Methods We calculated the SLICC scores (Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index) of 236 patients diagnosed as SLE between 1988 and 2000. Anti-ENA antibodies (U1-RNP, Sm, Ro, and La) were studied by ELISA in the first available serum sample for each patient. The SLE disease activity index (SLEDAI) of the first hospital study was calculated retrospectively.

Results Twenty-five (10.5%) of the 236 patients were men. Age, age at onset of SLE, duration of the disease, and SLEDAI score at the onset of the disease were similar in the men and women. There was no difference in the frequency of anti-ENA antibodies between the sexes. The SLICC cumulative tissue damage score obtained at the last follow-up visit was significantly higher in men (2.24; SD 1.89) than in women (1.40; SD 1.86) (Mann-Whitney $p=0.009$). Kidney disease, whether characterised by clearance of less than 50% of normal (9 of 25; 36% vs 23 of 211; 10.9%) (OR: 4.5) or proteinuria over 3.5 g/day and need for dialysis and transplantation (14 of 25; 56% vs 49 of 211; 23.2%) (OR: 4.2), was significantly more frequent in men (OR: 3.8). Cardiomyopathy (3 of 25; 12% vs 3 of 211; 1.4%) (OR: 9.4), avascular necrosis (3 of 25; 12% vs 3 of 211; 1.4%) (OR: 9.4), and retinal disorders (2 of 25; 8% vs 1 of 211; 0.5%) (OR: 18.2) also were more frequent in men than women. In the follow-up period, 5 of the 25 men died (20%) but only 3 of the 211 women died (1.4%) (Fisher $p=0.005$; OR: 17.33; IC 95%: 3.8–77.9).

Conclusion Mortality and the tissue damage scores were significantly greater in men with SLE than in women. The higher SLICC score was due mainly to the greater frequency of kidney and heart disease.

OP0104 LUPUS NEPHRITIS: CONTRASTING CLINICAL OUTCOME DURING THE LAST DECADE

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10.1136/annrheumdis-2001.188

Background According to Cameron (1999) 5-year renal survival of WHO class IV lupus nephritis (LN) after diagnosis and treatment is now 82%. It is claimed that when terminal renal failure (ERF) supervenes, it is within the first decade of follow-up.

Objectives As our recent experience is different, we evaluated the long-term renal outcome in patients with lupus nephritis treated in our institution in the last decade.

Methods During the last 10 years we followed 44 caucasians (35 females, 9 males, median age 40 years, range 18–73) with biopsy proven LN (WHO class II 5, III 2, IV 31, V 5, and VI 1). 15/44 patients had hypertension, 37/44 patients anti-DNA-ab (Farr test) (median 18, range 8–1493 U/ml), 41/44 positive ANA (median 1280, range 160–40960 titer), 44/44 proteinuria (20/44 with nephrotic proteinuria (NS) and 37/44 hematuria). Median creatinine on admission was 0.9 mg/dl, range (0.3–4.0), median haemoglobin 11.7 g/dl (range 8.2–14.3). All WHO classes were treated with glucocorticoids (1 mg/kg/day for 8–12 weeks, tapering off according to the clinical situation). WHO class IV was initially treated with 3 methylprednisolone boli (then 1 mg/kg/day) and cyclophosphamide (CP) (Austin scheme), sometimes supported by cyclosporine, azathioprine or mycophenolate mofetil. Hypertension and/or NS were treated with ACE inhibitors, hyperlipidemia with statins.

Results No patient died or attained ERF. Median creatinine was 1.1 mg/dl (range 0.5–5.4) after an median observation period of 39 month (range 1–128). Impaired but stable renal function was observed in 12/44 patients (classes III 2, IV 9, V 1). Progression to renal failure was observed in 1 patient with uncontrolled hypertension. From renal histology only classes III and IV had adverse outcomes.

Conclusion We attribute improved outcome of LN in the last decade to earlier detection and therapy of LN and treatment of LN-flares with CP and steroid boli. Moreover better medical management particularly the consequent treatment of hypertension with ACE inhibitors may contribute to a favourable prognosis.

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Vasculitis

AB0082 BEHCET'S DISEASE COMPLICATED WITH CELIAC TRUNK ANEURYSM

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10.1136/annrheumdis-2001.189

Background The aneurysms of the celiac trunk are the rarest aneurysms of the visceral arteries. Spontaneous celiac trunk aneurysm has not been described previously in Behcet's disease.

Objectives A 19-yr-old Turkish man was admitted to our hospital with a history of abdominal pain for 2 weeks. He described recurrent attacks of ulcerations of the oral and genital mucosa, blurred vision and painful lesions on the front of the legs for 2 years. He had had abdominal pain for 6 months, but this symptom had become more troublesome in the last 2 weeks. Physical examination revealed no pathology except for mild abdominal tenderness and deeply pigmented areas on the front of the legs. Ophthalmologic examination showed ocular inflammation and retinal vasculitis. Laboratory evaluations revealed haemoglobin of 14.4 g/dl, white blood cell count of 9650 cells/mm and platelet count of 470000 cells/mm. The erythrocyte sedimentation rate was 61 mm/h, C-reactive protein 11.5 mg/l (normal <0.8 mg/l), and fibrinogen 422 mg/dl (normal: 200–400 mg/dl for male). HLA-B51 is positive. Pathergy test is positive. Celiac trunk aneurysm was detected by computed tomography. A selective angiogram showed dilatation of the celiac artery and an aneurysm with a diameter 4.2 cm.

Methods Visceral artery aneurysm is a rare, but important form of vascular pathology. Aneurysms of splenic artery are the most common. Aneurysms of the celiac artery are uncommon and account for 4% of all splanchnic aneurysms. They can rupture in 2–50% of the cases with a mortality rate of around 80%. The most common etiologic factor of celiac artery aneurysms is atherosclerosis; other causes include infection, trauma, polyarteritis nodosa and fibromuscular dysplasia.

Behcet's disease is an uncommon multisystemic inflammatory disorder characterised by recurrent oral aphthous ulcers, genital ulcers, uveitis, and skin lesions. Involvement of the large vessels is less frequent. Vascular involvement in Behcet's disease is divided into venous and arterial thrombosis and arterial aneurysmal formation. Arterial involvement of Behcet's disease most frequently affects the abdominal aorta, followed by the femoral and the pulmonary arteries.

Results In this case, he fulfils the described criteria for Behcet's disease by the International Study Group for Behcet's disease in 1990. Our diagnosis of Behcet's disease was based mainly on the recurrent oral ulceration, recurrent genital ulceration, eye lesion, skin lesion, and positive HLA-B51 and pathergy test.

Conclusion To our knowledge, the present case is the first patient described with Behcet disease who developed celiac trunk aneurysms. We should be aware of this complication in patients with Behcet's disease who develop abdominal pain.

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AB0083 TAKAYASU ARTERITIS WITH BILATERAL RENAL ARTERY STENOSIS IN A 17-YEAR-OLD GIRL

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10.1136/annrheumdis-2001.190

Background Takayasu arteritis, commonly seen in young women, is a large vessel vasculitis, primarily affecting aorta and its major branches. Hypertension frequently accompanies Takayasu arteritis and in approximately 75% of the cases it is due to renal artery involvement, causing stenosis. Hereby, we report a 17-year-old girl with Takayasu arteritis, having bilateral renal artery stenosis, with unilateral atrophic and afunctional kidney.

This patient, who was diagnosed as Takayasu arteritis 3 years ago, with typical physical findings such as absent upper extremity peripheral pulses and classical angiographical findings involving arcus aorta and its major arteries, has been referred to our rheumatology department because of uncontrolled hypertension. Renal Doppler ultrasound studies and angiography revealed bilateral renal artery stenosis with atrophic right kidney. Successful angioplasty, followed by stent replacement corrected the stenosis in the left renal artery. However, the stenosis in the right renal artery involved a longer segment and angioplasty or stent replacement was not possible. From technical point of view, there was no way for surgical bypass for the stenosis in the right renal artery. Furthermore, no function was observed in the right kidney during renal scintigraphy. Nephrectomy of the right kidney was planned, but it was not accepted by the patient. Since the left renal artery stenosis was corrected, renovascular hypertension maintained by the right renal artery stenosis was treated successfully by captopril and the patient was closely monitored with respect to renal function tests.

In conclusion, renal artery stenosis should always be kept in mind in Takayasu arteritis with hypertension.

Objectives

Methods

Results

Conclusion

AB0084 BEHÇET'S DISEASE AND BUDD-CHIARI SYNDROME: TWO CASES WITH DIVERSE PROGRESSES

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10.1136/annrheumdis-2001.191

Background Behçet's disease (BD) is a systemic vasculitis of unknown aetiology that can lead to thrombosis of both veins and arteries of various sizes. Thrombosis of hepatic veins, known as Budd-Chiari syndrome (BCS) may be seen in BD, with variable clinical courses as described in 2 patients:

Case 1: Thirty-four year-old male patient had oral and genital ulcers and erythema nodosum for 2 years. Within two weeks after presentation, thrombosis of hepatic veins and vena cava inferior was found, while evaluating the patient for abdominal pain and liver function test abnormalities. The patient well responded to the treatment consisting high dose prednisolone, pulse cyclophosphamide and anticoagulant drugs with recanalisation of the veins. At the end of two years, the patient has still been in clinical remission.

Case 2: A twenty year-old male patient had oral and genital ulcerations for 4 years and had an episode of uveitis 4 years ago. He was referred to our clinic because of abdominal distension and oedema of lower extremities lasting for 4 months. The laboratory analyses disclosed ascites, grade III varicosity of oesophagus and thrombosis of hepatic veins and vena cava inferior. Mesoatrial shunt operation was performed when the initial treatment with pulse cyclophosphamide and high dose prednisolone failed. Obliteration of the shunt was observed by doppler ultrasonography, performed one week after the operation. He did not respond to the supportive therapy after the shunt operation and was lost with gastrointestinal bleeding.

Objectives

Methods

Results

Conclusion These two male Behçet's patients with BCS are examples of diverse prognoses, probably depending upon how early the diagnosis is made, and how efficient the treatment is. BCS in BD, may have a better prognosis if the diagnosis is made earlier and the treatment started sooner in the course of the disease.

AB0085 THE EFFECTIVENESS OF MELOXICAM IN TREATING ERYTHEMA NODOSUM SYNDROME

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10.1136/annrheumdis-2001.192

Background

Objectives We used meloxicam at a dose of 15 mg a day for treating erythema nodosum (EN) of various aetiology in 47 patients (44 females and 3 males aged 15-68). Not depending