defects on ventilation-perfusion scan, typical of acute pulmonary embolism (Figure 1). She was initially diagnosed with acute pulmonary embolism and treated empirically with heparin. Subsequently a negative venous Doppler, absence of changes of acute or chronic thromboembolic disease on pulmonary angiogram, elevated ESR of 70 mm/hr, and the typical angiographic changes (Figure 2), led us to make the correct diagnosis of Takayasu's arteritis. Her initial arch angiogram was normal (Figure 3A). She responded to treatment with prednisone, with both clinical improvement and a 50% increase in the calibre of both right and left pulmonary arteries on a MR angiogram done six months later. A repeat pulmonary angiogram 11 months later showed a decrease in pulmonary artery pressures from 65/12 mm Hg to 49/10 mm Hg. She was lost to follow-up and returned six years later with symptoms of dizziness and angiographic evidence of carotid artery involvement (Figure 3B), which required arterial bypass grafting. Histopathological examination of the artery specimen showed patchy acute and chronic inflammation of adventitia, media and intima, focal granulomatous foreign body reaction, and intimal proliferation, changes consistent with Takayasu's arteritis.

Conclusion In young women presenting with a clinical picture of acute pulmonary embolism without any risk factors or a previous history of thromboembolism, pulmonary TA must be considered in the differential diagnosis. A careful evaluation of the clinical features, laboratory findings and angiographic appearance, will often lead to the correct recognition of pulmonary Takayasu's arteritis and the initiation of appropriate therapy.

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AB0092 POLYARTERITIS NODOSA PRESENTING WITH BILATERAL PERIRENAL HEMATOMA IN PATIENT WITH FMF

¹M Çalgüneri, ¹S Apras, ¹MA Öztürk, ¹V Çobankara, ¹S Kiraz, ¹I Ertenli, ²B Çil, ²A Oto. ¹Rheumatology Department; ²Radiology, Hacettepe University School of Medicine, Ankara, Turkey

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Background

Objectives

Methods Case Report: A 17-year old Turkish boy was admitted to our clinic with complaints of weight loss, fatigue and abdominal pain. He had been on colchicine therapy for 6 years with the diagnosis of FMF. On January 2000, he was diagnosed as having acute hepatitis A infection in another centre. Two weeks later he was hospitalised due to severe abdominal pain and abrupt decrease in his haemoglobin values. A right renal subcapsular hematoma was demonstrated by computerised tomography (CT). During follow-up haemoglobin values progressively declined and the hematoma showed enlargement in control CT. Therefore right nephrectomy was performed. After the patient was stabilised he was discharged. However he was again hospitalised with similar complaints 4 days later. This time subcapsular hematoma

was demonstrated in the left kidney and he was referred to our hospital in March 2000. In physical examination, blood pressure was 160/125 mmHg. Laboratory evaluation demonstrated anaemia and elevated erythrocyte sedimentation rate and C-reactive protein. Serological tests for anti-hepatitis B surface antigen, anti-hepatitis A IgG and IgM were positive.

Nephrectomy slides were re-evaluated in our pathology department. Patchy and segmental areas of renal infarct were demonstrated. No amyloid deposits could be noted in the histopathology specimens. Renal angiography demonstrated 2 microaneurysms on intertubular arteries. Together with these findings the diagnosis of PAN was made and the patients was treated with intravenous corticosteroids, intravenous cyclophosphamide and oral corticosteroids together with antihypertensive therapy. He is still being followed-up by our clinic in good health.

Results

Conclusion Perirenal hematoma, which is a rare complication of isolated PAN, have been reported in approximately half of the patients with FMF who developed PAN. The exact pathogenetic relationship of FMF and PAN also is not clear. Immune-related mechanisms have been demonstrated in FMF. It has been proposed previously that infection-related antigenic stimuli could trigger the development of vasculitides. In the present case an usual response to hepatitis A infection and/or previous hepatitis B infection might have been among the triggers of vasculitic process. Additional reports demonstrating direct causal relationship between infectious agents and vasculitic syndromes in patients with FMF will help us to clarify the relationship between the two disorders.

AB0093 CALCIPHYLAXIS DUE TO LONG-TERM STEROID USING IN A PATIENT WITH RHEUMATOID ARTHRITIS

C Korkmaz, E Kasapoglu. *Rheumatology, Osmangazi University, Eskisehir, Turkey*

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Background Calciphylaxis is a rare and life-threatening condition of progressive cutaneous necrosis secondary to small and medium-sized vessel calcification seen almost exclusively in patients with end-stage renal disease and hyperparathyroidism.

Objectives We report a case with calciphylaxis due to long-term steroid using in a patient with rheumatoid arthritis (RA).

Methods Case report.

Results A 43-year-old man admitted with a history of progressive left leg pain and ulcerations. Several hemoragic bullae were present in these areas as well, and the patient offered that the ulcers began as blisters that then broke down. His past medical history included RA and he had been using steroid in doses of 5-40 mg/day according to his complaints for 20 years when he was admitted to our hospital. His examination revealed destructive and erosive arthritis affecting his bilateral wrists, MCP, and PIP joints, and ischaemic necrosis in the distal parts of the 2nd and 3rd digits of the left hand. Laboratory results were as follows: erythrocyte sedimentation rate 77 mm/h, urea 9 mg/dl, creatinine 0.8 mg/dl, calcium 7 mg/dl, phosphate 3.7 mg/dl, albumin 2 gr/dl, parathyroid hormone 43 pg/ml (n = 12-72), protein S 38% (n = 7-115%). Roentgenogram of the lower aspect of the leg showed calcified larger vessels and subcutaneous vessels. His lesions progressed and required above the left knee and left 2nd, 3rd digits amputations. Pathologic examination showed extensive arterial wall calcification. Now he has

been under the treatment of sulfasalazine and he has remained asymptomatic.

Conclusion Calciphylaxis may appear without renal failure and hyperparathyroidism. Long-term steroid using in RA patients may lead to calciphylaxis. Acquired protein S deficiency may contribute to this process.

AB0094 PREVALENCE OF TWIN BIRTHS IN BEHÇET'S DISEASE (BD)

S Masatlıoğlu, F Gödüp, B Yıldırım, R Tunç, I Fresko, H Yazıcı. *Rheumatology, Cerrahpaşa Medical Faculty, Istanbul, Turkey*

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Background

Objectives Studies of twins in BD have been few.^{1,2} Our clinical impression has been that patients with BD had a paucity of twin siblings. We studied the frequency of twin siblings among the BD patients in a controlled protocol.

Methods 1277 patients (773 males and 504 females with a mean age of 34.20 ± 9.59 years) attending a dedicated BD outpatient clinic were asked whether they had a twin sibling, living or dead. Inquiry into monozygosity was made by asking whether this twin (s) looked exactly like the proband as 'two peas in a pod'.³ As controls the same information was sought among i. a group of patients with either rheumatoid arthritis (RA) or systemic lupus (SLE), composed of 180 patients with RA -150 males and 30 females with a mean age of 49.08 ± 14.05 years- and 79 patients with SLE -64 females and 15 males with a mean age of 34.80 ± 12.33 years- all attending a rheumatology outpatient clinic; and ii. among 7761 first year university students -3848 males and 3913 females with a mean age of 18.55 ± 1.38 years- during their university registration. Chi-square test (2_{df}) was used to compare the findings between the three groups.

Results Among the 7761 university students, 120 twins (1.5%) were identified. 92 were dizygotic (1.2%) and 28 (0.38%) monozygotic. Among the 259 patients with connective tissue disease there were 7 (2.7%), all dizygotic twins. Two twins belonged to the SLE and 5 to the RA patients. Among the 1277 patients with BD there were 10 twins (0.08%; $\chi^2_{2df} = 7.17$, $p = 0.03$). Eight were dizygotic and 2 (0.15%; $p > 0.12$) were monozygotic. The monozygotic twins were discordant for BD in either case.

Conclusion The frequency of twin births among our controls was similar to that reported in general for twin-births.⁴ The rather marked decrease, 19 fold when compared to the group of university students and 34 fold when compared to the group of patients with RA or SLE, in the twin frequency among the BD patients could be due to the presence of in utero factors operative in the mothers of patients with BD, unfavourably affecting the successful initiation and/or completion of a twin pregnancy.

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AB0095 INTRAVENOUS IMMUNOGLOBULIN IN LIMITED POLYARTERITIS NODOSA

¹A Balbir-Gurman, ¹D Schapira, ²B Smolin, ²E Volfovitz, ¹AM Nahir. ¹B.Shine Department of Rheumatology, ²Department of Medicine "D", Rambam Medical Center, Faculty of Medicine, Haifa, Israel

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Background Polyarteritis nodosa (PAN) is severe systemic vasculitis that may affect almost all systems. Rarely isolated organ involvement occurs. These cases generally have more benign course and respond well to high dose of steroids. The addition of cytotoxic drugs is controversial. PAN limited to calf muscles is extremely uncommon. Painful tenderness and swelling of dorsal calf muscles, mild fever and general signs of inflammation in blood tests are the only features of this form of vasculitis. ANCA is not obligatory positive. Muscle biopsy is the only way to make the diagnosis.

Objectives

Methods

Results

Abstract AB0095 Table 1

	Case 1 24 M	Case 2 27 F
Calve's pain/tenderness	Yes/Yes	Yes/Yes
Walking difficulties	Yes	Yes
Fever	37.4	37.2
Regional muscle weakness	Yes	No
Haemoglobin (g/dl)	10.8	11.4
ESR (mm/1 h)	85	72
ANCA	pANCA positive	Negative
Prednisone 0.5 mg/d and IVIG 25 g/d for 5 days	10 weeks off steroids	8 weeks off steroids

ESR-Erythrocyte Sedimentation Rate, ANCA-Antineutrophil Cytoplasmic Antigen, pANCA-perinuclear ANCA, IVIG-Intravenous Immunoglobulin.

Conclusion The two herein reported cases (Table 1) of biopsy proven PAN of the calve show that the addition of single course of intravenous immunoglobulin to steroid treatment yielded rapid and stable clinical and laboratory remission with low doses of steroids from the beginning and rapid tapering off.

AB0096 TWO CASES OF BEHCET'S DISEASE WITH ULCERS ON GLANS PENIS

¹T Pirildar, ¹T Turk, ¹E Tunc, ¹V Inal, ¹G Keser, ²A Varol, ¹E Doganavsargil, ¹G Gumusdis. ¹Rheumatology, ²Dermatology, Ege University, Izmir, Turkey

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Background Behcet's Disease (BD), when first described in 1937, was consisted of three symptoms; recurrent oral, genital ulcerations and iridocyclitis. Today, it is known that BD is a multisystemic chronic vasculitic disorder which may involve both arteries and veins of all sizes. Although painful scrotal ulcers healing with scar formation are among the main clinical findings of BD, ulceration of glans penis is unusual.

Objectives

Methods

Results We present two cases of BD with ulcers on glans penis. The first case is a 46-year-old man who presented with