Case Report

Juvenile Systemic Sclerosis Management in Remote Area

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Background: Juvenile systemic sclerosis is a rare autoimmune disorder, often leads to delay in diagnosis. In limited resource setting, management could be challenging due to lack of treatment choices.

Aim: To describe the debilitating impact of juvenile systemic sclerosis in children and management in low resource setting.

Method: Data was collected from personal document, medical record and history taking.

Case Description: We report a case of systemic sclerosis in an eleven-year-old girl from remote island. Her fingers became stiff at first before spreading to upper arms and thighs in the following six months. History taking revealed dark-reddish skin lesion appeared three months before affected areas became stiff as board. She had already managed by Dermatologist as Morphea but was referred because her painful knees led to suspicion of systemic involvement. We found dark-shiny skin lesions, limited range of movement in four extremities, swollen and inflamed knees, and painless finger ulcers. Skin biopsy showed thick collagen deposition. EMG study revealed neuropathy. Indirect immunofluorescence ANA test showed speckled pattern with titer more than 1:1000. We established diagnosis of Juvenile Systemic Sclerosis (PRES/ACR/EULAR), and the clinical features suggested limited cutaneous type. Due to limitation in medication choices, we continued the corticosteroid and added joint mobility, muscle conditioning exercise, also occupational therapy.

Conclusion: Juvenile Systemic Sclerosis impacts beyond health problem. Holistic approach using available modalities should be considered to rehabilitate the child, especially in low resource area.

Keywords: juvenile, systemic sclerosis, systemic scleroderma, management, therapeutic

INTRODUCTION

Chronic disease impacts children heavily not only from physical growth and development, but also from social, educational, and psychological aspects. Juvenile systemic sclerosis is a rare chronic-autoimmune rheumatic disease, often leads to delay in diagnosis. In limited setting, management could be challenging due to lack of treatment choices. This case illustrates the rheumatic disease in children could be excruciating and management strategy should be comprehensive, considering available modalities.

CASE REPORT

An eleven-year-old girl came to our allergy-immunology clinic with the chief complaint hardening of her upper and lower extremities. Her fingers became stiff at first (Figure 1) before spreading to arms and thighs in the following six months. This condition affected her routine because she had limited walking ability, squatting when she needed to use bathroom would initiate pain, and disturbed small joint utilization limited her writing ability. She felt ashamed with her difficulties that she refused go to school or play with her peers.

Further history taking revealed that discolored skin lesions (Figure 2) appeared about three months before affected areas became stiff as board. She had already managed by Dermatologist diagnosed as Morphea but was referred to us because her painful knees led to suspicion of systemic involvement.

From physical examination we found dark-shiny skin lesions on her neck (Figure 3) and extremities those hard on palpation. Her range of movement of four distal limbs was limited, both knees felt warmth and swollen, and there were painless ulcers at the top of her fingers. Punch biopsy of the skin (Figure 4) showed epidermal atrophy and thick collagen deposition corresponded with scleroderma. EMG study revealed neuropathy of Peroneal and Tibial Nerve. Result of echocardiography examination was mild tricuspid regurgitation and trivial mitral regurgitation with normal left and right ventricle function. Indirect immunofluorescence ANA test showed speckled pattern with titer more than 1:1000. Other examinations were unremarkable.

Fig. 1
Sclerodactyly

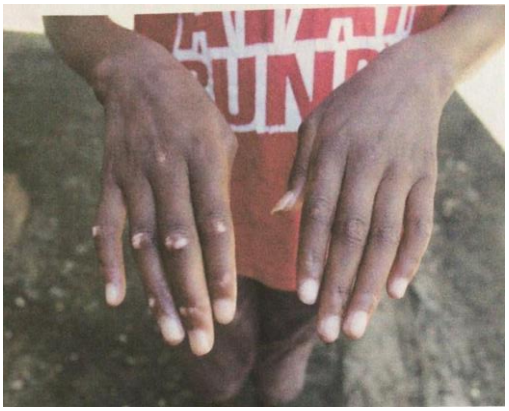

Fig. 2
Discolored skin lesion

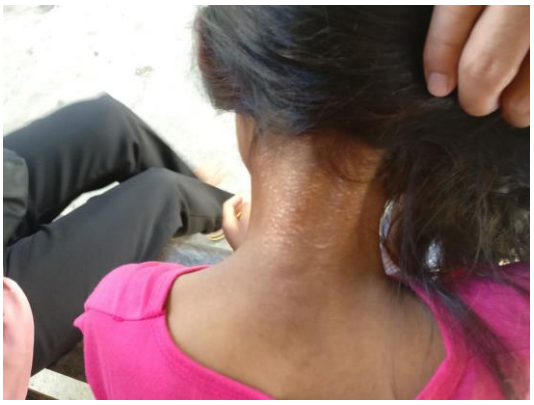

Fig. 3
Morphea

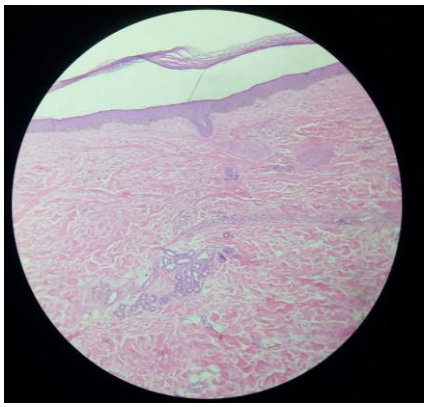

Fig. 4
Punch Biopsy

From the findings, using classification criteria from Rheumatology European Society (PRES), the American College of Rheumatology Pediatric (ACR), and the European League Against Rheumatism (EULAR), we established diagnosis of Juvenile Systemic Sclerosis, and the clinical features suggested limited cutaneous type.[1]

Considering natural course of the disease that is mild nor progressive,[2] availability of medication from where she lived and potential for long-term use also side effect monitoring we decided to continue corticosteroid that she got from Dermatologist before. Low dose methylprednisolone was given at 1 mg/kg body weight then tapered off once clinical manifestation alleviated. Along with medication, we suggested more fluid intake for connective tissue hydration, added excercise for joint mobility, muscle conditioning and occupational therapy to help her cope with the symptoms. We also assured caregivers to adhere with the treatment.

DISCUSSION

This case reports an eleven-year-old girl with diagnosis juvenile systemic sclerosis (SSc). This case is quite rare, there are no exact epidemiological data regarding incidence and prevalence of juvenile systemic sclerosis. All available data consisting of organ involvement and outcome available today are based on case reports and retrospective studies. It is suspected 3-10% of all systemic sclerosis patient, developed the disease before the age of 18 years, and estimated the mean age of onset is 8,8 years.[3] Skeletal muscle involvement occurred more than twice as often in the childhood onset compared to adult onset. Survival in childhood onset systemic sclerosis was better than in older adult onset. In our case**,** patient was female corresponds with the worldwide incidence. Female is more common to have this condition compared to male with the ratio 2-4:1. Course of the disease started at nine years old fits the epidemiological data found that stated the mean age at disease onset was 8.8 (±3.3) years. In two years of observation, the disease was no longer active, the skin and skeletal muscle symptoms stabilize, and patient was still on medication. There was no heart, renal, lungs, and central nervous system involvement during observation.

Progressive thickening and fibrosis of skin secondary to excessive collagen accumulation is the most evident and universal finding and can be associated with varying degrees of fibrosis of internal organs. Skin biopsies from SSc patients, compared with skin from healthy controls, show a significant increase in myofibroblasts, especially within the deeper dermis. Vascular dysfunction and abnormalities are often seen and can precede organ involvement by several years. Vasculopathy consists of fibro intimal proliferation of small vessels and vasospastic episodes triggered by cold or stress. This condition clinically referred to as Raynaud's phenomenon, usually was the earliest manifestation of SSc, could lead to tissue ischemia. In our case, the main complaint of the patient was caused by thickening and fibrosis of skin. These were a distinctive feature and universal finding found in scleroderma. She also had fingertip painless ulcers as one of the symptoms those appeared at first. The ulcer was the consequence of skin fibrosis and vascular dysfunction. The patient underwent skin biopsy that showed epidermis and dermis layer. There was atrophy at epidermal layer and thick collagen deposition with parallel distribution at dermal layer. This morphology was strongly suggested scleroderma. Unfortunately, the tissue specimen did not visualize blood vessel.

There are many treatments have been tried for limited cutaneous systemic sclerosis over the years. These have

included topical, intralesional and systemic corticosteroids, topical and systemic calcipotriol, topical tacrolimus, hydroxychloroquine, sulfasalazine, penicillamine, gamma-interferon and methotrexate, and phototherapy with UVA light, with and without psoralens. Unfortunately, the rarity of this disease and the difficulty in assessing outcomes in an objective way have limited the interpretation of most of these studies. There is one study addressing usefulness of oral corticosteroid in treating systemic sclerosis. The study reported that oral prednisolone could be beneficial in controlling skin sclerosis symptom in systemic sclerosis with no incidence of renal crisis.[4] In our case, the patient received oral methylprednisolone at 1 mg/kg body weight then tapered off as the skin sclerosis improved. Patient consumed the oral corticosteroid for as long as two years and stopped after the disease was inactive. There is no major adverse event developed by the end of observation.

Because there is no cure, comprehensive care involving combination of both pharmacologic and non-pharmacologic intervention may provide adequate milieu for the patient with systemic sclerosis to recover. Non-pharmacologic treatment including psychological, educational, and rehabilitation interventions provided to attenuate disability and support patient in coping with the disease. Multiple rehabilitation techniques including range of motion exercises, connective tissue massages, joint manipulation, splinting, heat/paraffin wax baths, and generalized physical therapy have been suggested to improve pain and joint motion. Non-pharmacological therapy with multidisciplinary team was proven to be effective in rehabilitate the patient with systemic sclerosis to execute daily living activities.[5] In our case, beside pharmacotherapy, the patient underwent physiotherapy excercise with multidisciplinary team. The program includes joint mobility and muscle conditioning exercise, also occupational therapy. We also provide motivational support and encouraging the caregiver to adhere with the treatment.

CONCLUSION

Juvenile Systemic Sclerosis in children impacts beyond health problem. Management of disease should consider holistic approach focusing on rehabilitating the child back to daily activities and mental status. In low resource setting, medication option needs to consider continuity of care accompanied by supportive environment to assure efficacy.

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