For the Eye Altering, Alters the All

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Case history

A 26-year-old African female presented initially with **chronic dacryoadenitis,** resistant to both topical and systemic treatment with short courses of antibiotics and corticosteroids**.** Screening for underlying systemic condition revealed **nephrotic range proteinuria (**2,72g/24u)**.**

Diagnosis

Renal biopsy confirmed diagnosis of **lupus nephritis class V. O**phtalmic exam showed **active vasculitis** in both eyes. Clinical and diagnostic features (joint pain, dacryoadenitis, retinal vasculitis and lupus nephritis) in combination with serology (positive anti-DNA antibodies, hypocomplementemia) confirmed **diagnosis of systemic lupus erythematosus.** Index for activity and severity (BILAG and SELENA/SLEDAI) showed a BILAG A for renal/neurological/muco-cutaneous, a B for musculo-skeletal with a SELENA-SLEDAI index of 24 points.

Management and outcome

Treatment was immediately initiated with **glucocorticosteroids (GCS), mycophenolate mofetil (MMF) and Hydroxychloroquine sulphate (Plaquenil®). Tacrolimus** was associated but no effect was observed with the proteinuria remaining in nephrotic range and secondary effects of the glucocorticosteroids becoming a real concern. Patient was started on **add-on belimumab** with quasi-immediate effect on the proteinuria, making it possible to reduce the dose of the other immunosuppressants and gradually stop them, even the GCS.

Discussion

Whether belimumab has a role in the treatment of CNS lupus or active lupus nephritis is still unknown, since these patients were excluded from the **BLISS trials.** However in those trials, 267 patients had renal involvement and treatment in this group was associated with significant improvement in proteinuria. A phase 3 study (**BLISS-LN**) is recruiting patients with lupus nephritis since these post-hoc analysis of the BLISS trials suggest that belimumab may offer renal benefit in patients with SLE.

Biopsy results

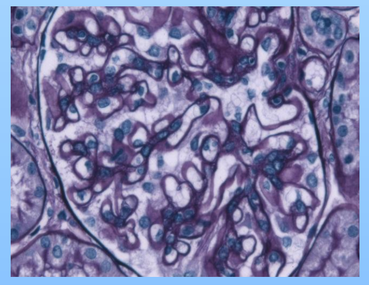

Fig 1: light microscopy: PAS staining: normal structure of the glomeruli, no proliferation, thickening of the basal membrane due to subepithelial immune deposits

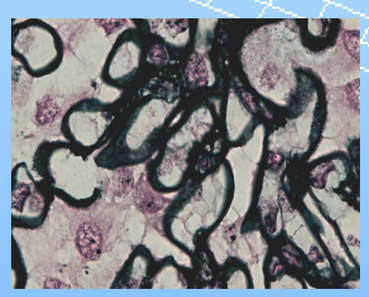

Fig 2: light microscopy, silver staining : clear visualisation of the thickened basal membrane and appearance of ‘spikes’ (formation of perpendicular projections of material similar to the glomerular basement membrane (GBM) in the external part of this one (between podocyte cytoplasm and GBM)

Treatment schedule

date	proteinuria	serum creatinine	GCS	Cellcept	tacrolimus	belimumab
27-12-12	2,72 g/24u	0,63 mg/dl	50 mg	1000->3000		
11-01-13	4,23 g/24u	0,82 mg/dl	50 mg	3000 mg		
7-02-13	3,72 g/24u	0,81 mg/dl	50 mg	3000 mg		
27-02-13	6,92 g/24u	0,75 mg/dl	50 mg	3000->1500	3mg-> TDM	
27-03-13	3,03 g/24u	0,76 mg/dl	40->20	1500 mg	6mg-> TDM	start
24-04-13	1,93 g/24u	0,78 mg/dl	20->15->10	1500 mg	6mg	640mg IV/m
6-06-13	0,19 g/24u	0,83 mg/dl	10->8	1500->1000	6mg	640mg IV/m
10-07-13	0,12 g/24u	0,9 mg/dl	8->6	1000->500	6mg	640mg IV/m
7-08-13	0,11 g/24u	0,93 mg/dl	6->5	500 mg	6mg	640mg IV/m
11-09-13	0,07 g/24u	-	5->4	STOP	5mg	640mg IV/m
10-10-13	0,07 g/24u	1,07 mg/dl	4->3	STOP	5mg	640mg IV/m
13-11-13	0,07 g/24u	-	3->2	STOP	5mg	640mg IV/m
19-12-13	0,07 g/24u	1,08 mg/dl	4 mg	STOP	5,5mg	640mg IV/m
7-01-14	0,15 g/24u	1,8 mg/dl	4 mg	STOP	STOP	640mg IV/m
17-01-14	0,29 g/24u	1,18 mg/dl	4 mg	STOP	STOP	640mg IV/m
4-02-14	0,42 g/24u	0,9 mg/dl	4 mg	STOP	STOP	640mg IV/m
24-02-14	0,39 g/24u	0,89 mg/dl	4->3	STOP	STOP	640mg IV/m
4-07-14	0,12 g/24u	0,91 mg/dl	3->2	STOP	STOP	640mg IV/m
10-09-14	0,07 g/24u	0,91 mg/dl	2 mg	STOP	STOP	640mg IV/m
8-10-14	0,16 g/24u	0,91 mg/dl	2/2d	STOP	STOP	640mg IV/m
17-12-14	0,13 g/24u	0,77 mg/dl	4mg/w	STOP	STOP	640mg IV/m
25-02-15	0,1 g/24u	0,81 mg/dl	STOP	STOP	STOP	640mg IV/2m

Conclusion

This case shows excellent results of belimumab in lupus nephritis with persistent nephrotic range proteinuria under conventional treatment. The bilateral ocular vasculitis, resistant to the initiated treatment, also responded to the initiation of belimumab but remission was only obtained after 6 months. Alternatives are scarce and mostly limited due to toxic effects and by failure to control disease.

References

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