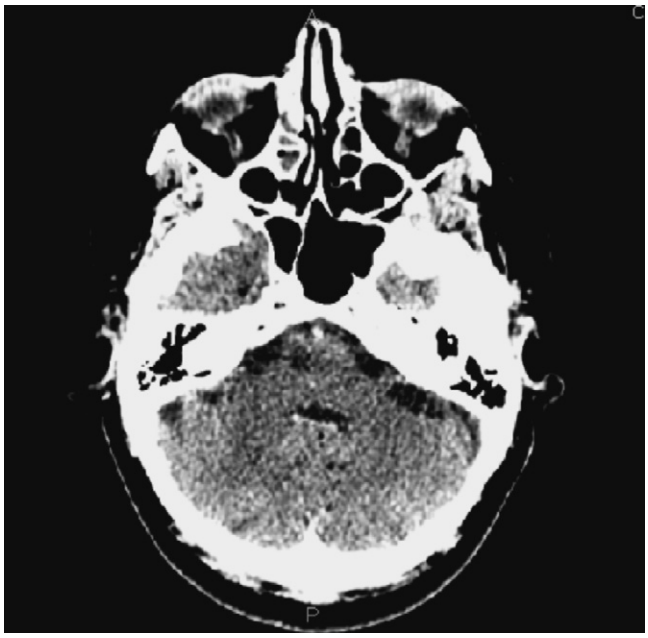

Fig. 1. Computed tomography, axial section through the pons: infarction seen as a hypodense zone in the right part of the pontine protuberance, at the site occupied by the pyramidal tract.

She returned to work on October 8, 2008. On November 15, her neurological evaluation was unchanged compared to the previous visit. On the right, pressure on eight of nine fibromyalgia points caused pain and flinching; whereas, on the left six points were slightly tender (discomfort without flinching). At the arms, the blood pressure cuff caused pain (visual analog scale pain score of 5/10) at 170 mmHg on the right and 220 mmHg on the left; corresponding figures at the legs were 120 and 220 mmHg.

The pathophysiology of fibromyalgia is complex and multiple factors are probably involved. A role for the central nervous system has been documented by studies showing central pain sensitization and hyperexcitability [2–4]. Recent data suggest impaired sensorimotor control or abnormalities in the motor system [5–7]. Our case-report supports a role for the central motor system. The infarction visualized by computed tomography seemed confined to the pyramidal tract, with no involvement of sensory pathways. The patient had isolated motor deficits. Thus, the resolution of the muscle pain on the paralyzed side was unexpected, as sensory function remained normal. Neither the spasticity nor the substantial muscular exertion required by the rehabilitation program exacerbated the muscle pain. Our case-report supports a role for a functional abnormality in motor control, which may be either primary or secondary to defective sensorimotor control. This functional abnormality may increase motor activity above the level required to maintain posture or to perform a movement, resulting in chronic muscular overload and in fatigue. The pyramidal tract involvement in our patient may have lowered this abnormally high level of central motor activity, leading to resolution of the muscle pain in the left side of the body.

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Intravenous immune globulins to treat eosinophilic fasciitis: A case report

Keywords: Eosinophilic fasciitis; Shulman syndrome; Eosinophilia; Immunoglobulins

1. Introduction

Eosinophilic fasciitis or Shulman's syndrome was first described in 1974 as a syndrome of diffuse fasciitis with high peripheral eosinophil counts [1]. This rare condition usually responds to glucocorticoid therapy. In patients with glucocorticoid-resistant or dependent disease, immunosuppressant therapy may be considered. We report the case of a glucocorticoid-dependent patient who experienced a partial response to methotrexate therapy followed by a full response to intravenous immune globulin (IVIg) therapy.

2. Case report

A 39-year-old male was admitted for swelling and induration of the hands and feet with severe functional impairment. He

reported Raynaud's phenomenon since adolescence but denied any recent exacerbation. For the last 5 months he had been experiencing inflammatory joint pain in the hands, wrists and shoulders, as well as muscle pain in the upper limbs. There was no fever. He had lost 8 kg and had dysphagia for fluids. The skin over the forearms and legs was thick and stiff to palpation. Abduction and flexion of the shoulders was limited and motion range limitation was also noted at the elbows, hands, and knees. His peripheral leukocyte count was 13,400/mm^3 with 3283 eosinophils/mm^3. The C-reactive protein level was elevated to 17.8 mg/L (normal, < 3.0) and the erythrocyte sedimentation rate was normal. Results were normal for serum assays of calcium, urea, creatinine and transaminases, whereas the serum aldolase level was high (13.1 U/L; normal, < 7.6). The serum protein assay and electrophoresis were normal. The immunoglobulin G (IgG) level was elevated to 2000 mg/dl (normal, 650–1500) and protein immunofixation electrophoresis detected a monoclonal IgG. No proteinuria was found and findings were normal from immunoelectrophoresis of a urine sample. The IgE level was normal. Tests were negative for antinuclear antibody, rheumatoid factor, anticentromere antibodies and anti-SCL70. Complement levels were normal. Tests for parasites were negative. An evaluation for cardiac or respiratory disease was normal.

Electromyography of the upper limbs showed a myogenic pattern at the biceps brachii muscle on both sides. Radiomanometry results indicated esophageal dysmotility. Capillaroscopy findings were normal. A bone marrow biopsy showed myeloid hyperplasia with relative eosinophilia and no evidence of malignancy. Findings from a biopsy specimen of skin, fascia and muscle taken from the right upper limb consisted of fibrous thickening and an infiltrate composed of lymphocytes and plasma cells with a few eosinophils (Fig. 1). The muscle contained foci of interstitial myositis with an inflammatory infiltrate but no necrosis (Fig. 2). The diagnosis was eosinophilic fasciitis.

Prednisone 30 mg/d and cimetidine 800 mg/d were started. Two months later, only small clinical improvements were apparent. The eosinophil count and aldolase level were normal, the monoclonal gammopathy had resolved and the C-reactive protein level was down to 1.6 mg/dl. When the prednisone dosage was tapered to less than 20 mg/d, the clinical symptoms flared and the aldolase level increased. Methotrexate was started in an increasing dosage of up to 20 mg/week. The skin abnormalities improved slightly. Seven months later, the persistent clinical symptoms and glucocorticoid dependency prompted IVIg treatment in a dosage of 0.5 g/kg/d on 3 consecutive days. One month later, he had very substantial improvements in general health and mobility and his skin was more pliable. He received five additional IVIg treatments at intervals of 1 month. His condition was then substantially improved and his aldolase level normal. The IVIg treatment was stopped. He had mild cutaneous induration over the legs and slight motion range limitation at the hands and was therefore kept on methotrexate therapy. At last follow-up 2 years after IVIg discontinuation he was free of symptoms and had normal laboratory tests with 2.5 mg prednisone/day and 10 mg methotrexate/week.

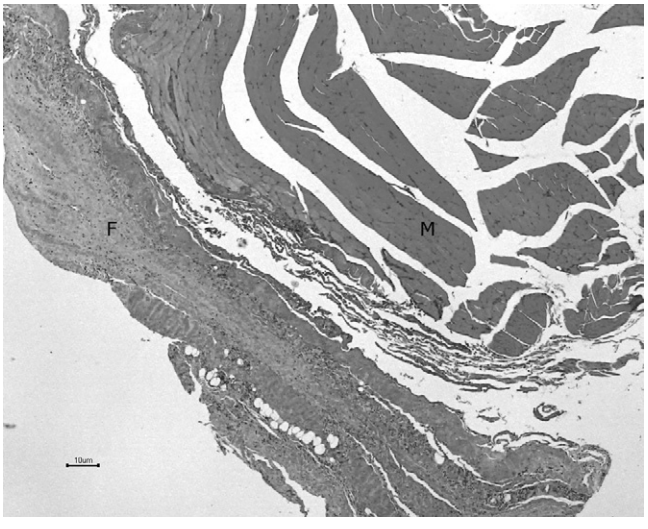

Fig. 1. Fibrous thickening and infiltrate of lymphocytes and plasma cells in the fascia (F) and striated muscle fibers (M).

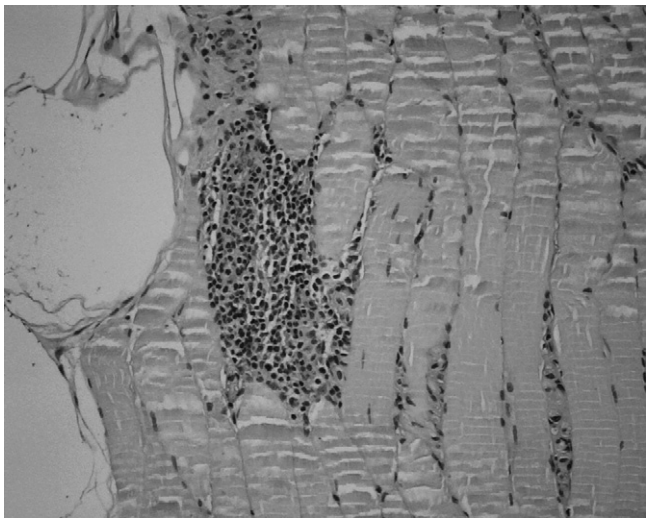

Fig. 2. Focus of interstitial myositis with an inflammatory infiltrate between the striated muscle fibers.

3. Discussion

Eosinophilic fasciitis is slightly more common in males than in females and can occur in all age groups [2,3]. The cause is unknown. The symptoms often set in rapidly, sometimes after an unusually strenuous physical activity [2,3], although this was not the case in our patient. Raynaud's phenomenon is usually absent and organ involvement is uncommon. The presence of Raynaud's phenomenon in our patient was probably coincidental. A number of hematological disorders have been reported in patients with eosinophilic fasciitis, apparently as a result of chance alone; they include Biermer's anemia [4], myelomonocytic leukemia [5], lymphoproliferative disorders and monoclonal gammopathies [6,7]. Others may occur as complications of eosinophiilic fasciitis, such as bone marrow aplasia.

Muscle manifestations are uncommon in eosinophilic fasciitis. Moderate muscle weakness may be found, as well as a

modest increase in muscle enzyme levels, which may mirror disease activity [8]. The electromyogram may show a myogenic pattern and the histological examination usually reveals interstitial myositis [9].

The treatment of eosinophilic fasciitis rests on glucocorticoid therapy in a dosage of 0.5 to 1 mg/kg/d. Patients may take 2 to 4 years to recover and relapses may occur. Medications that have been used in patients failing glucocorticoid therapy include cimetidine, hydroxychloroquine, methotrexate, cyclosporine and cyclophosphamide [10]. In our patient, cimetidine was unhelpful and methotrexate was inadequately effective. There have been two anecdotal case-reports of successful treatment with IVIg [11,12]. Thus, IVIg therapy may constitute a useful alternative to immunosuppressants. The treatment of eosinophilic fasciitis may carry a risk of iatrogenic events, most notably when the response to immunosuppressants is incomplete. Data from a larger number of patients are needed to better assess the usefulness of IVIg therapy.

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Bifocal sarcomatous transformation of Paget's disease: A case report

Keywords: Paget's disease; Extramammary; Cell transformation; Neoplastic; Bifocal; Osteosarcoma

1. Introduction

Paget's disease of bone is a benign dystrophic condition first described in 1877 by Sir James Paget [1]. Serious complications may arise, such as sarcomatous transformation, which is estimated to occur in less than 1% of patients [2]. We report a case of sarcomatous transformation at two different sites.

2. Case report

This 83-year-old man had been diagnosed more than 15 years earlier with polyostotic Paget's disease of bone, for which he had declined follow-up and treatment. A right pleural effusion developed. Computed tomography (CT) showed lysis of the neural arch of T4 and involvement of the soft tissues in the spinal canal, with mild spinal cord compression (Fig. 1). He reported mechanical pain in the right groin, and edema of the right leg was noted. The serum alkaline phosphatase level was elevated to 4962 IU/L. A radionuclide bone scan showed considerable extension of the pagetic lesions and a second lytic lesion, located in the right acetabulum (Fig. 2) and putting pressure on the iliac vein. Examination of a bone biopsy specimen established the diagnosis of osteoblastic osteosarcoma. Treatment consisted of 12 sessions of palliative radiation therapy to both sites aimed at decreasing the pain and compression, zoledronic acid, and supplemental vitamin D and calcium.

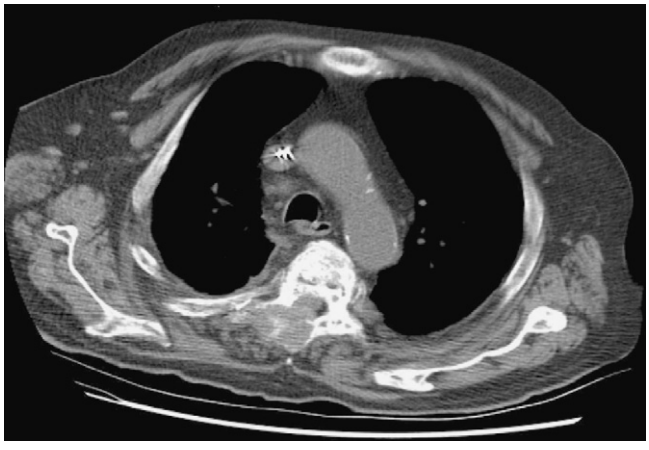

Fig. 1. Osteosarcoma complicating Paget's disease of bone. Computed tomography: lytic lesion in T4 with spread to the soft tissues.