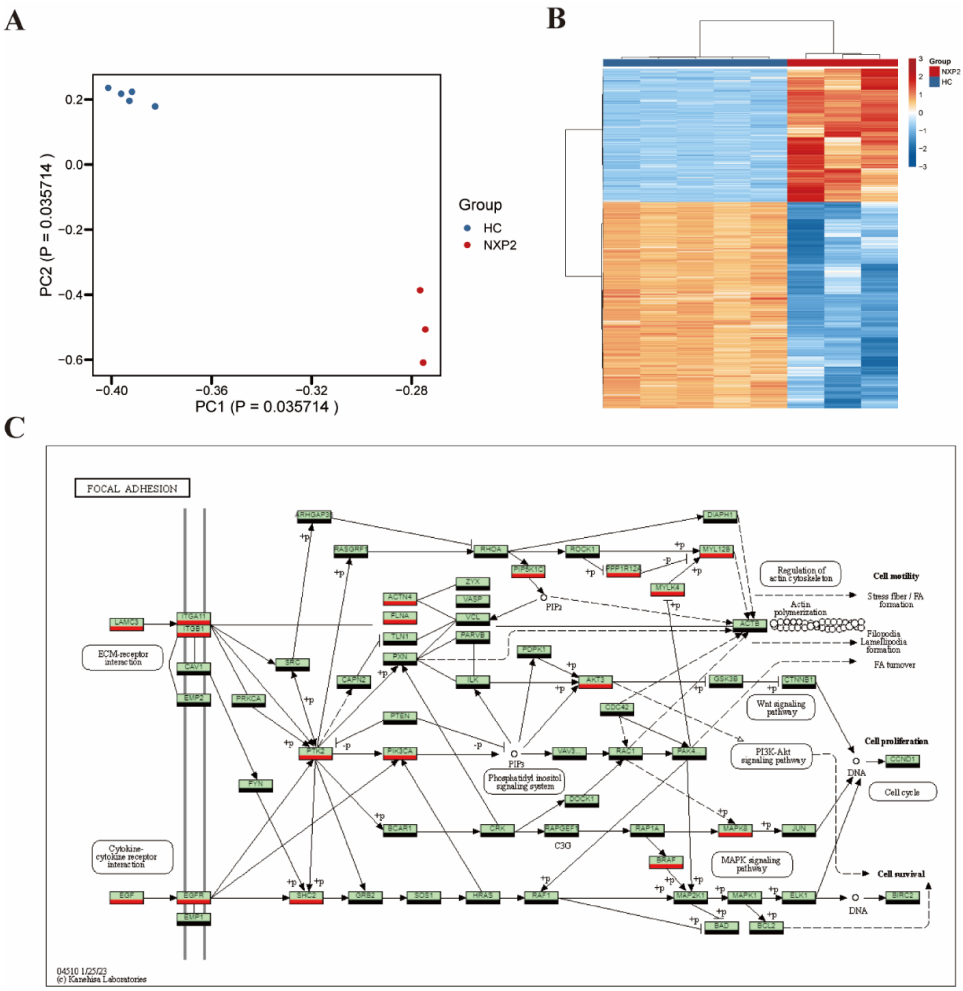

Fig S1. PCA plot (A) and heating map (B) indicated the alteration of transcriptomic profiles of NXP2^{+}DM patients compared to healthy control. (C) KEGG pathway of focal adhesion was activated in NXP2^{+}DM patients

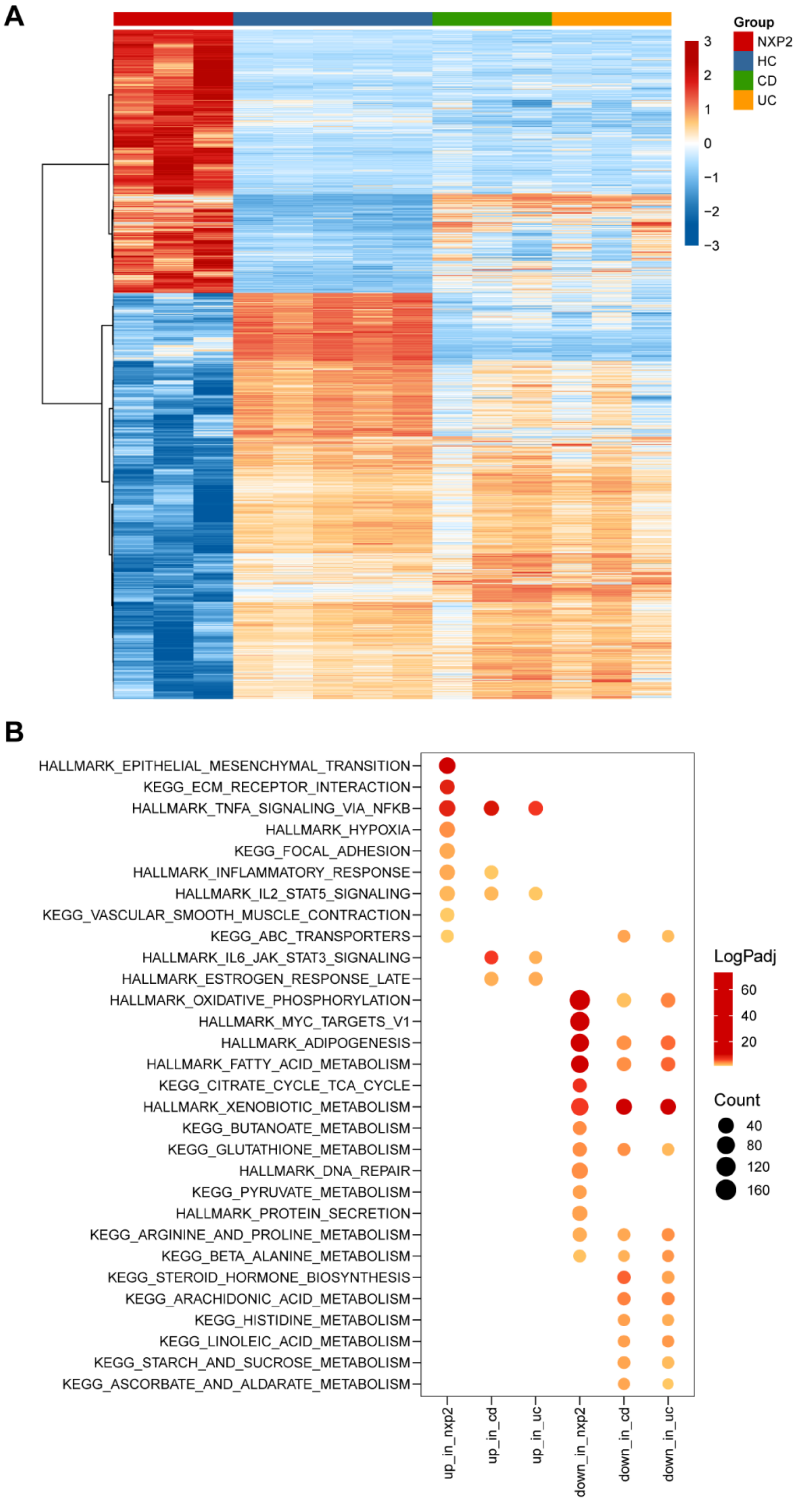

Fig S2. (A) The expression profile of NXP2+DM was different from IBD showing in heatmap. (B) Significantly differential gene pathways between NXP2+DM and IBD indicated by functional enrichment analysis. IBD, inflammatory bowel disease; UC, ulcerative colitis; CD, Corhn's disease.

	Case1	Case2	Case3	Case4	Case5	Case6	Case7	Case8	Case9	Case10
demographics										
gender	F	F	F	M	F	F	F	F	F	F
age (at diagnosis)	64	75	31	48	41	20	63	18	29	45
age (GI involvements)	64	75	33	49	43	31	64	18	29	46
disease courses to GI symptoms	3 months	4 months	31 months	14 months	15 months	11 years	9 months	5 months	7 months	10 months
duration from GI symptoms to perforation	2 months	3 weeks	2 months	2 weeks	3 months	-	-	3 weeks	2.5 months	2 months
clinical features										
cutaneous	+	+	+	+	+	-	+	+	+	+
muscle	+	+	+	+	+	+	+	+	+	+
calcinosis	-	-	+	-	+	+	+	+	-	+
interstitial lung disease	+	-	-	+	+	-	+	-	+	+
initial GI symptoms	abdonimal pain	abdonimal pain, melena	abdonimal pain, bloting	abdonimal pain	abdonimal pain, bloody stools	melena	intractable abdominal pain	abdonimal pain, vomiting	hematemesis, melena, abdonimal pain	abdonimal pain, hematemesis
perforation sites	multiple gastroduodenal perforations	jejunal perforation	ascending colon perforation	descending duodenum perforation	ileocecal perforation	-	-	stomach, duodenal bulb and descending perforations	jejunal perforation	ascending colon perforation
other GI findings	retroperitoneal hemorrhage	gastric ulcer with bleeding, extensive intestinal edema	colon edema	duodenum edema	colon edema	gastric ulcer with bleeding	jejunal ulcer, intestinal edema, mesenteric panniculitis	duodenum bleeding, intestinal edema	intestinal edema and upper gastrointestinal bleeding	descending duodenum ulcer with bleeding, extensive intestinal
Laboratory findings										
CK (U/L)	142	976	166	121	92	43	92	385	96	107
CRP (mg/L)	62.86	29.16	32.5	14.9	5.39	3.85	2.34	47.78	50.25	5.2
ESR (mm/h)	76	25	69	19	66	53	31	90	22	70
MSA	NXP2, Mi2α	NXP2, Ro52	NXP2	NXP2	NXP2	NXP2, Ro52	NXP2	NXP2, Ro52	NXP2, Ro52	NXP2
Infection	kebsiella pneumoniae, candida albicans	EBV	escherichia coli	escherichia coli	CMV, EBV, stenotrophomonas maltophilia	-	-	escherichia coli, pseudomonas aeruginosa, candida tropicalis	-	Acinetobacter baumannii, candida albicans
Treatments										
previous treatments	GCs, TCZ	GCs	GCs, MTX, MMF, Bari, CsA+tofa	GCs, tha, CyC	GCs, CyC, Infi	GCs, MTX, tofa	GCs, CyC, TCZ, tac, bari, Infi	GCs, tac	GCs, MTX, tac, tofa	GCs, tofa
last dosage of GCs before perforation	Pred 20mg/d	MP 80mg/d	MP 160mg/d	MP 80mg/d	MP 40mg/d	-	-	MP 120mg/d	Pred 20mg/d	MP 40mg/d
surgery	no surgery	no surgery	right hemicolectomy and colostomy	perforation repair	partial ileectomy and ileostomy	-	-	perforation repair, partial gastrectomy and Roux-en-Y	perforation repair	no surgery
sequential treatment	GCs	GCs	GCs+sirolimus	GCs	GCs+tofa	GCs	GCs+Vedo	GCs+thalidomide+bari/Vedo	GCs+Vedo	GCs+Vedo
follow-up after perforation/bleeding	1 week	3 weeks	3 weeks	1 week	2 years	2 years	1 year	14 months	3 months	3 month
outcome	deceased	deceased	deceased	deceased	improved	improved	improved	improved	improved	improved

Table S1. Clinical features of NXP2+ DM with gastrointestinal involvements

F is for female and M is for male. GCs, glucocorticoids; TCZ, tocilizumab; MTX, methotrexate; MMF, mycophenolate mofetil; Bari, baricitinib; CsA, cyclosporine A; tofa, tofacitinib; tha, thalidomide; CyC, cyclophosphamide; Infi, infliximab; tac, tacrolimus; vedo, vedolizumab.