Case Report

# A Child with Dermatomyositis and a Suspicious Lymphadenopathy

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***ABSTRACT.*** Dermatomyositis (DM) in adults is frequently associated with cancer. In contrast, juvenile DM (JDM) is predominantly idiopathic and rarely reported with occult neoplasm. We describe a patient who presented with DM that was found to be a paraneoplastic manifestation of nasopharyngeal carcinoma. Although rare, paraneoplastic JDM must be suspected in the presence of unusual features such as elevated inflammatory markers or lymphadenopathy. Accurate clinical assessment including appropriate biopsies is needed before starting glucocorticoid therapy. (J Rheumatol 2005;32:744–6)

*Key Indexing Terms:*
JUVENILE DERMATOMYOSITIS
PARANEOPLASTIC PHENOMENON
NASOPHARYNGEAL CARCINOMA
MALIGNANCY

Dermatomyositis (DM) in adults is frequently associated with cancer. In contrast, juvenile DM (JDM) is predominantly idiopathic and rarely reported with occult neoplasm. We describe a patient who presented with DM that was found to be a paraneoplastic manifestation of nasopharyngeal carcinoma.

## CASE REPORT

A 15-year-old Moroccan boy was referred to us because of malar rash, periorbital edema, and muscle weakness. Two months earlier he had been investigated at another hospital for an enlarged cervical lymph node. At that time there was investigation of full blood count, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), Mantoux test (PPD), and a neck ultrasound, which were all negative. Serological tests for cytomegalovirus, Epstein-Barr virus (EBV), and toxoplasma were negative for a recent infection, with IgM persistently negative. He was then treated with antibiotics, with mild improvement.

On examination there were Gottron's papules on his knuckles and elbows (Figure 1), and heliotrope and malar facial rash. Gower's sign was present and he had moderate proximal muscle weakness, particularly of the psoas and recto-abdominal muscles. Examination of heart, lungs, and abdomen was normal. In addition, lymph nodes that were enlarged and moderately hard to touch were palpable on the left side of the neck.

We made a clinical diagnosis of JDM and performed the following investigations: complete blood count was normal, ESR 58 mm/h, CRP 6.75 mg/l (normal up to 5 mg/l), creatine phosphokinase (CPK) 4760 U/l (normal 0–190), aldolase 43.0 U/l (normal 1.0–7.7), neopterin 20.0 mmol/l (normal 1.9–15), lactate dehydrogenase 1021 U/l (normal 190–338), SGOT 203 U/l (normal 10–45), and CD19+ lymphocytes 23% (normal 5–16). Complement factors, biochemical profile, and clotting tests were normal. Videocapillaroscopy showed thickening of capillaries and areas of dropout. Electromyography result was suggestive of myopathy and there was also a denervation pattern. Magnetic resonance imaging (MRI) of the thigh muscles revealed moderate hyperintensity consistent with edema of the crureus and both the internal and external obturator muscles. Thus, the patient fulfilled the Bohan and Peter criteria[1] for JDM.

Biopsies of crureus muscle and of the neck lymph nodes were performed before starting treatment. Under steroid treatment, the patient rapidly improved, with reduction of muscle weakness, improvement of the skin, and significant decrease of muscle enzymes (CPK 1793 U/l, aldolase 35.6 U/l, SGOT 138 U/l).

Surprisingly, the muscle pathology showed no abnormalities, while the lymph node showed well defined aggregates of epithelial cells surrounded by fibrous tissue and lymphoid cells with typical pattern of proliferation, and tumor cells showing a high mitotic rate. This was consistent with the diagnosis of undifferentiated nasopharyngeal carcinoma (NPC). The presence of EBV in the tumor tissue was investigated using EBV-encoded small RNA (EBER). A strong nuclear staining was observed in many tumor cells by *in situ* hybridization (Figure 2). An MRI of the head and neck showed a mass infiltrating the roof and the lateral walls of the rhinopharynx. The patient was given chemo-radiotherapy treatment, with complete remission of both NPC and JDM that continues after 3 years.

## DISCUSSION

The association between adult onset dermatomyositis and cancer is well known, and has been reported with a prevalence ranging from 7% to 60%[2-4]. Thus extensive screening for occult cancer is a common practice in all adult patients with DM, particularly when the onset is after 50 years of age[5]. Malignancies most frequently linked to DM are similar to those of an age and sex matched population and include carcinoma of the lung, ovary, gastrointestinal tract, breast, and testes[6]. By contrast, JDM is predominantly an idiopathic disease, and reports of association with occult neoplasm are extremely rare[6,7].

The temporal association of cancer with DM can help identify the underlying pathogenic relationship. In adults an

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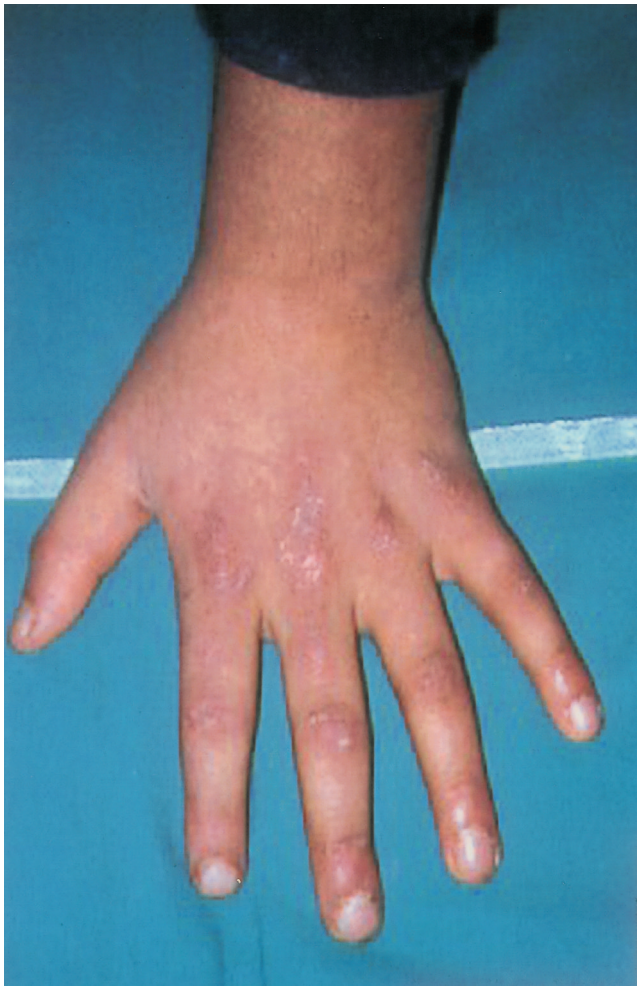

*Figure 1.* Classical clinical signs of JDM on the patient's hand: Gottron's papules, periungual erythema, and cuticle overgrowth.

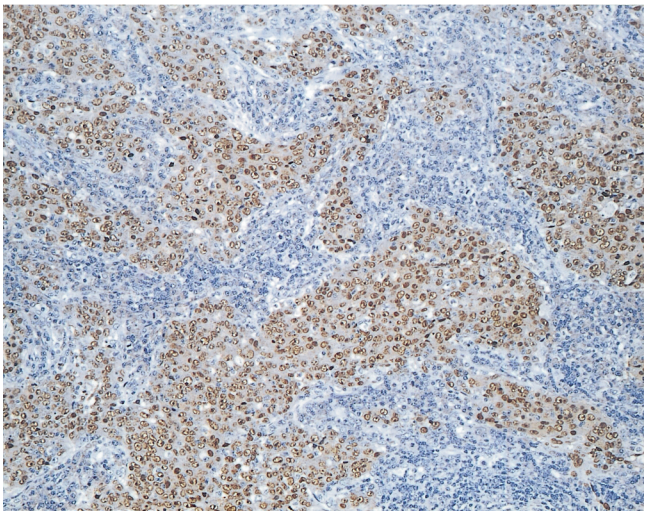

*Figure 2. In situ* hybridization for EBV. Well defined aggregates of epithelial cells surrounded by fibrous tissue and lymphoid cells (brown); the tumor cells show strong nuclear staining for EBV (original magnification ×20).

increased risk of cancer has been found in the 4 years preceding or following the diagnosis of DM[8], supporting the hypothesis that DM may represent a paraneoplastic phenomenon in some patients. This is confirmed by reports of improvement in DM after successful treatment and worsening with recurrence of the cancer.

In our patient the close temporal association between onset of the 2 conditions and the remission of all clinical and laboratory signs of muscle involvement after radiochemotherapy suggest that JDM was a paraneoplastic syndrome related to NPC.

NPC is frequent in southern China, rare in the Western countries, and has an intermediate incidence in the Mediterranean basin[9]. In children, NPC is very rare, constituting 1%–5% of all cancers, and an association with EBV infection has been reported[10]. EBV genome has been identified in tumor cells and high titers of IgG and IgA against EBV antigens are detected in patients with NPC[11,12]. A pathogenic role of EBV in JDM has not been observed[13]. To our knowledge, our patient is the first with JDM and documented EBV-associated NPC. By contrast, an association between NPC and DM has been reported in adults, particularly in Asian countries[14].

The etiology of DM in patients with cancer is still unknown. Proposed mechanisms include (1) a common environmental trigger for both cancer and myositis in genetically susceptible hosts, (2) tumor products causing muscle and skin inflammation, and (3) cross-reactivity between tumor and muscle or skin antigens inducing dysregulation of the immune system[15].

In adult-onset DM, necrotic skin changes, old age, itch, ESR elevation, HLA association, and autoantibody expression have been suggested to be predictive of malignancy. In particular, ESR > 35 mm/h has been found to be the most reliable predictive factor suggesting the presence of cancer in DM[16].

In our patient 2 features unusual for JDM were seen: persistent lymphadenopathy and elevated ESR. However, the fact that lymphadenopathy is common in childhood, the negative investigations one month earlier, and the characteristic features of JDM were falsely reassuring. Finally, the lymph node biopsy, which was done because of the persistent enlargement of the glands, led to the correct underlying etiology of the JDM syndrome.

This is the first report of an association between 2 extremely rare conditions, nasopharyngeal carcinoma and paraneoplastic dermatomyositis, in a child. This observation suggests that, although rare, paraneoplastic JDM must be considered in the differential diagnosis, particularly when unusual features such as high levels of inflammatory markers or suspicious clinical signs such as lymphadenopathy are present. It is crucial to identify these patients early because

steroid treatment can partially suppress the tumor growth and lead to delay in establishing the correct diagnosis.

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