association between asbestos exposure and risk of CP has been reported. The aim of this study was to investigate the role of occupational and environmental exposure to asbestos, metals, pesticides, silica, organic solvents in CP; we also evaluated the possible interaction with tobacco smoking.

Patients.– We enrolled 90 consecutive patients with CP diagnosed at or referred to our Department from all over Italy between 2004 and 2012; 270 subjects recruited from the general population and matched with the patients for age, sex and geographic origin served as controls. All the study subjects were asked to fill in a questionnaire in order to assess individual risk factors (such as cardiovascular disease, abdominal surgery, drug use and smoking) and occupational and environmental exposure to asbestos, organic solvents, metals, other industrial chemicals, and pesticides.

Results.– Asbestos exposure was over-represented among cases [OR 4.05 (2.28–7.19), P < 0.00001]; exposure to other agents also tended to be more frequent in the patient group, but for none of them statistical significance was reached. Among cases, there was an excess prevalence of smokers [OR 3.16 (1.67–5.99), P = 0.0003]. The pack-year index was also significantly higher in the patient group [median (IQR) 31(20–44) vs. 20(9–37), P < 0.0001]. Interestingly, we found a positive interaction between asbestos exposure and smoking [OR 10.10 (4.28–23.84), P < 0.0000001].

Conclusion.– Exposure to asbestos may be a significant risk factor for the development of CP. In addition, tobacco smoking also increases the risk of CP. Asbestos and tobacco smoking significantly interact and, together, substantially increase the risk of developing the disease.

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A59

Behçet’s disease in Budd-Chiari Syndrome

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Introduction.– Behçet’s disease (BD) is a systemic vasculitis characterized by oral and genital ulcerations, ocular inflammation and venous thrombosis. BD is a well recognized cause of Budd-Chiari Syndrome (BCS). Data are lacking regarding the relationship between BD and BCS.

Patients.– Using a cohort of 111 patients with BCS, we investigated the relationship between BCS and BD in 19 patients with combined diseases and compared the results to 92 BCS patients in whom BD was excluded.

Results.– Median follow-up for the study group (n = 111) was of 53 months (range 1–141 months). BD patients with BCS were younger (P = 0.025), more frequently of male gender (P < 0.0001), originated more frequently from North Africa (P = 0.0002), had higher plasma C-reactive protein level (P < 0.0001) and presented more frequently with inferior vena cava (IVC) obstruction (P < 0.0001), as compared to BCS without BD (n = 92). Among BD patients with BCS, three patients were treated with recanalisation of the vena cava and of the hepatic veins, none patient had a TIPS and one had liver transplantation. Transjugular intrahepatic portosystemic shunt (TIPS) was significantly less performed in BCS patients with BD as compared to those without BD (P = 0.003). Eighty nine percent of BCS patients with BD received corticosteroids and/or immunosuppressive therapy. The 5-years survival rate was of 74% (CI 95% 55–100) and 79% (CI 95% 71–88) in BCS patients with and without BD (P = 0.47), respectively.

Conclusion.– This study shows that despite a higher frequency of additional IVC thrombosis, short-term prognosis of BCS patients with BD does not differ from BCS patients without BD. TIPS is less performed because of technical issues and spontaneous improvement with medical therapy Medical treatment with anticoagulation and immunosuppressive agents seems to be efficient and sufficient to control BCS symptoms.

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A60

The unmet need in Behçet’s disease: Most patients are not in complete remission in the long-term follow-up

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Introduction.– Behçet’s disease (BD) is a systemic disorder with a remitting-relapsing course. As complete remission should be aimed in all inflammatory diseases, we investigated the frequency of complete remission in routine practice in BD.

Methods.– One hundred and thirty patients with BD (F/M: 67/63, mean age: 43.23 ± 11.7 years) classified according to ISG criteria were included, retrospectively. The data for active organ manifestations and treatment protocols were evaluated, both for the current visit and in the last month. Patients having at least one disease manifestations were categorized as active.

Results.– Totally, 857 visits were overviewed. Mean visit number was 6.5 ± 2.7 (1–10), mean follow-up duration was 53.54 ± 41.79 months.

![Figure 1: Clinical activity rates of 10 visits](http://dx.doi.org/10.1016/j.lpm.2013.02.061)
Cutaneous vasculitis: Report of 117 cases

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Introduction.— Cutaneous vasculitis (CV) is defined broadly as inflammation of the blood vessels of the dermis. We retrospectively analyzed a cohort of 117 patients with biopsy-proven CV.

Patients.— We performed a single-centre retrospective review of 117 patients who met the histologic criteria for cutaneous vasculitis between 2003 and 2012.

Results.— The mean age was 55.34 years (15–95) and 65 (55.6%) were females. The main hystopathological pattern was leukocytoclastic vasculitis (65.81%) followed by urticarial (10.25%), lymphocytic (7.6%), neutrophilic (4.2%), nodular (2.5%), granulomatous necrotizing and other type of CV (3.4%). Palpable purpura (65.81%) was the most frequent clinical lesion followed by plaques (17.09%), papules (5.9%) and others (11.11%). Systemic symptoms were observed in 47 cases (41.02%) while 70 cases (59.08%) did not have any symptom. The most common systemic symptom was fever (30.43%) followed by arthritis (18.84%), oral or genital ulcers and uveitis (19.1%), renal disease (8.60%), respiratory symptoms (5.7%) and others (26.08%). Only eight patients showed positive ANCA antibodies, being five MPO positive and three PR3 positive. Forty-nine cases (41.8%) had idiopathic cutaneous vasculitis being the leukocytoclastic type the most frequent. Sixty-eight patients (58.11%) had an aetiological condition. Thirty-four patients (29%) had a systemic autoimmune disease, systemic vasculitis was the most common disease (18), 16 (13.6%) cases were caused by drug reaction and 16 (13.6%) were due to infections. Only two cases had a malignancy cause.

Discussion.— In 58.11% of cases, CV occurred either as part of a primary systemic vasculitis or as secondary vasculitis related to an underlying disease, such as an autoimmune disease, drugs, infections or malignancy. In the remaining 41.8% of cases, CV occurred idiopathically. Leukocytoclastic vasculitis was the most frequent histological pattern observed.

Conclusion.— Cutaneous vasculitis is not one specific disease but a manifestation that can be seen in a variety of settings.

Further reading

A62 Takayasu arteritis – An outcome study in a UK cohort

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Introduction.— Takayasu arteritis is a granulomatous vasculitis affecting the aorta and its branches. Arterial inflammation leads to myofibroblast proliferation, fibrosis, stenoses and aneurysms. Although optimal treatment remains undefined, early diagnosis and treatment may improve outcomes.

Patients.— A longitudinal study of 98 Takayasu arteritis patients seen at Hammersmith Hospital between 2000 and 2012. Eighty-nine percent were female and mean age at diagnosis was 31.5 years.

Results.— Examples of all six TA subtypes were found, and a mean of three arteries involved (range 1–9). The aorta, left subclavian and left common carotid arteries were most commonly affected. Despite 91% of patients displaying “red flag symptoms”, mean delay in diagnosis was 3 years.

18FDG-CT-PET proved most useful for diagnosis. Ultrasound, MR and CT angiography aided diagnosis, with the capacity to identify prestenotic disease. 90% received prednisolone, plus methotrexate 43%, azathioprine 37%, and cyclophosphamide 10%. Biologics (TNFα & IL-6R antagonists) were used in nine patients to control refractory disease. Annual MRA and US monitored outcome and demonstrated that 81.5% had stable disease, 9.8% progressed and in 8.7% lesions improved. Thirty-three and 31 patients respectively underwent surgery or endovascular procedures, with a success rate of 66%. Five pregnancies and 14 live births were recorded and two patients died.

Discussion.— Morbidity due to arterial injury in TA remains high. In our series 18FDG-CT-PET facilitated early diagnosis, while MRA proved effective and was preferred for disease monitoring. Although 60% of patients respond to steroids alone, relapse and side-effect rates are high. We co-prescribed steroids with immunosuppressants to achieve good disease control and steroid-sparing.

Conclusions.— Delayed diagnosis of TA predisposes to severe arterial injury. TA must be considered in young patients with carotidynia, claudication, chest pain, hypertension, dyspnoea, and early immunosuppressive therapy prescribed.

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A63 Biologic agents offer an effective long-term therapeutic option for refractory Takayasu arteritis

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Introduction.— To analyse the efficacy of anti-TNFα and anti-IL-6 receptor (IL-6R) blockade therapy in the treatment of patients with refractory Takayasu arteritis in a large UK cohort.

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