Rituximab in pemphigus patients: A South Auckland perspective

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Background

Pemphigus is a rare immunobullous disorder of the skin and mucous membrane.[1] First line treatment for pemphigus has previously relied on high dose corticosteroids, and/or steroid sparing agents.[2] Rituximab is a chimeric monoclonal antibody against CD-20. It is the first biologic to be FDA approved, and it has superior efficacy over prednisone monotherapy for the treatment of severe pemphigus vulgaris.[3] It is recommended as first line for severe pemphigus in some guidelines. It is not funded in New Zealand for pemphigus but it is being considered by Pharmac.

Methods

A retrospective case series was carried out on patients who received rituximab under NPPA at Counties Manukau DHB. Patient data from 1st Jan 2003 to 31st December 2020 was collected.

Patient demographics (age, ethnicity, gender) and disease characteristics (skin biopsy, autoantibodies) were documented. Outcome after infusion was collected for up to 3 years from infusion (if available). These included serum antibody levels before and after treatment, any side effects of treatment, and any hospital admissions or mortality.

Figure 1: Clinical photographs of patients with pemphigus foliaceus (a), (b) and pemphigus vulgaris (c), (d)

Results

Five patients received rituximab at CMDHB. Four patients had pemphigus vulgaris and one pemphigus foliaceus. All had histologic, skin autoantibody and direct immunofluorescence proven diagnoses. Patient and disease characteristics are as Table 1.

All patients received rituximab using the protocol of 1000mg IV week 0 and 3. One patient received an additional 500mg IV.

All patients had recorded clinical benefit after treatment. Two patients had a documented reduction in skin autoantibody levels after treatment. Two patients subsequently moved out of Auckland. One has been lost to follow up with no further admissions. Two remain in Dermatology outpatient care on prednisone and prednisone with dapsone respectively.

Table 1: Pemphigus patient and disease characteristics, rituximab treatment and outcome at CMDHB

	Case 1	Case 2	Case 3	Case 4	Case 5
Age at Dx	14	25	49	44	66
Ethnicity	Fiji Indian	PI	Maori	Fiji Indian	Maori
Gender	M	F	M	F	M
Autoab titres at Rx	1:1280	1:1280	1:2560	NA	1:1280
Disease duration at Rx	8 months	3 years	1 month	8 years	2 months
Date of Rx	2018	2019;2020	2014	2014	2019
Adverse effects	Nil	Nil	Nil	Nil	Nil
Follow up duration	3 years	1 year*	3 years	3 years	1 year**
No of admissions					
-Before	3	38	2	0	1
-After	0	5 ; 0+	0	0	0

*lost to follow up; **moved DHB
+ no admissions since 3rd additional dose of rituximab
Autoab = skin autoantibodies; Dx = diagnosis; Rx = treatment

Conclusion

All five patients had significant clinical improvement with rituximab treatment for severe mucocutaneous pemphigus. The benefits were a rapid clinical response, early reduction of prednisone dose reduced hospital admissions. This rare skin condition could benefit from funded treatment in New Zealand to reduce morbidity.

References

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3.Joly P, Maho-Vaillant M, Prost-Squarcioni C et al. First-line rituximab combined with short-term prednisone versus prednisone alone for the treatment of pemphigus (Ritux 3): a prospective, multicentre, parallel-group, open-label randomized trial. Lancet. 2017;389:2031-40.