

A Peculiar Juvenile Progressive Systemic Sclerosis Case Report: The Egyptian versus the International Approach of Treatment

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Abstract

Progressive systemic sclerosis (PSS) is an autoimmune generalized connective tissue disease that affects the skin as well as internal organs. I report a 4 years old Egyptian female patient who is suffering from PSS for more than one and a half years. A peculiar clinical picture has deceived some clinicians at an early stage and a quite rapid progression has dramatically deteriorated the condition. A synopsis of the drugs which are being used in her treatment in Egypt compared to the international approach is also demonstrated in this case.

Keywords: Juvenile systemic sclerosis; Methotrexate; Prednisone

Case Report

Juvenile progressive systemic sclerosis (PSS) is a low prevalence autoimmune multisystem connective tissue disease of obscure etiology which is characterized by skin fibrosis and internal organ involvement [1,2]. A small well-circumscribed whitish patch has appeared on the lower right forearm of an Egyptian female child born to a father and mother who are cousins, she was two and a half years old - now she's 4 years old - when her parents first consulted some dermatologists, an initial misdiagnosis of vitiligo was made and topical corticosteroids and narrow band UV-B sessions were prescribed and followed for three months. Rapidly within these few months, the small patch has extended to involve most of the skin of the right upper limb, the elasticity of the skin was gradually lost and a contracture of the right hand has developed later (Figure 1). ESR performed according to the Westergren method was higher than normal (56 for the first hour and 74 for the second); ANA titer by IF was positive and negative anti Scl 70 as well as negative anti-double-stranded DNA titers by ELISA were also recorded. A skin biopsy confirmed a diagnosis of localized scleroderma (morphea) and only then a pediatrician prescribed a regimen of oral prednisone combined with a weekly low dose systemic methotrexate (MTX). Unfortunately, the condition has not improved and over nine months following this regimen, large patches have involved lower limbs (Figure 1), trunk and lower neck with marked anorexia, dysphagia, constipation and dyspnea. The parents have turned to ask the advice of a rheumatologist who prescribed a regimen of low dose systematic MTX combined with oral azathioprine, the condition has relatively been stabilized; however, no improvement of the systemic symptoms was felt. Moreover, cicatricial alopecia has developed over the last six months following the new regimen (Figure 2). Interestingly, this condition occurred for the first time in the family; the left upper limb is almost completely normal and the parents couldn't confirm the occurrence of any symptoms of Raynaud's syndrome. I've asked for echocardiography, chest x-ray and recommended to periodically check for proteinuria.



Figure 1: Diffuse fibrotic skin patches involving the right upper limb with contracture of the right hand as well as both the lower limbs.



Figure 2: Recent cicatricial alopecia spreading over the scalp

Juvenile progressive systemic sclerosis is a rare chronic childhood condition characterized by fibrosis of the skin and widespread

abnormalities of the viscera. Cardiopulmonary disease is the most frequent visceral involvement, leading to significant morbidity and progressive cardiopulmonary complications are known to be the main causes of death in these patients [2-4]. Further, patients with PSS often suffer from gastrointestinal tract disorders that may progress to significant dysmotility causing complications like oropharyngeal dysphagia, esophageal dysphagia, gastroesophageal reflux, gastroparesis, pseudo-obstruction, bacterial overgrowth, intestinal malabsorption, constipation, diarrhea, fecal incontinence, malabsorption, weight loss and severe malnutrition [5,6].

Additionally, renal disease is also considered an important cause of morbidity and mortality in PSS. The spectrum of renal complications includes scleroderma renal crisis, normotensive renal crisis, antineutrophil cytoplasmic antibodies-associated glomerulonephritis, penicillamine-associated renal disease, and reduced renal functional reserves manifested by proteinuria, microalbuminuria, or isolated reduction in glomerular filtration rate [7]. As regard to treatment, it's well established that long-term MTX therapy is considered a beneficial and well tolerated treatment for juvenile localized scleroderma (JLS) and that mycophenolate mofetil is considered to be effective in arresting disease progression in severe or MTX-refractory JLS and is also generally well tolerated [8,9]. Moreover, pulsed high-dose corticosteroids combined with orally administered low-dose MTX therapy were previously shown to be beneficial and safe in the treatment of patients with severe LS [10].

Recently, a combination of imatinib, corticosteroids, and MTX halted the progressive skin thickening and the hand and finger joint deformity in the early stages of a case suffering from progressive JLS [11] and a combination of infliximab and leflunomide have shown a similar or even more success in a case of an adolescent Caucasian female with severe progressive localized scleroderma [12]. Azathioprine and cyclosporine A were also tried with limited success to control a pediatric case suffering from a severe progressive chronic skin disease with clinical features of PSS, systemic lupus erythematosus and dermatomyositis [13].

Regarding the case represented in this paper, I believe that the regimens used in the management were highly subjective and in my opinion, the lack of solid protocol to manage the rare cases of Juvenile PSS is one of the main reasons for such a tragedy frequently encountered in the developing countries. However, socioeconomic reasons are also hindering trials of some of the new drugs like

infliximab or of course imatinib to be offered to this child belonging to a poor family.

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