Supplementary Table S1. Literature search strategy in Embase.

No.	Query
#12	#10 AND #11
#11	'neuro' OR 'neurological' OR 'neurologic'
#10	#8 AND #9
#9	'infliximab' /exp
#8	#1 OR #2 OR #3 OR #4 OR #5OR #6 OR #7
#7	'Behçet*' OR 'Behçet*'
#6	'adamantiades-Behçet*' OR 'adamantiades-Behçet'
#5	'silk route disease'
#4	'triple symptom complex'
#3	'parenchymal neuro-Behçet syndrome'
#2	'Behçet disease'
#1	'Behçet syndrome'

Note: The synonyms were combined from Medical Subject Headings terms (MeSH) and EMTREE trerms. No restrictions were set on publication types, languages, and year of publication.

Supplementary Table S2. Criteria for risk of bias assessment of eligible studies.

Domain	Item	Response
Selection bias	1.Did the study apply clear inclusion/exclusion criteria in the selection of participants?	Low risk, the study reported clear and appropriate inclusion/exclusion criteria; high risk, the criteria used in the study may lead to bias in the estimation of the response rate; unclear, there is no relevant information.
	2.Were the participants representative of the targeted population?	Low risk, the participants were recruited consecutively or using probability sampling method; high risk, the participants in the study were biased from the targeted population; unclear, there is no relevant information.
Performance bias	1.Did researchers rule out any impact from a concurrent intervention or an unintended exposure that might bias results?	Low risk, there was no concurrent or unintended intervention, or the existing concurrent intervention is unlikely to influence the response rate; high risk, there were some concurrent or unintended intervention that may influence the response rate; unclear, there is no relevant information.
	2.Did variation from the study protocol compromise the conclusions of the study?	Low risk, the reporting results are concordant with the information from registration and study protocol; high risk, there are some changes in the conducting of the study compared with the registration or study protocol; unclear, there is no available registration or protocol.
Attrition bias	1.Was the follow-up completed in all subjects?	Low risk, the primary outcome could be assessed in more than or equal to 90% of the participants, or there is solid evidence indicating that those who lose to follow-up were similar with those still staying in the cohort; high risk, less than 90% of the participants contributed to the primary outcome; or there is evidence indicating that those who lose to follow-up were different with those still staying in the cohort; unclear, there is no relevant information.
Detection bias	1.Were the outcome assessors blinded to the intervention or exposure status of participants?	Low risk, the outcome assessors were totally blinded to the intervention; high risk, the outcome assessor knew the intervention; unclear, there is no relevant information.
	2.Were the inclusion/exclusion criteria measured using valid and reliable measures, implemented consistently across all study participants?	Low risk, the personnel who recruited the participants were unaware of the intervention, or objective measures were used in the patients recruiting; high risk, the personnel who recruited the participants were aware of the intervention, or there is evidence that the recruiting of participants will lead to biased estimation of the primary outcome; unclear, there is no relevant information.
	3.Were primary outcomes assessed using valid and reliable measures, implemented consistently across all study participants?	Low risk, the personnel who assessed the outcome were unaware of the intervention, or objective measures were used in the primary outcome; high risk, the personnel who assessed the outcome were aware of the intervention, or there is evidence that the assessment of the primary outcome will lead to biased estimation; unclear, there is no relevant information.
Reporting bias	1.Were the potential outcomes pre-specified by the researchers? Are all pre-specified outcomes reported?	Low risk, all the predefined outcomes in registration or study protocol were reported in the study; high risk, the investigators selectively reported some predefined outcomes, or there are changes in the outcomes of interest; unclear, there is no available registration or study protocol.

Supplementary Table S3. Baseline characteristics of eligible studies.

First Author (year)	Study design	Country	Sample size	Diagnostic Criteria for BS	follow-up time (month)	Duration of IFX treatment (month)	Mean age (SD)	concomitant medication	IFX regimen
Kikuchi (2008)	Retrospective, single-centre, open-label, single-arm cohort study	Japan	5	ISG criteria	6	3.5	35.8(7. 2)	MTX, GCs	5 mg/kg at week 0, 2, 6, and 14
Haghighi (2011)	Case series	Iran	4	ISG criteria	411-36	3.5-5.5	40.5(1.9)	GCs, CTX, Col	3 or 5 mg/kg at week 0, 2, 6, and every 8 weeks
Hibi (2016)	Prospective, single-centre, open-label, single-arm cohort study (NCT01532570)	Japan	3	Japan criteria	13.5	11.5	38.5(12.0)	GCs	5 mg/kg at week 0, 2, 6, and every 8 weeks until week 46
Desbois (2016)	Retrospective, multicentre, observational, single-arm cohort study	France	13	ICBD criteria	3-163	≥12	40.2(9.4)	AZA, GCs, MTX, MMF	5mg/kg
Pipitone (2008)	Case series	Italy	8	ISG criteria (n=6), unclear(n=2)	3-24	3-24	50.3(13.8)	GCs, CsA, MTX	5 mg/kg[a]
Zeydan (2016)	Prospective, single-centre, open-label, single-arm cohort study	Turkey	14	ISG criteria	16-104.9	16-104.9	38.5(10.0)[b]	GCs, AZA, COL, CsA, CTX, IFN-α	5 mg/kg at week 0, 2, 6, and every 8 weeks
Yalcin (2021)	Retrospective, single-centre, single-arm cohort study	Italy	19	ISG (diagnosed before 2014) and ICBD criteria	11-79	11-79	36.6(11.6)	AZA, MTX	5 mg/kg at week 0, 2, 6, and every 8 weeks

SD: Standard Deviation; BS: Behçet's syndrome; p-NBS: parenchymal neuro-Behçet's syndrome; IFX: infliximab; ISG: the International Study Group criteria for BD; MTX: methotrexate; GC: glucocorticoids; CTX: cyclophosphamide; Col: colchicine; ICBD: the International Criteria for Behçet's Disease; AZA: azathioprine; MMF: mycophenolate mofetil; CsA: cyclosporin A; IFN-α: interferon-α
a: 4 patients with 5 mg/kg at weeks 0, 2, and 6 and bimonthly thereafter; 2 patients with 5 mg/kg at weeks 0, 1, 3, and 8 and bimonthly thereafter;1 patient with 5 mg/kg at weeks 0, 2, and 6; 1 patient with 5 mg/kg at weeks 0, 2, and 6 and every 6-8 weeks thereafter.
b: This is the mean age of 14 patients with pNBS and one patient with non-pNBS.

Supplementary Table S4. Risk of bias in eligible studies.

First author(year)	selection bias		Performance bias		Attrition bias		Detection bias		Reporting bias
	inclusion/ exclusion criteria	represent-ativeness	rule out intervention	variation from protocol	completed follow-up	blindness	measures for recruitment	measures for outcome	pre-specified outcomes
Kikuchi (2008)	Low	Low	Low	Unclear	Low	Unclear	Low	Low	Low
Haghighi (2011)	Low	Low	Low	Unclear	Low	Unclear	Low	Low	Low
Hibi (2016)	Low	Low	Low	Low	High	Unclear	Low	Low	Low
Desbois (2016)	Low	Low	low	Unclear	Low	Unclear	Low	Low	Low
Pipitone (2018)	Unclear	Low	Unclear	Unclear	Low	Unclear	Unclear	Low	Unclear
Zeydan (2016)	Low	Low	Low	Low	Low	Unclear	Low	Low	Low
Yalcin (2021)	Low	Low	Low	Low	Low	Unclear	Low	Low	Low

Supplementary Table S5. The adverse events reported by studies related to infliximab used in the treatment of Neuro-Behcet's disease.

First author (year)	Sample size (n)	AEs (n)	Severe AEs (%)	Detailed safety information
Kikuchi (2008)	5	2	1	One patient experienced a brief headache. Another patient showed signs of suspected subclinical pneumocystis pneumonia. A chest CT scan revealed a ground glass opacity lesion, with a slight elevation in sialylated carbohydrate antigen KL-6 levels. Despite normal β-D-glucan levels and no positive microbiological findings, the lesion exhibited improvement following low-dose treatment.
Haghighi (2011)	4	1	1	Varicella zoster infection was seen in one patient.
Hibi (2016)	3	3	0	One case of infections was observed, and specific details of the other two adverse reactions were not mentioned.
Desbois (2016)	13	3	3	Three patients experienced adverse reactions, including pneumonia, heart failure, and behavioural disorder. The latter two discontinued the treatment.
Pipitone (2018)	8	NA	0	It was not mentioned whether any adverse events occurred, but it was reported that no serious adverse events occurred.
Zeydan (2016)	14	5	0	One patient experienced a rash, two patients had headaches, and two patients had nausea.
Yalcin (2021)	19	1	1	One patient experienced allergic reaction and discontinued the treatment.
Data from PUMCH (2023)	11	1	1	Case 11 developed acute hematogenous TB four months after initiating IFX, despite having a negative TB screening.

NA: not available; AEs: adverse events; TB: tuberculosis.