

Successful IVIg-treatment of myelofibrosis developing in a patient with primary Sjögren's syndrome. A case report

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A man born 1927, without heredity for connective tissue or hematological diseases, developed in 1981 bilateral (most left-sided) swelling and enlargement of the parotid glands. Some discomfort and permanent mumps looking face due to the chronic parotitis followed. Shortly thereafter progressing symptoms of dry mouth and dry eyes started.

When first seen at SSRC Feb 1982 he furthermore complained of purpura hypergammaglobulinaemia Waldenström, dryness in the upper respiratory tract and slight tiredness with 8.5 h sleeping/resting time/day. Three different objective functional tests of both the lacrimal glands and the salivary glands were all abnormal. The ANA was negative, while both anti-SSA and -SSB ab were positive. P-IgG = 26.0 g/l and IgM-RF = 2.560. Blood cell counts were all within normal range.

Severe ongoing generalised urticarial-vasculitis started 1994/95 followed by decreasing hemoglobin and thrombocytes ($<100 \times 10^9/l$). There was no peripheral lymphoma or splenomegaly while a bone marrow aspiration showed no signs of malignant lymphoma or foreign body cells but changes compatible with myelofibrosis.

Treatment with blood transfusions, s.c. erythropoetin at various concentrations and prednisolone were initiated without any improvement in the clinical situation. Therefore we decided to try IVIg according to a previous report (Lupus 1997: 6; 408). The patient received 3 courses of IVIg 3 weeks apart. Each course consisted of 2 mg/kg IVIg given over 2 days.

	960410	960507	960604	960805
	Before 1st	Before 2nd	Before 3rd	After 3rd
Hemoglob:	83	93	89	108
Thromboc:	56	81	80	175

Nov 1998 the patient was still doing rather well. The myelofibrosis is treated with prednisolone 2.5 mg/day; iron 50 mg/day and erythropoetin 20.000 IU s.c./week. The Sjögren symptoms have been rather stable even after the IVIg procedures and are treated with bromhexine, gammalinolenic acid and ocuguttae.