Letters to the Editors

Uveitis and Henoch-Schönlein purpura: case report and literature review

Sirs,

Henoch-Schönlein Purpura (HSP) is the most common vasculitis of childhood (1) and to date only 8 cases of uveitis in HSP have been reported in literature. We describe a child with HSP and anterior uveitis, who attended our Rheumatology Unit.

A 6-year-old female, previously in good health and with a negative family history for autoimmune diseases, received the diagnosis of HSP based on the presence of typical palpable rash at lower limbs and buttocks, bilateral ankle arthritis and haematuria. Two weeks later, she was admitted to our Hospital due to unilateral, painful red eye with photophobia along with a concomitant HSP poussee. Ophthalmological assessment revealed an anterior uveitis of left eye: cellularity 3+, flare 2+, along with perikeratich injection, posterior synechiae and optic disk swelling. Optical coherence tomography (OCT) excluded sign of cystoid macular odema. Blood tests showed increased values of CRP (1.92 mg/dl) and ESR (69 mm/h), with normal complement levels and blood cell count. Urinalysis displayed mild haematuria and proteinuria. Immunoglobulin levels resulted increased: IgG 1850 mg/dl (n.v. 540-1330 mg/dl), IgA 244 mg/dl (n.v. 50-240 mg/dl) and IgM 72 mg/dl (n.v. 50-180 mg/dl). A comprehensive infectious work-up ruled out viral, bacterial and parasitic infections. HLA B27 and B51 typing were negative as well as autoantibody testing, including ANA, ANCA, ASCA and anti-phospholipid panel. Deficit of adenosine deaminase 2 (DADA2) was also excluded. Chest x-ray, abdominal ultrasound and echocardiography were negative. Brain and ocular MRI with gadolinium resulted negative for any central nervous system involvement. Due to the worsening of eye inflammation despite topical treatment, pulse intra-venous methylprednisolone (30 mg/kg/day) for 3 consecutive days was started, and then switched to oral prednisone (2 mg/kg/day). It resulted in a progressive ocular improvement along with skin, nephrological and articular manifestation. Therefore, prednisone was then gradually tapered over 2 months without disease recurrence. To date, at one-year follow-up, no further recurrence of disease has been experienced.

Uveitis is commonly associated with rheumatologic diseases, including Juvenile Idiopathic arthritis, and, more rarely, with vasculitis. Due to the vision loss, it represents a considerable morbidity issue. Our patient fulfilled the HSP-EULAR/PRINTO/PRES criteria (1). To date, only few cases of HSP with uveitis have been reported (2-3). The pathogenesis seems to be related to the increased levels of abnormally glycosylated serum IgA1, generated by the alternative complement activation. IgA1 containing circulating immune complexes may reach eye circulation inducing inflammation by leukocytes immigration (2-3).

We additionally performed a systematic review on Pubmed, Medline and Embase up to 30th September 2019 retrieving an association between HSP and uveitis and we found other additional 8 cases: three of them belonging to paediatric age. Clinical characteristic of these patients, are reported in Table I (2, 4-10). All of them showed cutaneous and articular involvement, whilst nephrological involvement in all cases except one. In 6/9 patients, uveitis was anterior, posterior in 3. In 8 patients uveitis was symptomatic with pain, redness, photophobia or blurred vision; in one patient this datum was not extractable. The main treatment was corticosteroid, administered topically, orally or intravenous.

Although rare, the presence of ocular involvement can occur during HSP. Notable, HSP associated uveitis is symptomatic. Renal involvement might be a risk factor for the development of eye involvement as in IgA nephropathies: conversely to HSP patients without eye disease, the rate of renal involvement appears more frequent, even of variable severity. However, besides the paucity of available literature, as Kaur *et al*. suggested (2), uveitis onset in HSP should not *a priori* exclude that diagnosis, as well ophthalmological examination might be part of evaluation in routine HSP setting in order to early identify and timely treat eye disease.

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Competing interests: none declared.

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Table I. Summary of 9 HSP patients with recurrent uveitis.

	Yamabe *et al*. 1988	Wu *et al*. 2002	Muqit *et al*. 2005	Erer *et al*. 2010	Kaur *et al*. 2012	Nicholson *et al*. 2013	Patheja *et al*. 2015	Ozlu *et al*. 2016	Our patient
Age	64 years	6 years	42 years	39 years	11 years	24 years	21 years	8 years	6 years
Rash	Y	Y	Y	Y	Y	Y	Y	Y	Y
Joint involvement	N	Y	Y	Y	Y	N	N	Y	Y
Nephritis	Y	Y	Y	Y	N	Y	Y	Y	Y
Abdominal pain	N	N	N	N	N	Y	N	Y	N
Anatomic subtype uveitis	Anterior	Posterior: central retinal artery occlusion retinal oedema	Anterior and punctate keratitis, corneal epithelial erosion	Anterior	Anterior	Posterior: macular oedema and cotton wool spot	Posterior: macular oedema/cotton wool spot	Anterior	Anterior
Bilaterality	N	Y	N	Y	Y	Y	Y	Y	N
Granulomatous	N	N	Y	N	N	N		N	N
Uveitis symptoms	NA	Visual loss	Pain	Photophobia and red eye	Pain	Blurred vision	Blurred vision	Pain and redness	Pain and redness
Recurrence of symptoms	Y	NA	N	Y	Y	N	N	N	N
Treatment	NA	Oral Prednisolone	Oral Prednisolone	P + cyclo-phosphamide	Oral Prednisolone	Oral Prednisolone	Steroids	P + Azathioprine	Metil-P pulses + oral Prednisolone

Y: yes; N: no; P: predinisolone; NA: not available.