PP19. EVALUATION OF THE USE OF COMBINED TREATMENT REGIMENS INVOLVING METHOTREXATE AND MYCOPHENOLEATE MOFETIL IN THE SHEFFIELD PAEDIATRIC UVEITIS SERVICE

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Background: A joint paediatric–adult uveitis transition clinic has recently been established in Sheffield involving staff from the Sheffield Children’s NHS Foundation Trust and the Sheffield Teaching Hospitals NHS Foundation Trust. Children and adults with uveitis are managed using similar treatment strategies encompassing a combination of MTX, MMF and biologic treatments. We conducted a service evaluation to describe response to treatment in individuals receiving combination therapy: MTX and MMF. Here we report data from the paediatric service.

Aims: To document the treatment pathway for children and young people with uveitis ultimately receiving triple immunosuppression therapy and to describe response to treatment in terms of disease activity and visual acuity.

Methods: A key word search of a comprehensive ophthalmic electronic clinical letter storage system was conducted. Keywords methotrexate and mycophenolate were used to identify patients who were diagnosed with uveitis at 16 years or younger, and who received combination therapy between April 2012 and April 2014.

Results: Of 64 patients with uveitis, 11 patients (18 affected eyes) met the inclusion criteria: 6 males, 5 females; 6 idiopathic uveitis, 5 JIA-associated uveitis. The mean age at diagnosis was 7 years 7 months (range 3–14 years). Seven patients had bilateral disease, four had unilateral disease. Ten patients followed the same treatment pathway, receiving MTX followed by addition of MMF followed by addition of anti-TNF if required. Combination treatment with MTX and MMF resulted in adequate to achieve disease control in 8/18 eyes. Eight patients (10 eyes) required the addition of anti-TNF to control their uveitis. Seven patients received infliximab, one patient was entered into the Sycamore trial to receive either adalimumab or placebo. 41% of eyes showed a reduction in anterior chamber cells with MTX alone, compared with 76% with MTX plus MMF, and 100% with MTX plus MMF plus anti-TNF. All patients achieved remission on anti-TNF, with anterior chamber cells <1+ in 100% (10/10 eyes) (Table 1). The results present suggesting that presenting visual acuity is a prognostic indicator for the final outcome. Those who presented with vision worse than logMAR 0.4 have a 40% chance of achieving a final visual acuity of logMAR 0.1 or better. Those who presented with vision better than logMAR 0.4 all have a final visual acuity of logMAR 0.1 or better.

Conclusion: Children in our cohort usually followed the same treatment pathway (MTX followed by addition of MMF followed by addition of anti-TNF if required). The addition of MMF to MTX resulted in adequate uveitis control in 44% (8/18 eyes). Addition of anti-TNF to MTX and MMF resulted in disease quiescence in 100% (10/10 eyes) of those who did not achieve quiescence with combination therapy alone.

Disclosure statement: The authors have declared no conflicts of interest.

Table 1 Mean visual acuity according to treatment

<table>
<thead>
<tr>
<th>Condition</th>
<th>Mean visual acuity, logMAR</th>
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<tbody>
<tr>
<td>At presentation 0.365</td>
<td>MTX alone</td>
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<tr>
<td>MTX + MMF 0.212</td>
<td>MTX + anti-TNF</td>
</tr>
<tr>
<td>In those requiring anti-TNF 0.143</td>
<td>MTX + MMF + anti-TNF 0.125</td>
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References

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PP20. BRUISING, BLEEDING AND RECURRENT COMPARTMENT SYNDROME: A CASE OF GARDNER–DIAMOND SYNDROME (AUTOERYTHROCYTE SENSITIZATION SYNDROME)?

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Background: Children are occasionally referred to Paediatric Rheumatology because of bruising or purpura. The differential includes vasculitides and Ehlers Danlos syndrome (EDS) as well as haematological disorders and non-accidental injury.

Aims: To describe the case of a girl who presented with bruising and bleeding leading to recurrent compartment syndrome requiring fasciotomies.

Methods: A 12-year-old girl was referred with a history of spontaneous bruising over her right upper arm. Over the following days she developed progressive right hand swelling and pain resulting from compartment syndrome which required a fasciotomy. She also appeared to develop bruises on her arms after sphygymomanometry and haematuria after abdominal US. Medical history included regional pain syndrome following a right arm injury in a road traffic accident several months before this presentation. Examination showed bruising on both arms and mild knee hypermobility. ANA was positive at low titre. Full blood count and film, extended clotting and platelet aggregation studies, inflammatory markers, dsDNA, ENA, ANCA, complement, aPL antibodies and viral serology were all normal. Magnetic resonance angiography of abdominal aorta, mesenteric and renal arteries was normal. An initial skin biopsy showed minimal inflammation around capillaries but no evidence of vasculitis. Repeat skin biopsy sent for electron microscopy and fibroblast culture, together with DNA for COL3A1 mutations, did not support a diagnosis of vascular EDS [1].

Results: During 3 months following initial presentation, she had seven further fasciotomies for episodes of compartment syndrome affecting the forearms and lower legs. On some occasions, the girl described feeling a pop in a limb followed within minutes by pain and swelling. She was empirically treated with s.c. DDAVP at the start of subsequent episodes. She has continued once-weekly DDAVP and has had no significant bleeding/ bruising in the last 5 months’ follow-up. Gardner–Diamond syndrome (GDS) was postulated after exclusion of other diagnoses. Also called autoerythrocyte sensitization syndrome, GDS is an autoimmune vasculopathy with sensitization to phosphotidylserine in erythrocyte cell membranes [2]. In most cases, the disease develops after psychological stress and is characterized by painful oedematous skin lesions progressing to ecchymoses within 24 h. Cases of GDS in adolescent females and associated with complex regional pain syndrome have been described. The diagnosis is usually based on typical history and positive tests with intracutaneous injections of 80% solution of washed autologous erythrocytes. No effective treatments have been identified.

Conclusion: GDS should be considered in the differential diagnosis of bruising/purpura after excluding haematological and collagen vascular disorders, vasculitides and non-accidental injury.

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References

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