

## Supplementary material

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Table S1 – Details of treatment.

Abbreviations used in the table:

PG- pyoderma gangrenosum

CsA – cyclosporin A

IVIg – intravenous immunoglobulins

MTX – methotrexate

AZA -azathioprine

CFS – cyclophosphamid

IFX – infliximab

ADA - adalimumab

Symptoms of the disease	Treatment (dose per day if not mentioned otherwise)	Treatment duration	Result of treatment	Remarks
Diffuse skin ulcerations, multiple lung infiltrates, fever, elevated inflammatory markers	Prednisone 0,6 mg/kg	11.2008	No response - progression of skin and pulmonary lesions	
As above - deteriorating	Prednisone 0,6 mg/kg Cyclosporin A 3,3-5,0 mg/kg	12.2008	No response - progression of skin and pulmonary lesions	
As above - deteriorating	Prednisone 0,6 mg/kg Cyclosporin A 5,0 mg/kg Intravenous immunoglobulins 2g / kg / pulse (over 4 days)	12.2008	Partial remission	
Residual skin and lungs lesions	Prednisone 0,6 mg/kg Cyclosporin A 5,0 mg/kg	12.2008-03.2009	Partial remission sustained	Development of arterial hypertension > cyclosporin A reduction to 1,7

				mg/kg
Skin and lungs lesions deterioration	Prednisone 0,6 mg/kg Cyclosporin A 1,7 mg/kg Infliximab 5mg/kg (0,2,6 weeks > every 8 weeks)	04.2009 – 07.2009	Skin lesions healed within a month, pulmonary changes within 3 months.	Prednisone was gradually reduced to 0,25 mg/kg
Active cutaneous lesions	Prednisone 0,25mg/kg Cyclosporin 3,3 mg/kg Infliximab 5mg/kg every 8 weeks	08.2209 – 10.2009	No significant response – skin lesions active	
As above	Prednisone 0,25mg/kg Cyclosporin 3,3 mg/kg Infliximab 5mg/kg infusion after 6 instead of 8 weeks	10.2009-11.2009	No significant response – skin lesions active	
As above	Prednisone 0,25 mg/kg Cyclosporin 3,3 mg/kg Infliximab 5mg/kg every 6 weeks Methotrexate 7,5mg > 20mg/week	12.2009 – 04.2010	Gradual regression with methotrexate dose increase; Skin lesions - constantly minimally active	Methotrexate gradually increased to 20 mg/week as cyclosporin gradually withdrawn
Minimally active skin lesions	Prednisone 0,25 mg/kg Infliximab 5mg/kg every 6 weeks Methotrexate 20mg/week	04.2010	Minimal activity of skin lesions	Osteoporhosis > prednisone cessation + supplementary hydrocortisone therapy; methotrexate dose increased
Minimally active skin lesions	Infliximab 5mg/kg every 6 weeks Methotrexate 25mg/week	04.2010-05.2010	Minimal activity of skin lesions	Skin lesions deterioration 6 weeks after prednisone cessation > prednisone reintroduced
Active skin lesions	Infliximab 5mg/kg every 6 weeks Methotrexate 25mg/week Prednisone 0,5 mg/kg	05.2010-09.2010	Complete remission	
None	Infliximab 5mg/kg every 8 weeks Methotrexate 25mg/week Prednisone 0,25 mg/kg	10.2010-04.2011	Complete remission	
None	Infliximab 5mg/kg every 10 weeks	05.2011-02.2012	Complete remission	Infliximab cessation in

	Methotrexate 25mg/week Prednisone 0,25-0,20 mg/kg			02.2012
None	Methotrexate 25mg/week Prednisone 0,2 mg/kg	03.2012-06.2012	Complete remission	05.2012 Cataract diagnosis
Single skin lesion	Methotrexate 25mg/week Prednisone 0,5 mg/kg	07.2012-08.2012	Minimal activity of skin lesion	08/09.2012 - Pneumonia and urinary tract infection due to Escherichia coli ESBL (+) > antibiotic therapy; MTX cessation > disease flare
Active skin lesions	Prednisone 0,5 mg/kg Mycophenolate mofetil 2g/d Co-trimoxazole 320/1600mg/d	09.2012-11.2012	Skin lesions slowly deteriorating	11.2012 MMF cessation
Active skin lesions	Prednisone 0,5 mg/kg Co-trimoxazole 320/1600mg/d Etanercept 50mg/week	12.2012-02.2013	Only temporary remission	06.02.2013 Etanercept cessation
Active skin lesions	Prednisone 0,5 mg/kg Co-trimoxazole 160/800mg/d Infliximab 3,3mg/kg (induction 0,2,6 weeks > every 8 weeks)	02.2013-04.2013	Partial remission	
Deterioration of skin lesions	Prednisone 0,5 mg/kg Infliximab 5 mg/kg every 8 weeks Cyclosporin A 5mg/kg Co-trimoxazole 320/1600mg/d (cessation 07.2013)	05.2013-01.2014	Partial remission	11.2013 Monoclonal gammopathy of unknown significance 01-04.2014 Polycystic lesion in perianal tissue > infliximab tapered > CT scan + laparotomy > cystis serosa multilocularis – benign lesion
Moderately active skin lesions	Prednisone 0,5 mg/kg Cyclosporin A 5mg/kg Dapsone 25-100mg/d	02.2014-04.2014	Progression of skin lesions	04.2014 CsA nephropathy with creatinine increase up to 2x upper limit of the norm > CsA

				cessation
Active skin lesions	Prednisone 0,5 mg/kg Dapsone 100mg/d	04.2014- 05.2014	Gradual progression of skin lesions	
Active skin lesions	Prednisone 0,5 mg/kg Infliximab 5mg/kg (0,2,6 weeks > every 8 weeks)	06.2014	Only temporary regression	
Numerous, active skin lesions	Prednisone 0,5 mg/kg Infliximab 5mg/kg every 8 weeks Azathioprine 200mg/d	07.2014	No response	Severe anaemia due to azathioprine > azathioprine cessation
Numerous, active skin lesions	Prednisone 0,4 mg/kg Infliximab every 8 weeks Co-trimoxasole 160/800mg/d	08.2014	Deterioration of skin lesions	08/09.2014 Infliximab cessation
Numerous, very active skin lesions	Prednisone 0,85 mg/kg Co-trimoxasole 320/1600mg/d Intravenous immunoglobulins 3g/kg/puls (over 4 days) Cyclophosphamid pulse 1g	09.2014	Partial regression	
Numerous, active skin lesions	Prednisone 0,85 mg/kg Intravenous immunoglobulins 2 g/kg/puls (over 3 days) Cyclophosphamid pulse 1g	10.2014	Slow regression of skin lesions	Severe anaemia after 2nd puls of CFS (haemoglobin 6,6 g/dl) > CFS cessation, red blood cells concentrate transfusion
Active skin lesions	Prednisone 0,85 mg/kg Intravenous immunoglobulins 2-3g/kg/pulse (over 3-4 days) – pulses monthly	11.2014- 05.2015	Moderate improvement of skin lesions	Severe pain of skin lesions > Phentanyl transdermally
Active skin lesions	Prednisone 0,85 mg/kg Intravenous immunoglobulins 2-3g/kg/puls (over 3-4 days) – pulses monthly Methotrexate 15-20mg/week	06.2015- 07.2015	No further improvement of skin lesions (lesions stable but active)	07/08.2015 IVIg tapered
Moderately active skin lesions	Prednisone 0,85 mg/kg Methotrexate 20mg/week Adalimumab 40mg / 2 weeks	08.2015- 09.2016	Spectacular effect – complete remission of skin lesions	Prednisone was gradually reduced to 0,25 mg/kg 09/10.2016 Productive cough, rales and fever after

				6 weeks of therapy – suspected to be ADA – induced pneumonia > antibiotic therapy was induced and ADA withdrawn but pulmonary symptoms deteriorated > CT scan > active PG lesions in lungs, spleen and mesorectum.
Active lungs, spleen and mesorectum lesions	Prednisone 0,25 mg/kg Methotrexate 20mg/week Adalimumab 80mg / 2 weeks	01.2017- 02.2017	Rapid regression of symptoms and healing of visceral lesions	
None	Prednisone 0,25 mg/kg Methotrexate 20mg/week Adalimumab 40mg / week	03.2017- 08.2017	Complete remission	08.2017 Cough with hematoptysis > Pneumocystis jirovecii infection was suspected > received co- trimoxazole, adalimumab was tapered > CT scan, bronchofiberoscopy , BALF and bacteriological examination > infection was excluded > symptoms due to minimally active pulmonary PG lesions
Cough and hematoptysis	Prednisone 0,25 mg/d Methotrexate 20mg / week Adalimumab 40/week Co-trimoxazole 320/1600mg/d	08.2017- 09.2017	Complete remission of skin lesions Constantly minimally active pulmonary lesions	Co-trimoxazole therapy was maintained due to combined immunosuppressive therapy (P. jiroveci prophylaxis)
Cough and hematoptysis	Prednisone 0,85 mg/d Methotrexate 20mg / week Adalimumab 40/week Co-trimoxazole	09.2017- 02.2018	Complete remission	Prednisone was gradually reduced to 0,25 mg/kg 02.2018 Urinary tract

	320/1600mg/d			infection + moderate anaemia – adalimumab was temporarily withdrawn during antibiotic therapy > due to no signs of flare at that time – ADA cessation
None	Prednisone 0,25 mg/d Methotrexate 20mg / week	03.2018-06.2018	Complete remission	
Active skin lesions, cough, hematoptysis	Prednisone 0,25mg/d Methotrexate 20mg / week Adalimumab 40mg/week	07.2018 - now	Complete remission	