Results The indications of IVIG therapy were: severe thrombocytopenia with active disease despite corticosteroid and immunosuppressive therapy in 7 patients and concurrent mycobacterial infection in 2 patients with SLE; resistance to standard treatment in 7 patients with PM/DM and 3 patients with WG. Fifteen of 20 patients received concomitant high dose prednisolone, methotrexate, azathioprine and/or pulse cyclophosphamide therapy. In two patients, IVIG was used because of concomitant viral infections (one patient with DM+chronic active hepatitis due to Hepatitis B virus and 1 patient with WG+cytomegalovirus chorioretinitis). Two patients with DM died of underlying malignancies and 3 patients with SLE died of uncontrolled disease activity. No serious side effects related to IVIG therapy were observed.

Conclusion High dose IVIG is an effective and tolerable option when standard therapies are failed or a rapid response is needed in patients with PM/DM and WG. In SLE the main indication for IVIG is steroid resistant severe thrombocytopenia. IVIG therapy may also be preferable in patients with active disease and concurrent severe infections.

AB0213 RARE COMPLICATIONS OF PAGET'S DISEASE

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Background Paget Disease's (PD) myelopathy and chondrosarcoma are rare entities. The spinal cord compromise is due to the direct compression of the growing bone and also to spine cord ischaemia. Chondrosarcoma is a rare malign degeneration of the bone disease.

Objectives

Methods

Results Case 1: A 74 years old man with the previous diagnosis of polyostotic PD was admitted to our inpatient Unit with lower limbs paraparesis that began 2 months earlier. He had a spastic gait, lower limbs paresis 4/5, increased patellar and Achilles tendon reflexes (more prominent on the left), extension plantar reflexes, T6/T7 sensory level without sphincter dysfunction. Both alkaline phosphatase and hydroxyprolinuria were high. Thoracic spine CT scan and MRI showed myelopathy at T5, T6 and T7 due to spine PD compression. Therapy consisted of pamidronate 180 mg (2 infusions of pamidronate 90 mg, 2 weeks apart) and 200 UI i.d. of inhaled calcitonin. After therapy a clear clinical and laboratorial improvement was observed. After 1 year the gait and neurological observation were normal.

Case 2: A 76 years old woman with PD involving the thoracic spine and iliac bone, was admitted to our Unit with lower limbs paraparesis, nocturnal back pain and right gluteal pain irradiating to the posterior aspect of the thigh with 4 months duration. Neurological evaluation showed paraparesis grade 1–2/5 on the right lower limb and grade 4/5 on the left, decreased right patella reflex and sensory level T12–L1. A large stony mass was palpable in the right iliac fossae and in the gluteal area. Bone turnover markers were high. Spine CT scan and MRI showed myelopathy due to PD vertebrae in T12–L1. Thoracic and abdominal CT scan revealed a bone tumour of the right iliac and pulmonary metastases. Histology of the tumour revealed a chondrosarcoma. She started calcitonin 200 UI i.d. and intravenous dexamethasone 5 mg t.i.d. with neurological improvement. She suffered an episode of sudden death 1 month later at home.

Conclusion In cases of PD with spinal cord symptoms, PD myelopathy should be the first clinical suspicion. Prompt diagnosis and therapy should be instituted to prevent irreversible neurological damage. Chondrosarcoma is an aggressive tumour, non responsive to standard therapies for bone malignancies.

AB0214 THE SECOND PHYSICIAN BEHIND THE USE OF COLCHICINE FOR THE TREATMENT OF FAMILIAL MEDITERRANEAN FEVER

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Background In 1972 Dr. Goldfinger of the Massachusetts General Hospital in Boston published one of the most quoted 'letters to the editor' in New England Journal of Medicine, 'Colchicine for Familial Mediterranean Fever', (FMF).¹ Here he described 5 patients whose FMF attacks resolved by the continuous administration of colchicine. This 30 year old observation has been reproduced by numerous controlled studies and changed the prognosis of this disease dramatically.

Objectives To document the contribution of Professor Emir Özkan MD, on the use of colchicine in FMF.

Methods

Results What is not widely known is the contribution of another physician, Dr. Emir Özkan, professor of cardiology in the Medical Faculty of University of Istanbul, to the treatment of FMF. Dr. Özkan and his colleagues published their findings in an article entitled 'A New Approach to the Treatment of Periodic Fever' in the Medical Bulletin of Istanbul Medical Faculty.² This article included their favourable results in 14 patients with FMF and also reported the disappearance of proteinuria in a patient with amyloidosis after 6 months of colchicine treatment. The authors had submitted this article on April 17, 1972, some 8 months before the appearance of the Goldfinger article in the New England Journal of Medicine which appeared in the December 21 issue of the journal.

Conclusion A Goldfinger-Özkan Award for Excellence in Clinical Science and Management of FMF, was given for the first time during the II. International Congress of FMF in Antalya, Turkey, in May 2000.

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AB0215 MULTICENTRIC RETICULOHISTIOCYTOSIS: A CASE REPORT

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Background Multicentric reticulohistiocytosis is a rare systemic illness which primarily affects joints and skin. The illness has a female predominance and usually begins during the fourth decade.¹

Objectives We describe a patient with multicentric reticulohistiocytosis whose manifestations have started at a much younger age than mentioned in the literature.

Methods

Results 18-year-old female patient presented with a complaint of arthralgia at her wrists, knees and ankles. Her complaints had started 6 months before admission with arthralgia and painful swellings at the wrists and knees. On admission she had active symmetric arthritis at the shoulders, elbows, wrists and knees. Synovial hypertrophy was prominent at the wrists, knees and MCP joints. Cutaneous examination showed a single 0.5 × 0.5 cm erythematous nodular structure at her 3 MCP joint. There were orange coloured hypertrophic lesions at her nail-folds. Her systemic examination was not remarkable. Her ESR was 29 mm/h, CRP 96 mg/dl, RF(-), Hb: 11.5, WBC: 10.300, platelets 289.000. Liver and kidney functions were normal. x-rays of the hands showed periarticular osteopenia, there were no erosions. Arthrocentesis showed the characteristics of an inflammatory synovial fluid. She was thought to have active rheumatoid arthritis and was started on 7.5 mg prednisolone and sulphasalazine 2 g/day. The dosage of prednisolone was gradually increased to 30 mg/day as there was no improvement in her active arthritis. Methotrexate 7.5 mg/week was added. During her follow-up she developed orange papulonodular lesions on her forehead and pinnae. Biopsy taken from the 3MCP joint revealed reticulohistiocytoma. 10 months after the diagnosis she is still under close surveillance as the disease can be associated with malignancy in 25% of patients. She is currently on methotrexate 12.5 mg/week and prednisolone 20 mg/day. The active arthritis at the shoulders and knees has resolved but she still has active arthritis at the wrists.

Conclusion Although multicentric reticulohistiocytosis affects predominantly middle-aged women the disease should be kept in mind in the differential diagnosis of active arthritis for younger age groups as well.

REFERENCE

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AB0216 TRANSFORMATION OF PRIMARY HYPERTROPHIC OSTEOARTHROPATHY INTO SECONDARY CAUSED BY BRONCHOGENIC CARCINOMA

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Background

Objectives This paper reports on the two cases of PHO that transformed into SHO under the influence of bronchogenic carcinoma.

Methods

Results The patients had not have any sign of PHO activity until the last 3 and 4 months, when a malignant process of the lungs developed and reactivated all symptoms of the disease (joint swelling, moist palms and soles, unpleasant odour of perspiration, deeper folds on the forehead and deeper nasolabial furrow, enlarged fingers, erythema around the nails). Scintigraphy showed increased radioisotopes accumulation in particular regions of the skeleton.

Conclusion To our knowledge, similar cases have not been described in literature.

AB0217 INTRAVENOUS PULSE CORTICOSTEROID THERAPY IN PATIENT WITH EOSINOPHILIC FASCITIS

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Background Eosinophilic fascitis is a disorder characterised by the appearance over weeks or months of tender swelling of the arms and legs with inflammation and sclerosis of the deep fascia and subcutis. Long lasting therapy with prednisone is often unsatisfactory.

Objectives A 40-year-old man suffering from eosinophilic fascitis is reported with skin rash and induration of subcutaneous tissue of the forearm, typical "groove sign" and contractures of the elbow joints (pict.1,2,3). Skin involvement was accompanied by peripheral eosinophilia (0,33) hypergammaglobulinemia (0,24), an elevated erythrocyte sedimentation rate (28/h) increased concentration of acute phase reactants (C-reactive protein 52 mg/l), alpha-1 acid glycoprotein 1489 mg/l) as well as leukocytosis (14,5 G/l).

Methods The typical treatment with dosis of 20 mg prednisone caused only slight improvement. Moreover the patient has suffered from typical poststeroidal skin changes. Therefore we decided to perform an intravenous pulse methylprednisolone therapy as follows: 3 days-every day 500 mg methylprednisolone as solution in 5% NaCl (two courses with 4 weeks pause). As a continuation the patient has got orally dosis 7,5 mg prednisone every second day in the morning.

Results After pulse therapy the remission of the symptoms with regression of skin changes and improvement of the acute phase reactants levels (ESR 14/h, CRP 28 mg/l) as well as the normalisation of leukocytosis (10,8 G/L) and eosinophilia (0,03) were observed. After one year observation the patient was still in remission without signs of active inflammatory process and without eosinophilia in blood wet.

Conclusion We recommend in patients with fascitis eosinophilica the intravenous pulse corticosteroids therapy and reduced dosis of prednisone as an alternative to high-dose oral corticosteroids.

AB0218 AMYOPATHIC DERMATOMYOSITIS ASSOCIATED WITH PNEUMONITIS: IS METHOTREXATE A RISK FACTOR ?

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Background We described a case of dermatomyositis which subsequently developed interstitial lung disease. 35 years old woman was admitted to our department complaining of dermal eruptions without weakness of muscles. Characteristic skin changes included Gottron's papules on the dorsal aspect of interphalangeal joints, elbows and patella, heliotrop discoloration of eyelids with associated periorbital oedema, macular erythema of anterior neck and chest (V sign), face and forehead and mechanic's hands. Serum CK levels and EMG was normal and no histological abnormality by muscle biopsy was found.

Since she had not claimed evidence of muscle disease she was diagnosed as having dermatomyositis sine myositis (Amyopathic dermatomyositis) and initially treated with prednisolone (40 mg/day) and methotrexate (10 mg. per week). Since her condition didn't changed high dose intravenous immunoglobulin (IVIG)