ELISA analyses were used to assess membrane expression and serum levels of TNFRSF1A, respectively.

Results A missense mutation of exon 4, resulting in an amino acid substitution (F112I) close to a conserved cysteine and disulphide bond, was detected in 4 affected family members and in one asymptomatic family member. Impaired shedding of TNFRSF1A after phorbol myristate acetate stimulation was detected in peripheral blood monocytes and granulocytes from affected individuals. The soluble TNFRSF1A levels of individuals carrying the mutation ($n = 5$) were about half of soluble TNFRSF1A levels of individuals not carrying the mutation ($n = 4$).

Conclusion Dominantly inherited autoinflammatory syndrome should be suspected whenever a patient presents with a history of intermittent fevers accompanied by abdominal pain, arthritis or skin rashes, particularly in the presence of a positive family history. Among such patients, low serological levels of soluble TNFRSF1A give an indication of those individuals who are likely to have TRAPS.

REFERENCE

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THU0214 EFFECT OF TRIAMCINOLONE DIACETATE INJECTIONS IN EARLY DUPUYTREN'S CONTRACTURE AND STENOSING TENOSYNOVITIS OF THE HAND

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Background Dupuytren's contracture and other fibrosing conditions of the hand are generally progressive, causing limitation of function and decreasing grip strength. Long term results of surgical procedures are less than optimal with high recurrence and extension rates. Other non-operative therapies such as slow skeletal traction, radiation, splinting, ultrasound and enzymatic fasciotomy have not proven to be clinically useful.

Objectives The objective of this study was to determine the effectiveness of steroid injections in slowing the progression of fibrosis and contractures in early Dupuytren's disease and conditions causing stenosing tenosynovitis of the hand.

Methods Twenty-three patients were studied. There were 12 men and 11 women with an age range of 35 to 72 years. Of these patients four also had diabetes mellitus and three had rheumatoid arthritis. Eighteen patients had Dupuytren's contracture. Fifteen had bilateral contractures and three in one hand only. None of them had contractures in the proximal interphalangeal joints. One had mild flexion contracture at the second, third and fourth metacarpophalangeal joints. Two had bilateral De Quervain's tenosynovitis and three had isolated fibrous nodules in one flexor tendon in the hand. All patients received one or two triamcinolone diacetate injections in the fibrous nodules, cords and tendon sheaths. Additionally, five patients received oral non-steroidal antiinflammatory agents for one to two weeks. Patients with Dupuytren's contracture demonstrated softening of the symptomatic chords, improvement of grip strength and partial resolution of nodules. The other patients had complete resolution of their condition.

Results Patients were followed for one to seven years and maintained their functional improvement. No patient's contracture progressed or required surgery or radiotherapy. There were no

complications such as hematoma, skin necrosis, infection, granuloma formation, decrease in grip strength, transient paresthesia or flexion deformity of the fingers.

Conclusion In conclusion, early Dupuytren's disease and other fibrosing tenosynovitis of the hands can be safely and effectively treated with injections of triamcinolone diacetate and this appears to prevent progression of these conditions.

THU0215 ASYMPTOMATIC GASTROCNEMIUS MUSCLE BIOPSY: AN EXTREMELY SENSITIVE AND SPECIFIC TEST IN THE HISTOLOGIC CONFIRMATION OF SARCOIDOSIS PRESENTING WITH HILAR ADENOPATHY

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Background

Objectives The purpose of our study was to evaluate asymptomatic gastrocnemius muscle biopsy as a tool in the histologic confirmation of the diagnosis of sarcoidosis.

Methods Consequently, twenty two patients, admitted over a two year period to our department, with bilateral hilar adenopathy and a variety of symptoms compatible with sarcoidosis, were studied prospectively. Besides a complete history, physical and routine laboratory, serum angiotensin converting enzyme (SACE) determination, pulmonary function, slit lamp eye examination, PPD skin test, gallium 67 scan, and gastrocnemius muscle biopsy under local anaesthesia, after informed consent, were performed.

Results The biopsy revealed non caseating granuloma in all the patients, confirming the diagnosis of sarcoidosis. No other patient in our department received this diagnosis over the two year period of the study. The procedure was well tolerated by all patients and almost zero morbidity was noted. Erythema nodosum was present in 68.2% of the patients, PPD was negative in all of them, SACE was elevated in 59.1%, and pulmonary function was normal in the majority.

Conclusion The impressive sensitivity of the employed biopsy, its safety and ease of performance, along with the extreme rarity of muscle involvement by other granulomatous diseases, included in the differential diagnosis, may render it the procedure of choice for the histologic confirmation of sarcoidosis, presenting with hilar adenopathy.

THU0216 NSAIDS SELF-PRESCRIPTION AS A CAUSE OF UPPER GASTROINTESTINAL HEMORRHAGE

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Background Non-steroidal anti-inflammatory drugs (NSAIDs) are compounds easily to acquire over the counter that are extensively used for symptomatic relieve of pain of any origin and for platelet antiaggregation purposes.

Objectives To describe the epidemiology of NSAIDs usage and its association with upper gastrointestinal hemorrhage in a community hospital from Spain.

Methods We prospectively studied the clinical usage, indication, duration, dose, type of NSAID therapy, presence of NSAIDs self

prescription and use of concomitant drugs in all patients admitted to the hospital with upper gastrointestinal haemorrhage from October-1999 to November-2000. All patient underwent endoscopy to assess the type of gastroscopic lesion and the presence of *Helicobacter pylori*. Requirements of blood transfusion and surgery were also noted.

Results A total 63 (29%) out of 216 patients admitted with upper gastrointestinal haemorrhage were using NSAIDs (60% aspirin, 16% piroxicam, 13% diclofenac, 6% combined and 5% other) during a median of 7.5 days (range 1- >100 days). Median patients' age was 65 years (range 18-90). NSAIDs were prescribed for pain relieve (75%), platelet anti-aggregation (19%) or both (6%). Indication for prescription was done by rheumatology consultants (0%), general practitioners (25%), other consultants (33%) and self-prescribed (41%). Location of lesions at endoscopy were: esophageal (5%), gastric (35%), duodenal (43%) and, multiple lesions (17%). *Helicobacter pylori* was present in 28 (70%) out of 40 samples. A total of 44 (70%) patients had at least one risk factor for NSAIDs gastropathy of whom only 2 (4%) were receiving prophylaxis with protein pump inhibitors and 1 (2%) with H2-blockers. NSAIDs gastrointestinal haemorrhage accounted for a total of 346 admission days, 40 (63%) patients needed blood transfusion and one (1%) suffered surgery.

Conclusion Self-prescription is a common cause of NSAIDs gastrointestinal haemorrhage. Only a minor proportion of patients with risk factors for NSAIDs gastropathy receive appropriate prophylaxis with protein pump inhibitors.

THU0217 JOINT HYPERMOBILITY SYNDROME: A PROSPECTIVE STUDY OF ARTICULAR AND NON-RHEUMATIC MANIFESTATION IN A VENEZUELAN POPULATION

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Background

Objectives Although Joint Hypermobility Syndrome (JHS) is a common condition, there are clearly a number of articular and non-articular manifestations. Because of this, one study of subjects with JHS was performed in our country.

Methods A prospective, descriptive and analytical of 41 patients attending our Unit from February up to September 1999, and who fulfilled the Beighton Scale for JHS (Beighton P, Grahame R, Bird H (1999) Assessment of hypermobility. In Beighton P (ed) *Hypermobility of Joints*. Spring, London, pp:9-22) was made. The mean age was 32 years, there were 35 females (85.4%) and 6 males (14.6%). All over the group were evaluated through Beighton Scale (BS), being 4 points the lowest and 9 points the highest. All subjects were examined for echocardiogram and a gynaecological exam was performed in all sexually active women.

Results 11 patients (26.8%) had 9 points in the BS and 1 patient (2.4%), being the mean of the group of 6 points. The dorsiflexion of fifth finger was found in all subjects. When comparing passive apposition of the thumb of the flexor aspect of the forearm, with the highest point, there was a good correlation ($p = 0.003$). Patients under 30 years had higher points in the BS ($p = 0.005$). Arthralgia was present in 29 subjects (70.78%), mainly in women ($p = 0.001$). Flat feet was found in 23 patients (56.09%) and Marfanoid habitus in 23 patients. Mitral valve

prolapse (MVP) was found in 24 patients (58.53%), the great majority under 30 years and with higher points in the BS. Uterine prolapse was diagnosed in 12 women of the 24 examined (50.0%), most of them over 30 years, no significance correlation was showed between uterine prolapse and points in the BS.

Conclusion In our study arthralgias particularly of the knees was found, and among extrarticular MVP was the commonest one. We also found a higher incidence of uterine prolapse than expected for women in this age.

THU0218 SUCCESSFUL TREATMENT OF WEBER CHRISTIAN DISEASE (WCD) WITH THALIDOMIDE IN A PATIENT FAILING MULTIPLE OTHER MEDICAL THERAPIES

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Background

Objectives A case of idiopathic nodular panniculitis (WCD) with recurrent febrile episodes resistant to corticosteroids and methotrexate (MTX) in alternating combin. with hydroxychloroquine (HCQ), azathioprine, cyclosporine, colchicine, and doxycycline is described. Thalidomide 50-100 mg/d induced a remission - now since 2y.

Methods CASE REPORT: The patient is a 65y.o. German male who presented first in 3/1998 with a 2-y. hist. of recur. febr. attacks with 1-2 cm, painful, palp., subc., non-draining, eryth. nodules occur. on lower legs, backside of upper thighs, lower back, and upper arms. 3 consec. biop. showed lobular and septal panniculitis. Lab. exam. revealed mild leukopenia (2500/mm³), mild thrombocytopenia, CRP 2.5 mg/dl, sed.-rate 50 mm/h, ferritin 900 ng/ml. Blood cultures, ANA, ANCA, RF, C3, C4, C1q-Inhib., cardiolip.-AB, immunoelpho., borr. burgd. ser., a1-anti-trypt., serum amyl. and lip., and UA were all normal or neg. Bone marrow biopsy showed unspecific changes. Chest x-ray was normal. Abdominal CT gave no hint for lymphoma or malignancy. Despite 25 mg/d of prednisolone and 20 mg weekly MTX recurrent attacks occurred. Various drugs were tried in combination with MTX in order to lower steroids such as azathioprine 100 mg/d, HCQ 400 mg/d, doxycyc. 200 mg/d, colchicine 1.2 mg/d, and cyclosporine (200 mg/d), all without improvement.

In 3/1999 sympt. worsened and CRP rose intermitt. to 16 mg/dl, further lab. exam. showed persistence of leukopenia 3400/mm³, normal platelet counts, but drop of haemoglobin to 9.5 without clinical evidence of a GI-bleed. A new bone marrow biopsy revealed lymphocytic hyperplasia with no histol. or cytol. support for erythrophagocytosis.

Thalidomide initially 150 mg/d at night, then after 2 weeks at 100 mg/d was started and induced disappearance of febrile attacks and subcutaneous nodules. CPR, sedimentation-rate, and haematological findings normalised. Glucocorticosteroids and MTX were discontinued. After 3 months thalidomide was reduced to 50 mg/d with infrequent mild relapses (with mild subcutaneous nodules only), which disappear within 1-2 day with temporary thalidomide dose increase to 100 mg/d.

Results

Conclusion Rarity of WCD has so far hampered a controlled therapeutic trial with any drug. Only case reports with various drugs exist suggesting mostly corticosteroids, MTX, HCQ, cyclosporine, which all failed in this case. The use of thalidomide in WCD is suggested by clinical similarities to erythema