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.

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

Z Jajic, I Jajic. Department of Rheumatology, University Hospital "Sestre Milosrdnice", Zagreb, Croatia

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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

W Samborski, R Eaba. Rheumatology, K. Marcinkowski University Medical School, Poznań, Poland

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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 ?

I Tekeoglu, A Akyokus, T Tuncer, B Butun, C Kacar, E Gilgil, G Arslan. Department of Rheumatology, Akdeniz University Faculty of Medicine Hospital, Antalya, Turkey

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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)

therapy (1 gr/kg in three divided doses) was administered each month for six months.

Marked improvement in dermal eruptions was observed following the second administration of IVIG therapy. Now in the eighth month of follow up she has been complaining of dyspnea on exertion for twenty days. High resolution computerised tomography (HRCT) of lung revealed paranchimal changes suggesting that bronchiolitis obliterans organising pneumonia (BOOP).

Could this be the result of methotrexate therapy or a result of amyopathic dermatomyositis? Lung involvement in amyopathic dermatomyositis is rare. Pulmonary drug toxicity is increasingly being diagnosed as a cause of acute and chronic lung disease. BOOP, which is commonly caused by cyclophosphamide and bleomycin as well as methotrexate and gold salts, appears on HRCT scans as poorly defined nodular areas of consolidation, centrilobular nodules, and bronchial dilatation. Knowledge of the drugs most frequently involved can facilitate diagnosis and institution of appropriate treatment. The results of lung biopsy will be discussed in poster session. If lung biopsy will reveal the pathology methotrexate will be discontinued.

Objectives

Methods

Results

Conclusion

AB0219 QUANTITATIVE HEMOSTATIC INDICES IN SECONDARY AND MYELOPROLIFERATIVE THROMBOCYTOSIS

¹J Parada-Turska, ²B Sokolowska, ²A Walter-Croneck, ²A Dmoszynska, ¹L Szczepanski, ²J Brzozowska, ²M Sledzinska, ²M Kokoszka. ¹Department of Rheumatology; ²Department of Hematology, Medical University, Lublin, Poland

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Background An elevated platelet count may lead to an increased incidence of systemic thromboembolism or unexpectedly to thrombohemorrhagic complications.

Both, incidences of systemic thromboembolism and unexpected thrombohemorrhagic complications (bleeding) were described in patients with an elevated platelet count.

In the present study the quantitative hemostatic indices were measured in 46 patients with thrombocytosis (platelet count > 400.000 per microliter), including 23 patients with secondary thrombocytosis (17 patients with rheumatoid arthritis, 2 with lung cancer, 1 with systemic lupus erythematosus, 1 with granuloma Wegeneri) and 23 patients with myeloproliferative thrombocytosis (12 with chronic myeloid leukaemia, 8 with polycythemia vera, 3 with idiopathic thrombocytosis).

In both studied groups, von Willebrand factor activity and epinephrine-induced platelet aggregation were decreased, and activated partial thromboplastin time was prolonged, thus, suggesting increased risk of bleeding complications.

On the other hand, the concentrations of fibrinogen and d-dimers were enhanced and bleeding time was shortened. These findings argue for an enhanced risk of thrombosis.

Since no differences between two studied groups were found, it seems that the observed abnormalities were directly related to the elevated platelet count and were not associated with primary disease.

The low incidence of hemostatic complications in patients with secondary and myeloproliferative thrombosis may suggest that the coexistence of an additional component is critical for the clinical manifestation of such disturbances.

Objectives

Methods

Results

Conclusion

AB0220 OCHRONOTIC SPONDYLOARTHROPATHY WITH MULTISYSTEMIC INVOLVEMENT: A CASE REPORT

K Nas, A Gür, R Çevik, AJ Saraç, S Söker (Çakmak), S Em, M Karakoç, S Kiliç. *Physical Medicine and Rehabilitation, Dicle University, Diyarbakir, Turkey*

10.1136/annrheumdis-2001.741

Background Ochronosis is a musculoskeletal manifestation of alcaptonuria, a rare hereditary metabolic disorder characterised by the absence of the enzyme homogentisic acid oxidase and associated with various systemic abnormalities related to the deposition of homogentisic acid pigment.

Objectives We aimed to review a case with ochronosis, a disorder of rare and multisystemic involvement, in the light of literature.

Methods A housewife, 53 years of age, was admitted to our clinic with complaints of low back and left shoulder pains. Due to low-back pain complaints, she was operated on two times with diagnosis of lumbar discal hernia.

Results The radiographic appearance in the lumbar spine of the patients was almost pathognomonic of ochronotic spondyloarthropathy. Typical water-like calcifications were seen in the intervertebral discs, with narrowing of disc spaces and osteoporotic rarefaction of the vertebral bodies. Shoulder joint, narrowing of the joint space, a marked subchondral sclerosis, and small osteophytes were observed; similarly, knee, sacroiliac joints and symphysis pubis show at narrowing of the joint spaces and subchondral bone sclerosis.

In the eyes of the patient, ochronotic pigmentation was seen by light microscopy. The blue-black pigmentation was bilateral and asymmetric in the cornea, conjunctiva and sclera. In addition, black-grey pigmentation was present in the ear cartilage and hand nails. In the laboratory test, homogentisic acid was found increase (2740 mmol HJA/mmol creatinin; N: 10 mmol HJA/mmol) creatinin in urine.

Conclusion

AB0221 FAMILIAL MEDITERRANEAN FEVER: ANALYSIS OF 45 TUNISIAN PATIENTS

SS Turki, CH Ben Taarit, R Goucha, E Abderrahim, F Ben Moussa, H Ben Maiz. *Department of Nephrology and Internal Medicine, Charles Nicolle Hospital, Tunis, Tunisia*

10.1136/annrheumdis-2001.742

Background Familial mediterranean fever (FMF) is an inherited multisystem disease of unknown aetiology, characterised by recurrent, painful, self-limited episodes.

Objectives The objectives of this study is to describe clinical profile, course and complications of 45 cases of FMF observed over a period of 25 years.

Methods Retrospective review of 45 cases with FMF.

Results Forty patients (88%) started their illness below the age of 10 years. Peritonitis occurred in 84,4%, arthritis in 35,5%, pleuritis in 17,7% and erysipelas-like lesions in 31,1%. 21 patients developed renal amyloidosis and 17 patients were subjected to unnecessary operative surgery.