 1500 MCG, calcitrol 0.25 mcg for a period of 90 days. During the period a perceptible overall amelioration was a benchmark for tapering a dose of prednisolone 9-10 mg by every 30 days until a maintenance dose of 10 mg azathioprine 10 (Imuran, Azasan) to 50 mg 2 times a day, an adjunct therapy was added as a replacement therapy. Incidentally, the patient had high fluctuating blood sugar levels, which were detected over a period of time, apparent in the form of frequent urination, increased thirst, and increased hunger, non-insulin dependent/adult onset diabetes mellitus [9-12] type-2. She is also been treated by human mixtard 30/70 100 iu injection 35 unit before the major meals in the morning and 35 units in the evening. During the course of the treatment the patient was taken aback by sudden appearance of blisters affecting the right side of the face and forehead, and reported to the outpatients' with the complaints of excruciating pain, redness, swelling, and blistering affecting exclusively the right side of the face of 8 days duration. Initially, she had experienced numbness, tingling, and pain, which during the 48 h usher in with preceding features [13].

On skin surface examination, facial appearances were conspicuous, and studded with vesicles and/or blisters largely in groups/clusters. They were numerous. A few discreet vesicles or blisters intersperse between the groups were also seen. The background of these lesions was erythematous, there was marked edema of the face. Essentially, the lesions were ipsilateral altering right half of the face only (Figure 1). Accordingly, acyclovir [14] (Zovirax, Sitavig) 800 mg was administered 5 times daily for a period of 10 days, with complete regression of lesions leaving behind pigmented macules.

Figure 1: Herpes (varicella) zoster /shingles in Systemic lupus erythematosusmyositis overlap syndrome and Adult-Onset Diabetes Mellitus: Herpes zoster displaying group vesiculo bullous lesion on the erythematous background, occupying the right forehead and the cheek with prominent face edema, alopecia totalis wearing a wig.

Discussion

Multiple [2,3,5] autoimmune syndrome, is a fascinating caption has been in vogue ever since its inception -1, in the year 1988 and exclusive account of which was emphasize later as Multiple Autoimmune Syndrome (MAS). The scope of this term can be enlarge to include one or several autoimmune disease for one autoimmune disease may predispose itself to yet another autoimmune disease(s), inferring their overlap, an Overlap Autoimmune Syndrome (OLAS) thus Multiple Autoimmune Syndrome (MAS) and Overlap Autoimmune Syndrome are interchangeable and could be conveniently make use of in an exigency.

However, only a few scintillating report could be identified, despite it being an astounding challenge not only for its clinical, but also far
laboratory collaterals, considered essential to arrive at their precise diagnosis. It is true for both SLE as well as dermato-polymyositis.

The two well recognized autoimmune diseases the challenging pillars, inviting strenuous endeavors both for diagnosis and therapy. Non-Insulin dependent type-2/Adult Onset Diabetes Mellitus may yet be another component of multiple/overlap autoimmune syndrome, adding new dimensions Furthermore, it is therefore worthwhile to form an overview of three or more autoimmune diseases available thus far, with a focus, in particular, based on the analysis of 87 cases till 1988 from across the globe-1 suggesting a comprehensive classification for the future use. The recitation of which may provide glimpses into the future guidelines.

**Type-I** comprises myasthenia, thymoma, polymyositis and giant cell myocarditis.

**Type-II** includes the Sjögren’s syndrome, rheumatoid arthritis, primary biliary cirrhosis, scleroderma and autoimmune thyroid disorders.

**Type-III** groups together 10 autoimmune diseases (autoimmune thyroid disease, myasthenia and/or thymoma, Sjögren’s syndrome, pernicious anaemia, idiopathic thrombocytopenic purpura, Addison’s disease, insulin-dependent diabetes, vitiligo, autoimmune haemolytic anaemia, systemic lupus erythematosus).

An elaborate search on multiple/overlap autoimmune syndrome was found to be absolutely depleted, except for an occasional report-2 in the year 2014. This particular case report therefore, is paramount and had vitiligo, alopecia areata, crohn’s disease, psoriasis vulgaris and oral lichen planus to focus attention to the subject in order to create awareness for picking and choosing such cases in the future. Accordingly, reports of permutation and combination of multiple autoimmune diseases corresponding to type-1 [15], type-2 [16] and type-3 [17,18] were identified and delineated. The minute details of drug delivery (management) and approaches (strategies) challenging perspective for need careful designing to achieve optimum amelioration in the condition. Herpes (varicella) zoster/shingles may be one of them and has rarely been reported in SLE [19] and polymyositis [20] which may either be due to iatrogenic/coincidental or immunocompromised [21].

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