Dermatomyositis *

DERMATOMYOZİS

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SUMMARY

A rare case of a 9-year-old girl diagnosed as dermalomyosilis with a characteristic facial erythema, periorbital oedema, "heliolropic eyes" sign atrophic skin findings similar to psoriasis on the extremities, proximal muscle weakness is presented. Muscle biopsy showed inflamatory changes suggestive of connective tissue disorder.

TJte patient was treated with Prednisolone 2 mg/kg daily and the skin lesions were beared on the day of len-and the dose of Prednisolone was gradually decreased. During 6-months follow-up the patient was healthy without any recurrence.

Key Words: Juvenile dermatomyositis, Polymyositis, CPK

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ÖZET

*Yüzde kelebek tarzında rash, periorbital ödem, gözlerinin etrafından heliotropik renk değişikliği, ekslremiteleri üzerinde psöriasise benzer atrofik deri bulguları ve proksimal adalelerde kuvvetsizlik mevcut olan dennatomyozis tanısı konulan **9** yaşındaki bir hasta sunuldu. Hastanın serum kreatinin fosfokinaz seviyesi yüksek bulundu. Yapılan kas biyopsisinde infamatuvar myozilis bulguları mevcuttu. Hastaya 2 mg/kg/gün dozda Prednizolon tedavisi başlandı ve tedavinin onuncu günü deri lezyonlan iyileşmeye başladı ve kullanılan doz giderek azaltıldı. Altı ay takip edilen hastada rekürrens görülmedi.*

Anahtar Kelimeler: Jüvenil dermatomyozis, Polimyozit, CPK

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Juvenile Dermatomyositis (JDMS) is a rare connective tissue disorder in children. It is one of the inflammatory myopathies and characterized by vasculitis with skin and muscle involvement (7). The diagnosis is mainly based on theclinical features of symmetrical proximal muscle weakness and typical cutaneous manifestations, elevation of muscle enzymes, and/or abnormal eleetromyogram (EMG) and/or muscle biopsy showing inflammatory myositis (1,5,7).

This paper presents a 9-year-old child with JDMS with typical clinical and suggestive laboratory findings.

Case Report

S.Y., a 9-year old girl was referred to Karadeniz Technical University Medical Faculty Hospital for evaluation of easy fatigability, puffiness around the eyelids and facial redness for 10 days' duration. The personal history was negative. The parents and 3 other siblings were alive and healthy.

On physical examination the blood pressure was 100/60 mmHg., pulse rat 82/min. She had a

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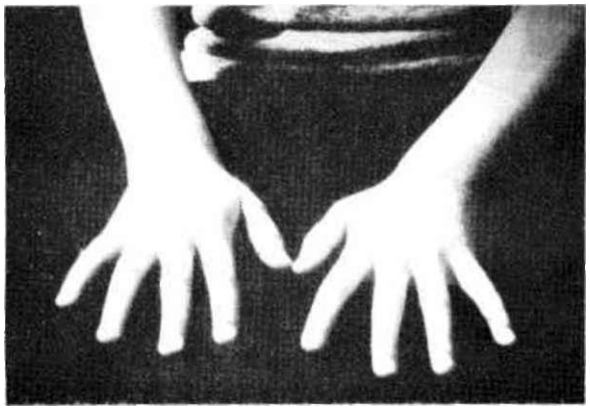
§ekil 1, Dermalomyositis. **Ilcliolropic** eyes of the patient and a malar rash crossing the nasal bridge.

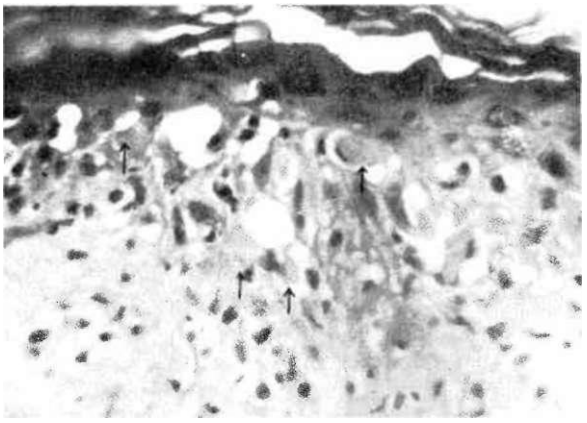


Şekil 3. Dermatomyositis. The skin over the exlremites was hypertrophied with **a** pale red colour, evolving into pale colourless bands of atrophic skin (Gottron's sign).

§ckil 2. Dermalomyosiiis. V-shaped erythema of the upper tarse, the extensor and flexor surfaces of the arms ad the extensor surfaces of the legs, over the elbows and knees.

malar rash crossing the nasal bridge and severe and erythema of the eyelids (Heliotrope) (Fig 1). Other areas of skin involvement included a V-shaped erythema of the extensor surfaces of the legs, over the elbows, knees, (Fig 2). The skin over the extremities was hypcrtrophied with a pale red colourless bands of atrophic skin (Gottron's sign) (Fig 3). The patient was in difficulty in climbing stairs and walking on toes. Soft palate dysfunction was not present and the speech was normal. The results of the rest of the physical and neurologic examination were normal.

The haemoglobin, cryhrocyte sedimentation rate (ESR) and white blood cell counts total protein, albumin, scrum urea, creatinine, sodium, potassium, chlorur, sugar and uric acid were within normal limits. The other test results are as follows: Direct Coombs' test was negative, ASO 100 Todd units, CRP, Latex, LE cell, VRDL, antinuclear antibody (ANA), anti-dcoxyribonuclease (anti-DNA) were also negative. Serum lactic dehydrogenase (EDM) level was 85 units/L (normal range 95-200 units/L). SGOT 80 units and SGPT was 44 units. Serum creatinine Phosphokinase (CPK) was found to be elevated. 484 units/L, more than four fold above the normal value (normal range 35-114 units/L). A skin biopsy carried out from the lesions showed the histological findings suggestive of connective tissue disorder (Fig. 4). Flattening of the epidermis hydropic degeneration of the basal layer, oedema of the upper dermis, lmyphocytic infiltration and fibrosis around the capillaries and skin appendages were seen. PAS-positivc fibrinoid deposits (colloid bodies, Civattc bodies) at the dermal-epidermal junction and around the capillaries of the upper dermis were also observed. The patient



Sjekil **4.** Dermatomyositis. The epidermis is atrophic and devoid of rate ridges. There is hydropic degeneration of basal cells. Colloid bodies, cferred to also as Civatte bodies, are present in the lower epidermis and in the pupillary dermis (arrows). (B-89-87) (II+1: .\600).

had no any muscle biopsy. Prednisolone was started in a dose of 2 mg/kg/day for treatment.

On the 10th day of treatment the skin lesions started to heal and the dose of prednisolone was decreased gradually. The duration of treatment was totally 30 days. On 6-monlhs follow-up period we did not record any recurrence.

DISCUSSION

Juvenile dcrmalomyositis is one the inflammatory myopathics which is thought to be pail of the spectrum including polymyositis (PM). Pfvf occurs more frequently in adults and is not associated with cutaneous involvement (1,5). In other words, dcrmalomyositis is diagnosed when polymyositis is present in association with a characteristic set of inflammatory skin changes. In our case we mainly thought the probable diagnosis of dcrmalomyositis because of the severe skin findings. Occasionally other autoimmune diseases may show cutaneous manifestations similar to JDMS-such as SLE. Although our case had a malar rash similar to "butterfly" type the test results were negative related to SLE and she had a typical periorbital oedema which is unusual for SLE. Gollron's sign can be mimicked by psoriatic lesions, which may be accompanied by healing foci of hypopigmenlation-found in locations usually unaffected by JDMS. We suggest, in addition to skin biopsy findings, the negative history about the healing of the skin lesions with sunlight exposure would exclude psoriasis in our case.

Although we did not have E M G in our case, we would think that the presence of proximal muscle weakness clinically constitutes a proof for the diagnosis of JDMS. Furthermore, E M G findings are not specific for JDMS and the similar findings may occur also in the muscular dystrophies and in early acute inflammatory myopathy (7).

Laboratory tests arc of limited value in the diagnosis of childhood dcrmalomyositis. Of the serum muscle enzymes the most sensitive is creatine phosphokinase (CPK). This was found to be elevated about four fold above the normal value in our case. Although elevated CPK level may be the most sensitive marker of myositis it is still a non-specific criterion for the diagnosis of dcrmalomyositis in children. The normal enzyme level can be explained by the fact that the patients were already so wasted that even in an exacerbation of the disease the CPK did not rise.

Colloid bodies, referred to also as "Civatte bodies" arc round to ovoid, eosinphilic, homogeneous, PAS positive structures which are approximately 10 mm in diameter (3). They form as the result of dyskeratosis of individual epidermal cells. Colloid bodies are not specific for the disease. They may be seen in several diseases with damage to the basal cells, such as lupus erythematosis, poikiloderma and dermatomyositis (6). These fibrinoid bodies were located in the lower epidermis and in papillary dermis in the skin biopsy of our case.

Muscle biopsy is not essential for the diagnosis and the normal muscle histology does not exclude the clinical diagnosis of JDMS. Ten to 40 per cent of biopsies have been reported as normal even in patients with clearcut active disease, presumably due to sampling error (2,9).

Although muscyc biopsy or E M G was not certainly necessary for the diagnosis of JMDS, both of these tests were still accepted being helpful for the diagnosis (10).

The prognosis of the patients with JMDS has been improved dramatically with aggressive steroid therapy (prednisolone 1 to 2 mg/kg/day) commenced early in the course of the acute disease (4,12). Immunosuppressive agents and plasmapheresis have been tried in severely ill children (8). In our case high dose steroid was used and in ten days the skin lesions started to be healed. Fol-

lowing the 30 days treatment period with prednisolon the patient started to feel better clinically and normal CPK level was also. The duration of therapy is variable in the literature (4,7) but in an uncomplicated case, the steroids rctapcrcd slowly over a 1-ycar interval. Periodic clinical and laboratory evaluation is required to monitor the possible development of other rheumatic disease such as Sjogren's syndrome, mixed connective tissue disease, overlap syndrome, rheumatoid arthritis, or scleroderma (11).

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