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## Juvenile Dermatomyositis Associated with Acute Lymphocytic Leukemia

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### Introduction

A link between dermatomyositis and neoplasia has long been suspected. The precise incidence of this association remains controversial, owing to varying criteria used to diagnose dermatomyositis. Although juvenile dermatomyositis is believed to display no association with neoplasm, there have been few reported cases of dermatomyositis associated with tumors of any kind in pediatric age group (1-4). We describe another such case and review briefly the current status of the association of neoplasia with juvenile dermatomyositis.

This female child was normal until the age of 15 months when she began to have fever and swelling of extremities. When she was 21 months old she was brought to Pediatrics Clinic of University Hospital with the complaining of fever, flashing on the face, pain and swelling of the arms and legs and difficulty in walking. She was born to 17-year-old mother and parents were first cousins. Physical examination revealed nonpitting edema on dorsal side of the extremities, patchy erythematous and nonitching rash in the skin, periorbital purplish discoloration associated with mild edema (heliotrope), proximal muscle weakness and hypertension (150/80 mmHg). Liver was palpable 2 cm below the costal margin. Laboratory investigations showed Hb, 9.7 gr/dl; Hct, 31%; ESR,

68mm/h; CPK, 97 IU/L; LDH, 913 IU/LP; AST, 65 IU/L; ALT, 85 IU/L. IgM were increased, 1720 mg/dl and 261 mg/dl respectively. Rest of the biochemical tests were normal. EMG revealed small, short duration polyphasic motor unit potentials. Muscle biopsy showed perifascicular muscle fiber atrophy and scattered muscle fiber necrosis and regeneration, and mononuclear cell infiltration of muscle fibers. Immunocytochemical studies demonstrated endomysial IgG deposition. Skin biopsy from sclerotic area showed increased collagen fibers and atrophic sweat glands. Renal biopsy was normal. On the basis of the presence of proximal muscle weakness, chronic inflammatory changes in skin and muscle biopsy and characteristic EMG findings the diagnosis of dermatomyositis was made.

Hypertension was treated with antihypertensive drugs. Prednisolone therapy (2 mg/kg) was instituted to treat active dermatomyositis. ESR was gradually decreased from 68 mm/h to 20 mm/h. Prednisolone was reduced to 5 mg/d within one month. After the following-up as out-patient for five months, she was hospitalized again because of the gradually developing subcutaneous solid nodules and plaques. She had difficulties in walking and sitting. Physical examination revealed marked proximal muscle weakness and hepatosplenomegaly. Laboratory studies showed ESR, 50 mm/h; LDH, 250 IU/L; CPK, 250 IU/L. Rest of the laboratory results were unremarkable. Plain radiographs

of the extremities showed widespread subcutaneous calcification (Figure 1). With these clinical and laboratory findings, she was diagnosed to have universal calcinosis as a complication of juvenile dermatomyositis. Since high level of CPK and increased ESR were accepted as an activation criteria, full dose of prednisolone (2 mg/kg) was given again for two more months and continued with the dose of 10 mg/d. At the end of the six-month-follow-up period, she showed remarkable improvement clinically and steroid treatment was stopped. Calcified nodules and plaques were nearly disappeared at that time. Within the following six months, at the age of three years and eight months, all subcutaneous calcified deposits disappeared radiographically.

The child was symptom-free until the age of five when she began to show paleness and weakness in addition to masses in neck and abdominal enlargement. Physical examination revealed axillar, cervical and inguinal multiple lymphadenopathy, and hepatosplenomegaly. No skin lesion other than localized scar tissues all over the body was observed. La-



Figure 1. Plain radiograph showing widespread subcutaneous calcifications.

boratory investigations showed Hb, 6.8 gr/dl; Hct, 20% WBC, 32000/mm<sup>3</sup> with over 90% blasts; ESR, 72 mm/h; platelets, 72000/mm<sup>3</sup>; ALT, 24 IU/l; AST, 14 IU/L; CPK, 60 IU/L; LDH, 746-2406 IU/L (in multiple measurements). Bone marrow examination revealed 95% lymphoblasts (33% L<sub>2</sub>, 67% L<sub>1</sub> type blasts; according to FAB classification) with PAS (-), Sudan black (-), peroxidase (-). Flowcytometric examination showed CD19(+), CD20(+), CD22(-), CD10(+) and HLA DR(+). Rest of the laboratory tests were normal and final diagnosis was pre B cell acute lymphoblastic leukemia and ALL BFM 90 therapy protocol was begun.

Juvenile dermatomyositis closely resembles its adult counterpart. Important differences distinguish juvenile dermatomyositis from the adult-onset disease, however, apart from the age of the patient (1). Calcinosis is much more likely to occur in children than adults as a complication and chronic sequel even after the active disease process is arrested. Angiitis of muscle is generally more intense in children, and vasculitis of other organs is more common than in adults (5). Finally, it is rarely associated with malignancy in childhood except for a few reports (1-4).

Although most of them are retrospective, recent studies have shown an increased incidence of malignancy among adult patients with dermatomyositis when compared with controls without myositis (6,7). The types of neoplasms found in association with dermatomyositis seem to parallel those observed in general population (7,8). Malignancies with hematopoietic origin have also been described, such as lymphoma (7,9), and leukemia (10). The relationship in time between the onset of dermatomyositis and the discovery of malignancy is quite variable. The preceding neoplasm is not unusual (30%), although it is somewhat more common for the dermatomyositis to be diagnosed before the tumor (70%); in either sequence, the two processes generally are discovered within 2-3 years of each other, though longer gaps have been noted (11). This time interval in our case was about three years after the initial diagnosis of dermatomyositis. It may be suggested that diagnosis of dermatomyositis and malignancy separated widely in time are probably coincidental findings. Although only 16-20% of the patients with dermatomyositis have onset childhood (5), namely juvenile dermatomyositis has been found in the literature, except a few case reports (1-4).

In sum, we believe our case is a rare example of

dermatomyositis associated with leukemia. Although the concurrens of these two events could still be a chance association, Such reports will rise, from time to time the question whether there is linked between

juvenile dermatomyositis and malignancy. This will prompt us to perform larger and collaborative studies to determine the exact relationship of these two events.

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