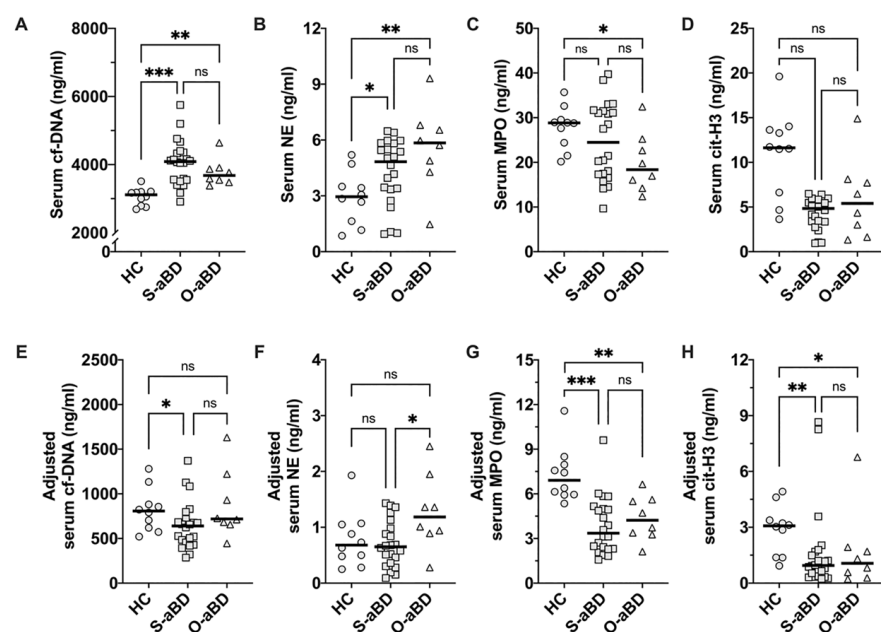


Supplementary Table S1. Characteristics of systemically and isolated orally active patients.

	Systemically active BD (n=22)	Isolated orally active BD (n=8)
Sex, male, n (%)	16 (72.7)	5 (62.5)
Age, year, mean $\pm$ SD	34.8 $\pm$ 10.1	36.8 $\pm$ 12.9
Disease duration, m, med (IQR)	48 (9-180)	73 (53-119)
Family history of BD, n (%)	4 (18.2)	2 (25)
HLA-B51, n (%)	7 ^y (77.8)	3 ^z (75)
Smoking, n (%)	7 (31.8)	1 (12.5)
Pathergy positivity, n (%)	15 ^s (78.9)	4 ^e (57.1)
Cumulative involvement, n (%)		
Mucocutaneous	22 (100)	8 (100)
Arthritis	9 (40.9)	3 (37.5)
Uveitis	7 (31.8)	1 (12.5)
Neurologic	2 (9.1)	0 (0)
GIS	1 (4.5)	0 (0)
Vascular	7 (31.8)	0 (0)
Other	1 (4.5)	0 (0)
Treatments, n (%)		
Naïve	5 (22.7)	1 (12.5)
Colchicine	16 (72.7)	7 (87.5)
Azathioprine	6 (27.2)	1 (12.5)
Corticosteroid	3 (13.6)	0 (0)
TNFi	0 (0)	0 (0)
Laboratory values		
Neutrophil, 10 ³ /μl, med (IQR)	6.6 (5.3-8)	5.0 (4.0-5.6)
CRP, mg/l, med (IQR)	10.75 (4.28-46)	6.7 (1.9-14.77)
ESR, mm/h, med (IQR)	11.5 (8-38)	7.5 (5.5-17)

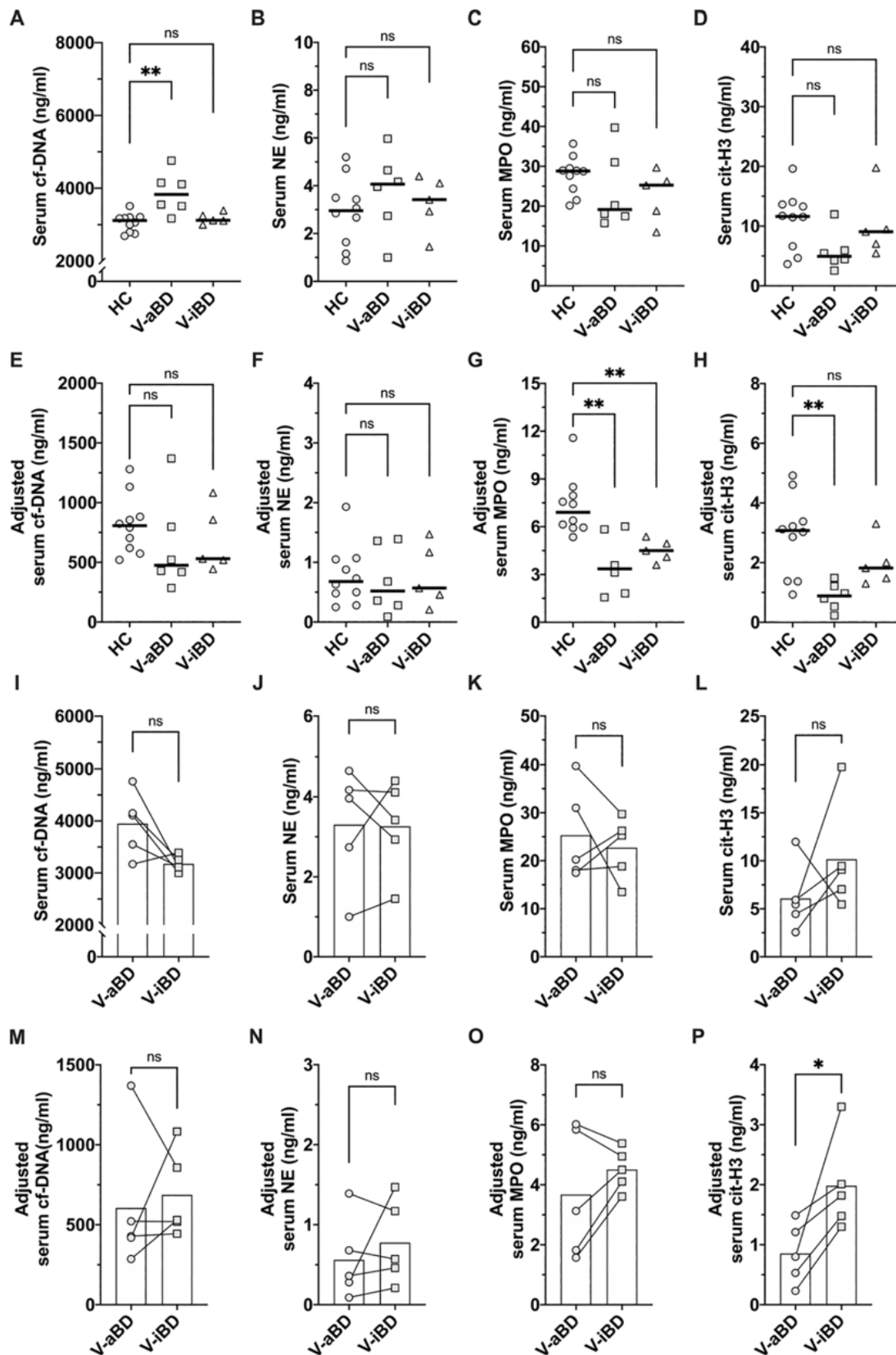
BD: Behçet's disease; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; GIS: gastrointestinal system; IQR: interquartile range; m: month; med: median; n: number of patients; SD: standard deviation; TNFi: tumour necrosis factor inhibitor.

(^y n=9, ^z n=4, ^s n=19, ^e n=7).

**Supplementary Fig. S1.** Unadjusted serum NETosis markers (A, B, C, and D) and adjusted serum NETosis markers (E, F, G, and H) in HC, systemically aBD and isolated orally aBD. Mann-Whitney *U* test; ns=non-significant.

* $p < 0.05$, ** $p < 0.01$, *** $p < 0.001$.

S-aBD: systemically active Behçet's disease; O-aBD: isolated orally active Behçet's disease; HC: healthy control.



Supplementary Fig. S2. Unadjusted serum NETosis markers (A, B, C, and D) and adjusted serum NETosis markers (E, F, G, and H) in HC, V-aBD, and V-iBD. Mann-Whitney *U* test; ns=non-significant, ** $p < 0.01$. Alteration in unadjusted serum NETosis markers (I, J, K, and L) and adjusted serum NETosis markers (M, N, O, and P) during longitudinally follow-up among the patients with vascular involvement. Wilcoxon signed-rank test; ns=non-significant.

* $p < 0.05$.

V-aBD: vascular active Behçet's disease; V-iBD: vascular inactive Behçet's disease; HC: healthy control.

Supplementary Table S2. Correlation analyses between NETosis markers and laboratory findings in the patients with BD.

	Neutrophil count		CRP		ESR	
	r	p [‡]	r	p [‡]	r	p [‡]
Unadjusted serum cf-DNA	0.300	0.039	0.558	<0.001	0.343	0.017
Unadjusted serum NE	-0.196	NS	-0.042	NS	-0.042	NS
Unadjusted serum MPO	0.313	0.030	-0.164	NS	-0.189	NS
Unadjusted serum cit-H3	0.051	NS	-0.206	NS	-0.220	NS
Adjusted serum cf-DNA	n/a	n/a	-0.036	NS	0.007	NS
Adjusted serum NE	n/a	n/a	-0.218	NS	-0.186	NS
Adjusted serum MPO	n/a	n/a	-0.380	0.008	-0.307	0.034
Adjusted serum cit-H3	n/a	n/a	-0.344	0.017	-0.314	0.030
Saliva cf-DNA	0.427	0.047	0.552	0.008	0.203	NS
Saliva NE [‡]	0.077	NS	0.231	NS	0.212	NS
Saliva MPO	-0.224	NS	0.232	NS	-0.123	NS
Saliva cit-H3	-0.167	NS	0.061	NS	-0.133	NS

[‡] Spearman's *rho*. [‡] n=13.

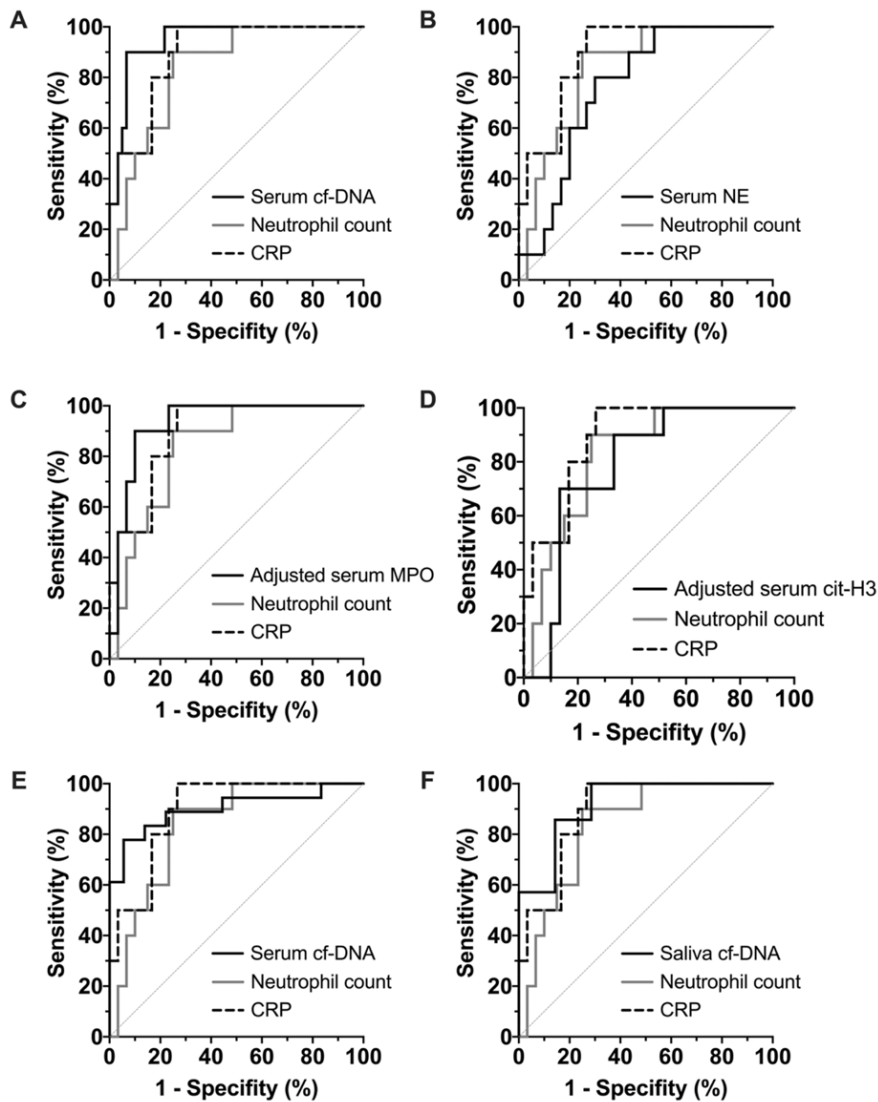
cf-DNA: cell-free DNA; cit-H3: citrullinated histon-3; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; MPO: myeloperoxidase; NE: neutrophil elastase; n/a: non-applicable; NS: not significant; r: correlation coefficient.

Supplementary Table S3. Correlation analyses between NETosis markers in the patients with BD.

		r	p [‡]
Unadjusted serum cf-DNA	Unadjusted serum NE	-0.067	NS
Unadjusted serum cf-DNA	Unadjusted serum MPO	0.076	NS
Unadjusted serum cf-DNA	Unadjusted serum cit-H3	0.022	NS
Unadjusted serum NE	Unadjusted serum MPO	0.003	NS
Unadjusted serum NE	Unadjusted serum cit-H3	-0.021	NS
Unadjusted serum MPO	Unadjusted serum cit-H3	0.457	0.001
Adjusted serum cf-DNA	Adjusted serum NE	0.711	<0.001
Adjusted serum cf-DNA	Adjusted serum MPO	0.572	<0.001
Adjusted serum cf-DNA	Adjusted serum cit-H3	0.367	0.010
Adjusted serum NE	Adjusted serum MPO	0.626	<0.001
Adjusted serum NE	Adjusted serum cit-H3	0.366	0.010
Adjusted serum MPO	Adjusted serum cit-H3	0.550	<0.001
Saliva cf-DNA	Saliva NE [‡]	0.275	NS
Saliva cf-DNA	Saliva MPO	0.378	NS
Saliva cf-DNA	Saliva cit-H3	0.151	NS
Saliva NE [‡]	Saliva MPO	0.011	NS
Saliva NE [‡]	Saliva cit-H3	0.203	NS
Saliva MPO	Saliva cit-H3	0.266	NS
Unadjusted serum cf-DNA	Saliva cf-DNA	-0.049	NS
Unadjusted serum NE [‡]	Saliva NE [‡]	0.052	NS
Unadjusted serum MPO	Saliva MPO	-0.291	NS
Unadjusted serum cit-H3	Saliva cit-H3	0.308	NS
Adjusted serum cf-DNA	Saliva cf-DNA	-0.347	NS
Adjusted serum NE [‡]	Saliva NE [‡]	-0.027	NS
Adjusted serum MPO	Saliva MPO	-0.021	NS
Adjusted serum cit-H3	Saliva cit-H3	0.382	NS

Spearman's *rho*. [‡] n=13.

cf-DNA: cell-free DNA; cit-H3: citrullinated histon-3; MPO: myeloperoxidase; NE: neutrophil elastase; NS: not significant; r: correlation coefficient.



Supplementary Fig. S3. ROC curves of NETosis markers and inflammatory parameters for differentiation of aBD from HC (A, B, C, and D). ROC curves of NETosis markers and inflammatory parameters for differentiation of aBD from iBD (E and F).

aBD: active Behçet's disease; iBD: inactive Behçet's disease; HC: healthy control.