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