

Table SI. Clinical data and changes of some complement and coagulation component levels between 2006–2010. Normal ranges are given in parentheses

Dates	Clinical data	APTT (24–37s)	fVIII:C (0.5–2.0 kIE/l)	C1inh function (70–130%)	C1q (78–131%)	C1inh level (72–153%)	fXII (0.70–1.50 IU/l)	Bradykinin (median 11 pg/ml ^a)
July 2005	Pericarditis							
Oct. 2005	Bruise-like skin symptoms							
June 2006	Diagnosis acquired haemophilia	93	0.04				0.57	
Nov. 2006	Angioedema of hands, palpable purpura, skin biopsy: leucocytoclastic vasculitis		0.71		149	142		
Feb. 2007	Angioedema of face and neck, urticarial vasculitis		1.21		125	137		
March 2007	Angioedema of upper airways, face, hands, urticarial vasculitis	38		135	168	185		
April 2007	Angioedema, urticarial vasculitis	36	1.69		143	180	0.56	1,680
May 2007	Improved symptoms	32						960
July 2007	Remission	28						279
March 2008	Remission			126	126			
Feb. 2010	Treatment tapered out		0.74				0.76	
Feb.–March 2010	Angioedema of face, urticarial vasculitis	34					0.56	
April 2010	Angioedema of face, hands, upper airways, urticarial vasculitis	42	1.79		143	180	0.71	
May 2010	Angioedema of face, hands, upper airways, urticarial vasculitis		1.54		194		0.63	
June 2010	Remission	36			96	151		

Numerous values of C3 and C4 were normal during both disease and remission.

^aApproximate values based on a paediatric study (11), median 11, range 1–304.

fVIII:C; factor VIII; C1inh function: C1 inhibitor function; fXII: factor XII. Bradykinin was measured according to (11, 12).

APTT: activated partial thromboplastin time.

Abnormal values are in bold.