Discussion

Although complete remission is the primary target in rheumatology, it is difficult to achieve complete remission in BD with current therapies. The reluctance of the clinician to be aggressive for some manifestations such as mucocutaneous lesions, might be influencing the continuous, low-disease activity state in BD. Some manifestations such as mucocutaneous lesions, might be influenced by current therapies. The reluctance of the clinician to be aggressive for some manifestations such as mucocutaneous lesions, might be influencing the continuous, low-disease activity state in BD.

Conclusion

The majority of Behcet patients are not in complete remission in long-term follow-up.

http://dx.doi.org/10.1016/j.lpm.2013.02.062

A61

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 histopathological pattern was leukocytoclastic vasculitis (65.81%) followed by urticarial (10.25%), lymphocytic (7.6%), neutrophilic (4.2%), nodular (2.5%), granulomatous necrosis 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 (10.14%), 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 PR-3 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


http://dx.doi.org/10.1016/j.lpm.2013.02.063

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 outlook.

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 yrs.

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%, mycophenolate mofetil 7%, 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%. Fifteen 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.

Conclusion

Delayed diagnosis of TA predisposes to severe arterial injury. TA must be considered in young patients with carotidynia, claudication, chest pain, hypertension, discreet blood pressure or absent pulses, and early immunosuppressive therapy prescribed.

http://dx.doi.org/10.1016/j.lpm.2013.02.064

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.