

Letters to the Editor

Butterworth Heinemann, 1998.

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Juvenile dermatomyositis: Medical and psychosocial team approach

Sirs,

In its extreme manifestation, juvenile dermatomyositis (JDM) may involve the loss of muscle function sometimes to the degree of total disability, with a need for assistance in everyday life (1,2). The patient and his family need to adjust to the chronic nature of the disease, various therapies, and changes in the equilibrium of the entire family. The patient we describe is unique in having experienced three major catastrophic life events in a short period of time, which he and his immediate family had to overcome. He was an 8-year-old boy, previously healthy, and a talented soccer player who was admitted to our pediatric ward with fatigue, muscle weakness and skin rash. Ten months earlier his grandmother, with whom he had a daily close relationship, passed away due to cancer following prolonged hospitalization at our hospital. At the end of the week of mourning his father unexpectedly vanished, departing to another country because of severe financial complications and threats to his life. About 4 months before his admission our patient developed rash and weakness. Gradually he discontinued his soccer playing, social activities and later refused to go to school. The family thought the child was malingering as a response to the traumatic life events that he had to confront.

Upon examination the patient was bedridden. The clinical diagnosis of JDM was supported by EMG, MRI, US and muscle biopsy. Considering the advanced stage of his illness, the prolonged course until diag-

nosis, and his depressed mood, we decided on aggressive management - a combination of corticosteroids, methotrexate, immunoglobulins, an intensive physiotherapy program, and daily supportive talks. To encourage him and boost his spirits, we managed to arrange for the number one soccer star in the country to pay him an encouraging visit. The boy gradually regained his previous physical abilities. In a follow-up of 2 years he has resumed normal muscle strength and is off medications.

The psychiatrist diagnosed that the patient was suffering from two major interacting problems. The more obvious one was the physical diagnosis of JDM with its accompanying pain, weakness, and loss of mobility, which rendered him bedridden. This physical blow was superimposed on the pre-existing acute grief reaction due to the loss of his grandmother, to whom he was attached, and the loss and betrayal of the father. The accumulated effect of these frightening catastrophic events brought about a reaction that may be likened to a post-traumatic stress disorder. The observable symptoms and signs were loss of interest and severe social withdrawal, low threshold to social stimuli, and frequent outbursts of anger, listlessness, sleep disturbances, and marked regression.

We perceived the complexity of the patient's emotional reaction from the start and responded accordingly. While considerable attention and empathy were provided, we insisted on reciprocity and active participation. The road to making real emotional contact was through his hobby - soccer, discussion regarding the sport, and physical therapy fitted to soccer-related themes. The idea to bring in the national super-star soccer player to his bedside was a turning point in the course of his hospitalization. From the boy's point of view there was a sense of meaning and even of pride replacing the shame, fear and bewilderment that he had experienced before.

Our case highlights the importance of a cooperative team approach in treating the patient, not only his body but also his soul. Both are related; emotional stress may lead to or enhance and aggravate a chronic disease (3,4). Stress has also been suggested to be a predisposing factor in polymyositis/dermatomyositis (5).

The coping-adjusting skills of the patient and his family are key factors in the treatment and control of the disease (6). Hence, a multidisciplinary team approach and psychosocial interventions are crucial and in this case proved to be an integral part of the treatment (7, 8).

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MRI diagnosis and successful treatment of upper cervical spine synovitis in a patient with juvenile chronic arthritis

Sirs,

Cervical spine involvement in juvenile chronic arthritis (JCA) may lead to typical permanent changes, which may cause disabilities and increase the risk of complications for the rest of the patient's life (1-5). Early diagnosis and active treatment of the inflammation may be preventive, which was demonstrated here in the patient's history.

A 2 1/2 year old girl came to our hospital because of arthritis in her right knee lasting for two months. ESR was 50 mm/h and CRP 30 mg/l. JCA (antinuclear antibody positive) was diagnosed. Local triamchi-

nolone injection was given and treatment with naproxen, hydroxychloroquine (HCQ) and auranofin was started. Auranofin and naproxen were discontinued after 23 months because of remission. HCQ was stopped 5 months later as the remission seemed to continue. The girl was symptom-free for 10 months after that, but subsequently began to complain of pain and stiffness in her neck. No neurological symptoms or signs were present. HCQ and naproxen treatments were resumed and she used a stiff collar to relieve cervical symptoms. The pain continued, but dynamic cervical spine radiographs were still normal 5 months later.

Because of continuous symptoms in the neck, magnetic resonance imaging (MRI) of the cervical spine was undertaken 9 months after the radiography. Synovitis was found in the atlanto-axial joints (Fig. 1a). CRP was 0 mg/l and ESR was 17 mm/h. Because of the definite inflammation in a rather critical joint area, methotrexate was added to the treatment. The neck complaints were gradually relieved and she was symptom-free 7 months later when the MRI examination was repeated. The former synovitis had disappeared (Fig. 1b). Cervical spine radiographs were taken 16 months later to check whether any permanent changes had developed during the inflammation period, but they were normal.

Chronic inflammation of JCA may cause several types of cervical spine abnormalities. Some of these are typical of JCA, e.g. disturbance in the growth of the vertebrae and ankylosis of the intervertebral joints,

but changes typical of adult onset rheumatic diseases may also develop. These changes are potentially dangerous, and even fatal complications have been described (1-5). The aim of treatment is to prevent or at least retard the development of permanent joint damage and disabilities. Thus, the inflammation has to be diagnosed early and treated aggressively. It has been shown in RA that the development of cervical spine disorders may be retarded by DMARDs, but there are also possibilities for active treatment if some abnormalities are already present (6, 7).

It is difficult to diagnose active synovitis in the cervical spine, especially if no other joints are inflamed. It cannot be done by clinical examination, laboratory tests or ultrasound, and some permanent changes have already developed by the time plain radiography is diagnostic (2-5). MRI is an excellent means of diagnosing synovitis in the cervical spine (1,8-10). A definite diagnosis can be made early, the approach is safe, and it does not expose the patient to radiation. Unfortunately MRI is rather expensive and children may need light general anaesthesia during the time-consuming examination. The availability of MRI is also frequently a problem, but hopefully will be less so in the future.

MRI offers a viable means for the early diagnosis of synovitis in cases with treatment-resistant neck pain. It is not needed if the findings will not influence treatment decisions. For example, in active polyarthritis the need to improve systemic treatment can be evaluated by clinical examination,

and MRI is not essential to describe any local synovitis. In our case the MRI finding gave us a reason to treat the JCA more aggressively, and this time successfully, with methotrexate.

We have found only one report of the use of MRI in the examination of the rheumatoid cervical spine in patients with JCA (1). Further research is needed on this important topic.

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(a)



(b)

Fig. 1. (a) A T2-weighted coronal magnetic resonance image (F8E 3900/126, 90°, 3 mm, 256 x 256) of the upper cervical spine in a 7-year-old girl with JCA (oligoarthritis) and long-lasting neck pain and stiffness. The bright signal in the lateral atlanto-axial joints (arrows) is caused by active synovitis. (b) The same after active medical treatment. The synovitis has disappeared.