

Neuro-Behçet disease: spectrum of imaging findings

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Pathway

- 1.Introduction
- 2.Neuro-Behçet spectrum of imaging findings:
 - 2.1.Parenchymal
 - Typical
 - Atypical
 - 2.2.Non-parenchymal
 - Vascular complications
 - Intracranial hypertension
 - Aseptic meningitis
- 3.Differential diagnosis
- 4.Take home messages



Introduction

- Behçet disease is a inflammatory multisystemic disorder of uncertain etiology clinically characterized by **recurrent genital and oral ulcers and uveitis**, but its manifestations can arise in almost any tissue, consisting in **inflammatory perivasculitis**.
- **Neurological involvement in Behçet (Neuro-Behçet)** is maybe one of the most uncommon manifestations of the disease spectrum, but surely is one of the most disabling, occurring mostly on late onset, being present on the hall of other inflammatory / demyelinating CNS diseases.
- Neuro-Behçet consists in **CNS involvement** (parenchymal and non-parenchymal patterns), but **PNS and other uncommon syndromes** (such as acute meningeal and psychiatric symptoms) are some other recognized manifestations, giving this disorder a **multifaceted presentation**.

Neuro-Behçet disease: spectrum of imaging findings

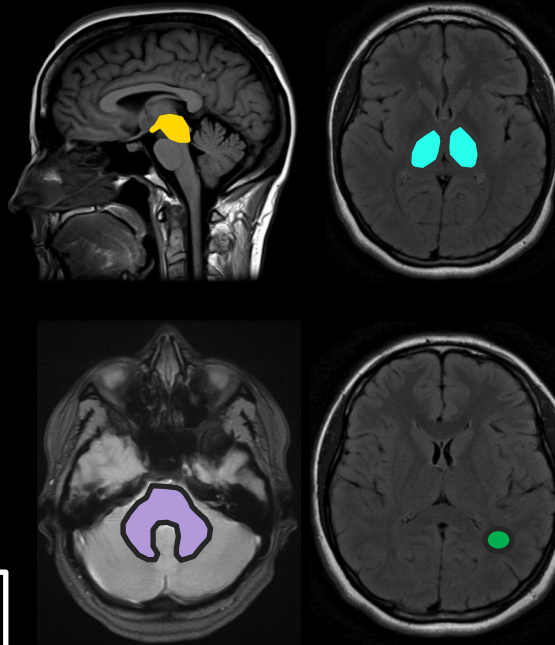
NEURO-BEHÇET

Parenchymal

Typical

Midbrain-diencephalic >
Cerebellar peduncles / Pons >
Basal ganglia > Medulla > Spinal
cord

Subcortical lesions



Intracranial hypertension

Dural sinuses thrombosis
arterial dissection, occlusion,
aneurysms

Aseptic meningitis

Non-parenchymal

CSF (70-80%)

Pleocytosis (neutrophilia □ lymphocytosis);
High protein levels; Normal glucose

CSF

High pressure – normal contents

Neuro-Behçet disease: spectrum of imaging findings

NEURO-BEHÇET

Parenchymal

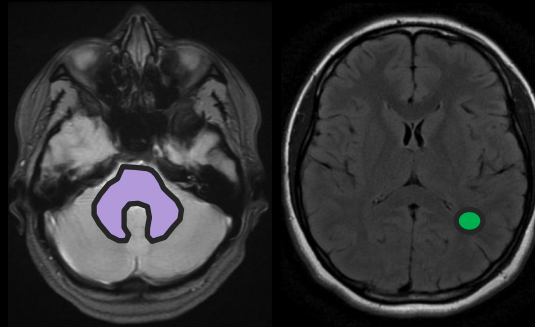
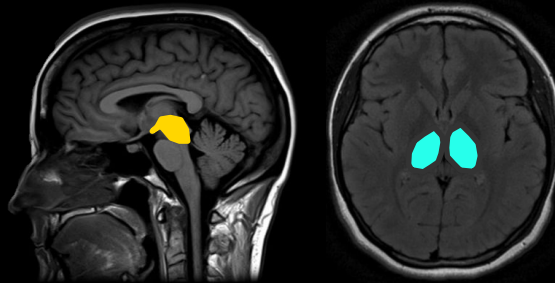
Typical

Midbrain-diencephalic >
Cerebellar peduncles / Pons >
Basal ganglia > Medulla > Spinal
cord

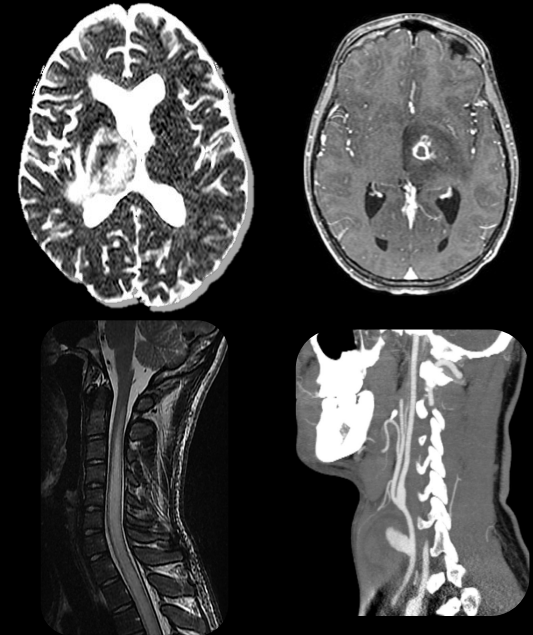
Subcortical lesions

CSF (70-80%)

Pleocytosis (neutrophilia □ lymphocytosis);
High protein levels; Normal glucose



Atypical



Neuro-Behçet disease: spectrum of imaging findings

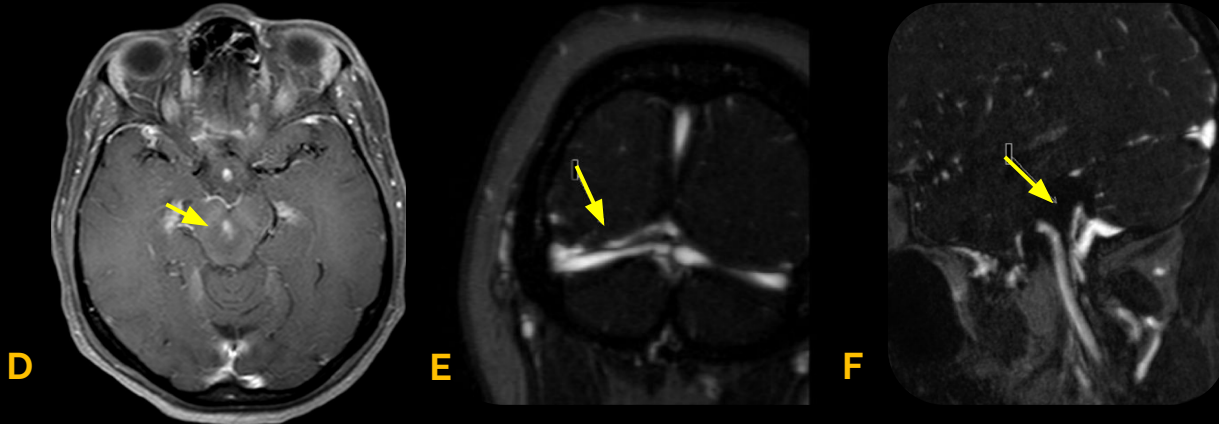


Typical

Neuro-Behçet

Coronal T2, Axial FLAIR (A-C):

Mesodiencephalic hyperintense lesion, sparing red nucleus (red arrow) extending to superior cerebellar peduncle (red arrow).



Axial T1 FS C+ (D):

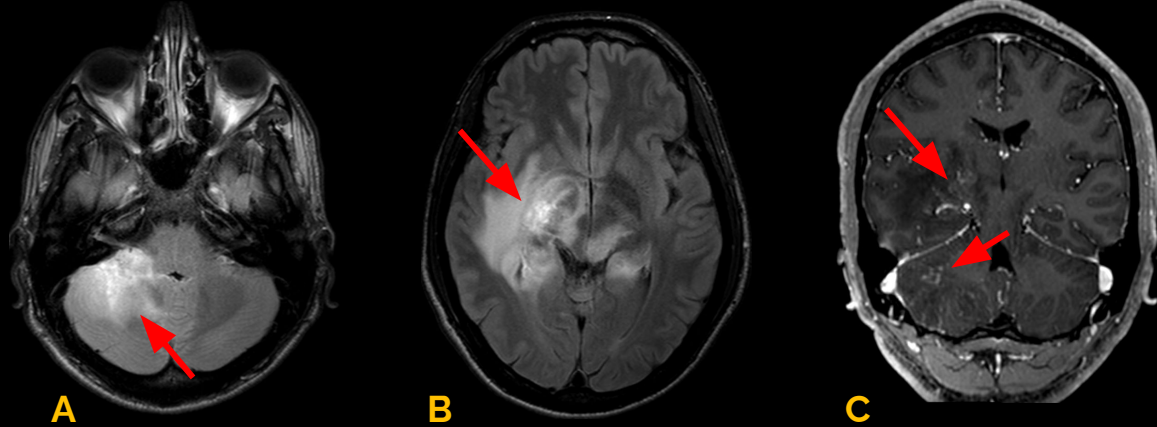
Thick linear contrast enhancement (yellow arrow).

Coronal and Sagittal MRI

venography (E-F):

Chronic partially recanalized transverse and sigmoid sinus thrombosis (yellow arrows).

Neuro-Behçet disease: spectrum of imaging findings



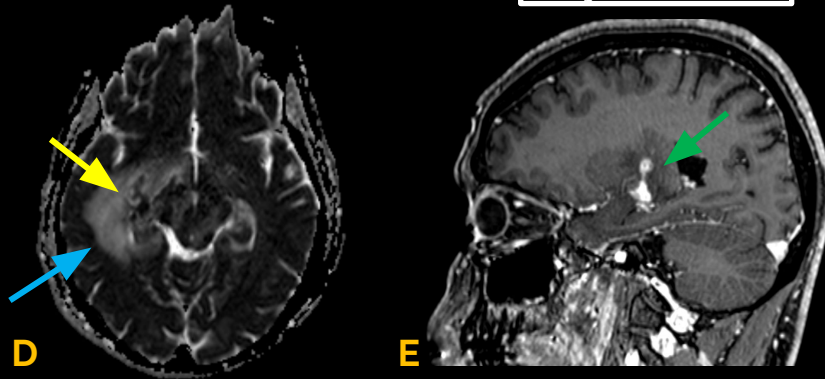
Typical Neuro-Behçet Axial FLAIR (A,B):

High signal intensity tumefactive areas involving mesodiencephalic junction, basal ganglia, and cerebellum (red arrows).

Coronal T1 C+ (C):

Scattered area of contrast enhancement (red arrow).

2 years later...



ADC map (D):

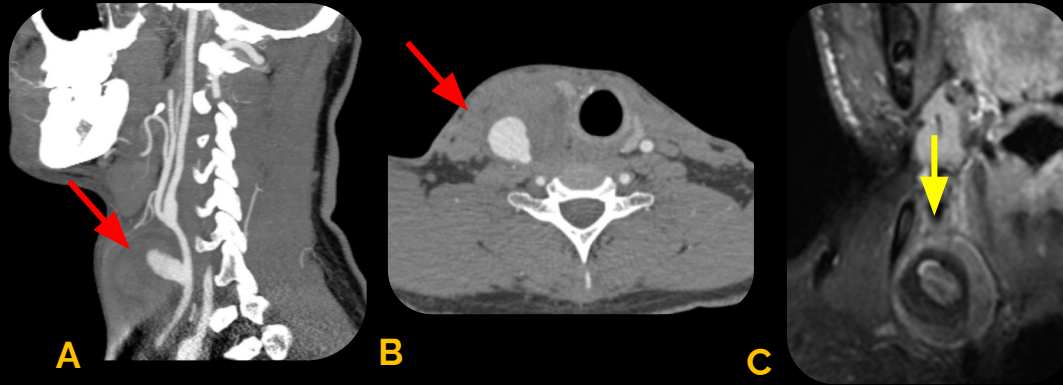
Regions of restricted diffusion (yellow arrow), but ruling vasogenic edema (blue arrow).

2-year control MRI (E):

New lesions affecting right temporoinsular region (green arrow) and brainstem (not shown).

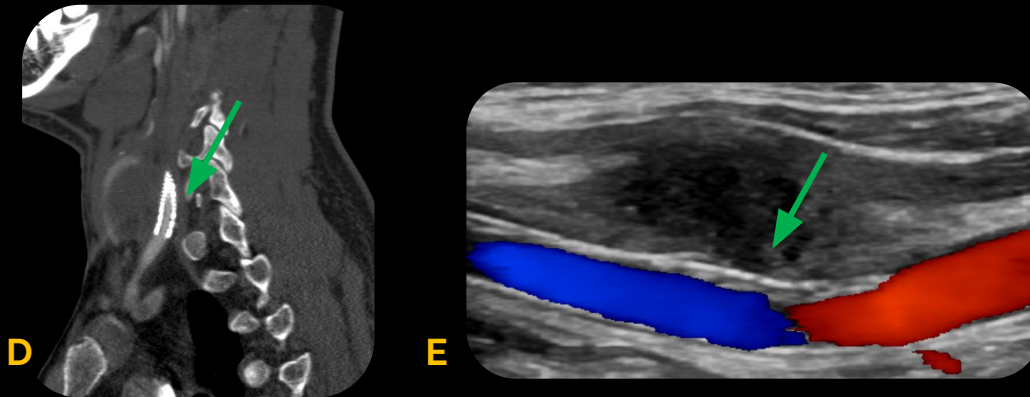
Neuro-Behçet disease: spectrum of imaging findings

Atypical Neuro-Behçet Carotid pseudoaneurysm



Sagittal (A); Axial (B) CT angiography:
Right common carotid artery partially thrombosed pseudoaneurysm (red arrows).

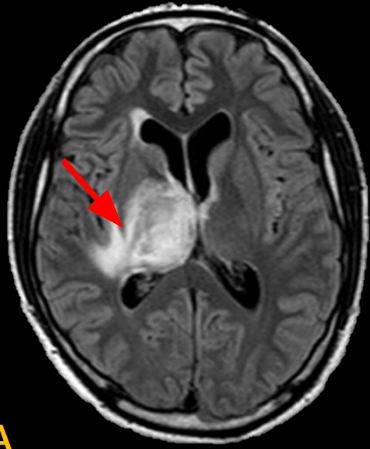
Postcontrast Black-Blood (C):
Peripheral pseudoaneurysm contrast enhancement around the (yellow arrow).



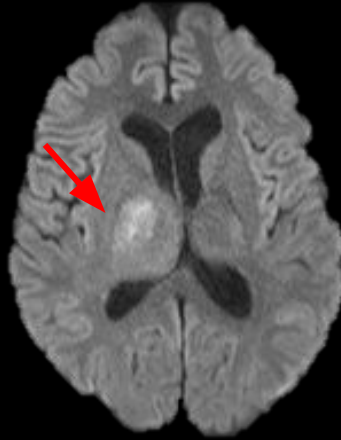
Posttreatment CT angiography (D) and
US Doppler (E):
Patent stent (green arrows) in the right common carotid artery.

Neuro-Behçet disease: spectrum of imaging findings

Atypical
Neuro-Behçet
Pseudotumoral



A

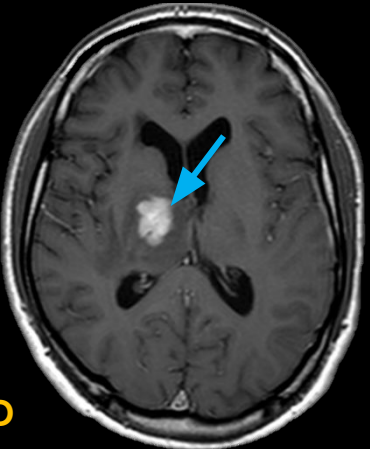


B

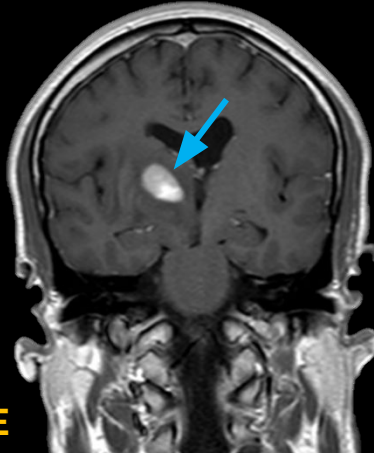


C

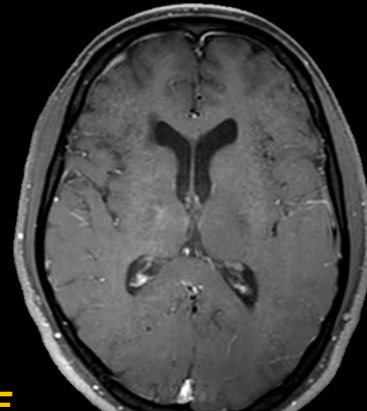
Axial FLAIR (A), DWI (B) and ADC (C):
High signal tumefactive right capsulo-thalamic lesion, with central restricted diffusion (red arrows).



D



E



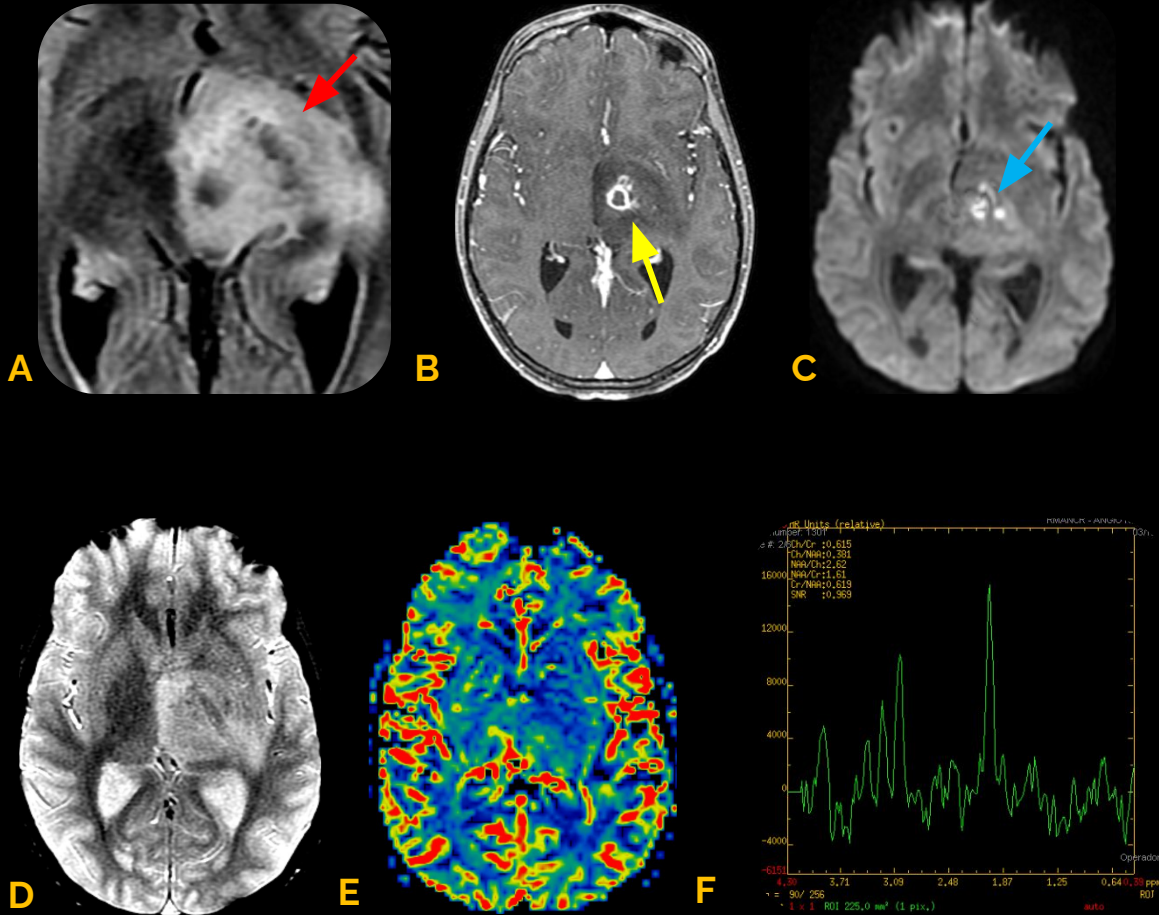
F

Coronal T2 FS (D) and Coronal T1 FS post-contrast (E):
Nodular homogeneous enhancing lesion in the same area of central restricted diffusion above (blue arrows).

Control MRI after corticosteroids(F):
Lesion regression was noted.
Patient fulfilled Behçet's disease criteria.

Post treatment

Neuro-Behçet disease: spectrum of imaging findings



Atypical
Neuro-Behçet
Pseudotumoral
meningitis

Axial FLAIR (A):
Left mesodiencephalic junction tumefactive lesion (red arrow).

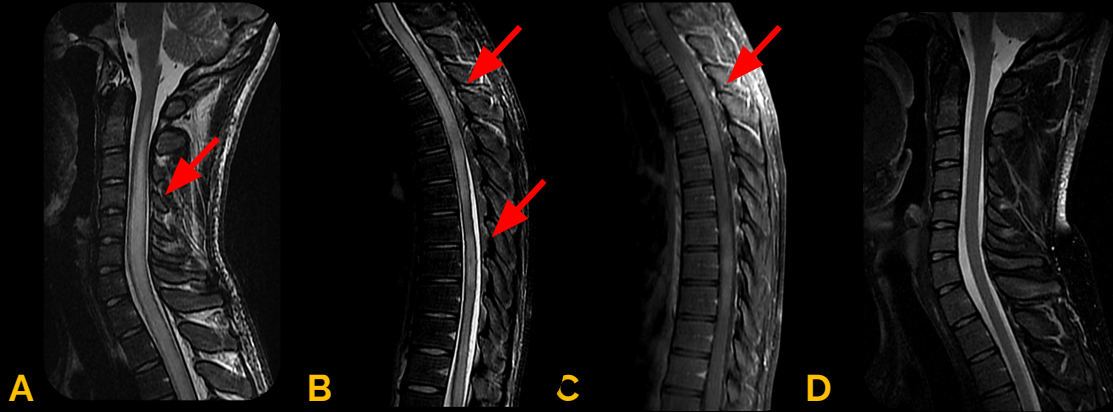
Axial T1 FS C+ (B):
Anular enhancing areas (yellow arrow) with areas of restricted diffusion (C) regions (blue arrow).

Axial T2* GE (D):
No signs of bleeding or calcifications.

Perfusion MR imaging (E) and spectroscopy (F):
Low rCBV and no abnormal peaks.

Neuro-Behçet disease: spectrum of imaging findings

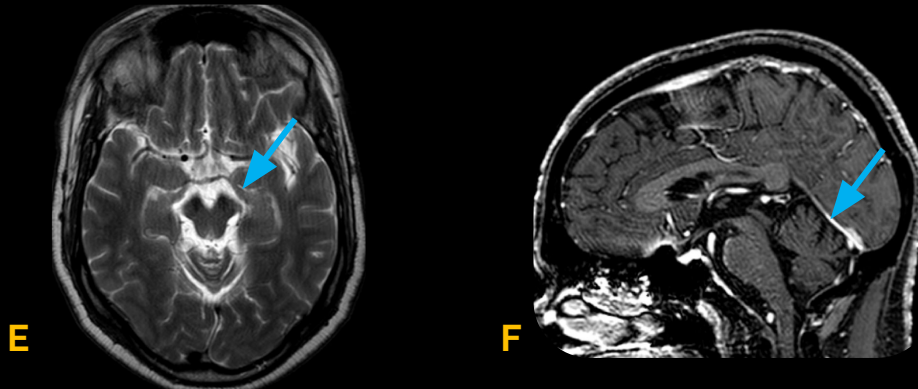
Treatment



Atypical
Neuro-Behçet
Longitudinally extensive myelitis

Sagittal T2 FS and T1 FS C+ on cervical and thoracic levels (A-C):
Longitudinally extensive myelitis with patchy enhancement (red arrows).

Control MRI after treatment (D):
Complete recovery.



Brainstem and cerebellar atrophy

Axial T2 (E):
Brainstem atrophy, predominantly in midbrain (blue arrow).

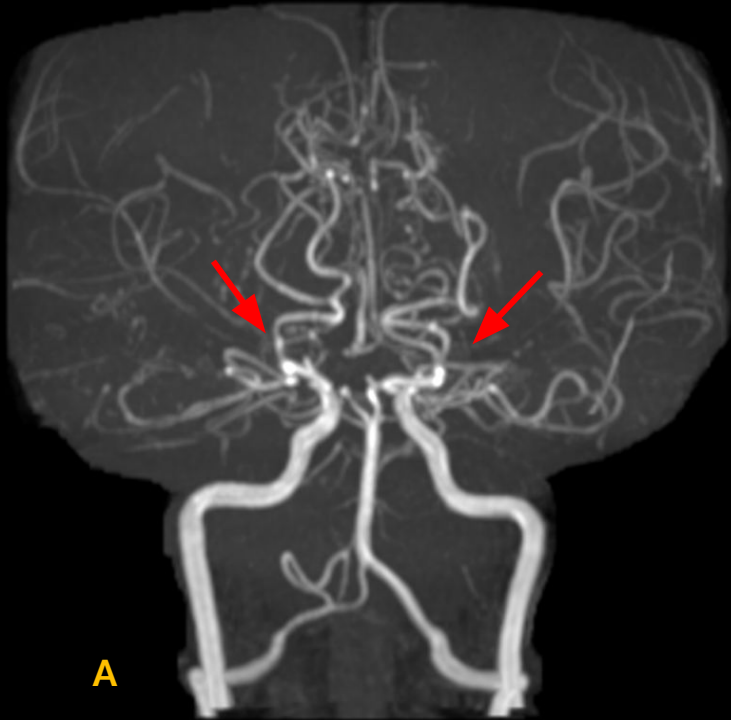
Sagittal T1 FS C+ (F):
Mild cerebellar atrophy (blue arrow).

Patient presenting with ataxia

Neuro-Behçet disease: spectrum of imaging findings

Atypical
Neuro-Behçet

Arterial occlusion ("Moyamoya like")



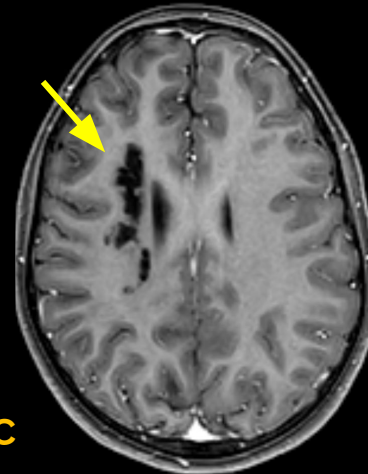
A



B

MRI angiography (A,B):

Occlusion of the supraclinoid portion of both internal carotid arteries (red arrows) with abnormal collateral vessels and M1/A1 subocclusion or occlusion.



C

Axial T1 FS C+ (C):

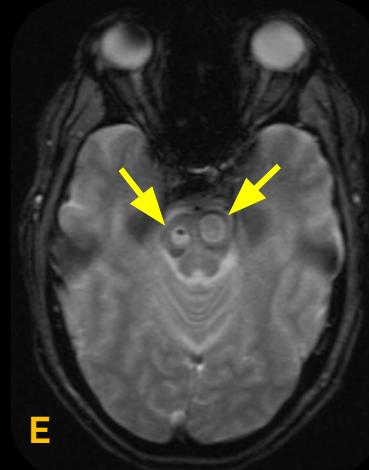
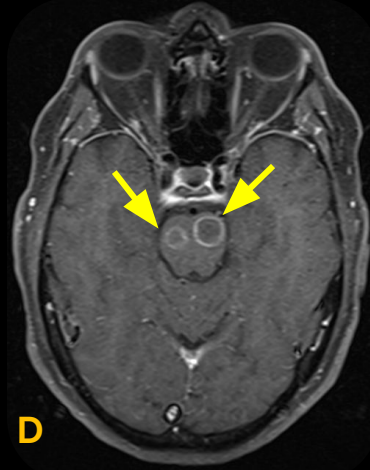
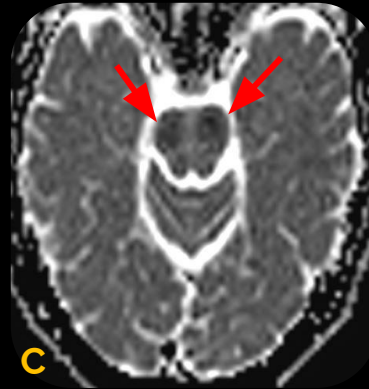
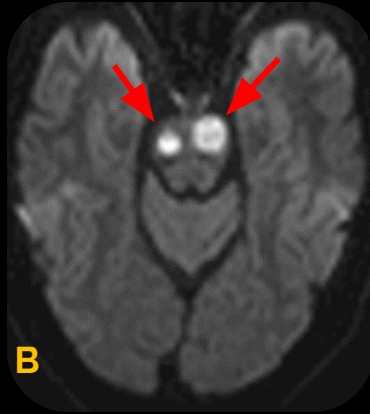
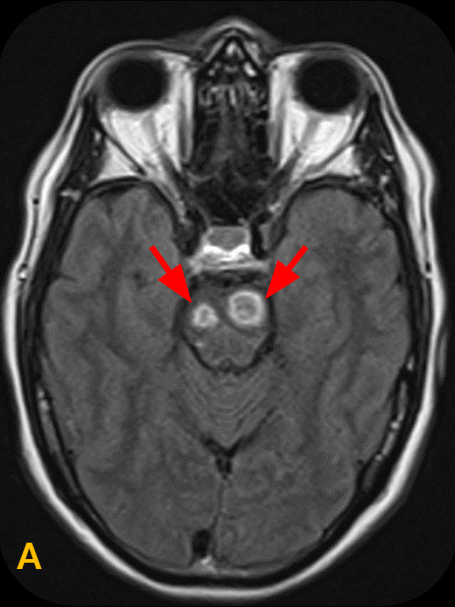
Patient with Behçet diagnosis presenting sequel ischemic areas at centrum semiovale / corona radiata's white matter (yellow arrow).

Neuro-Behçet disease: spectrum of imaging findings

Atypical
Neuro-Behçet
Aseptic Abscess

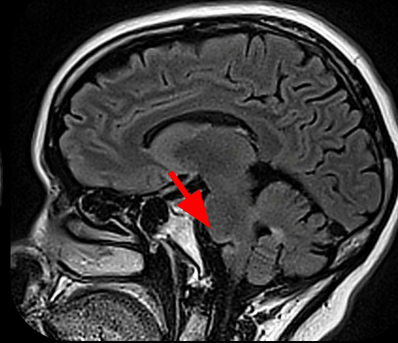
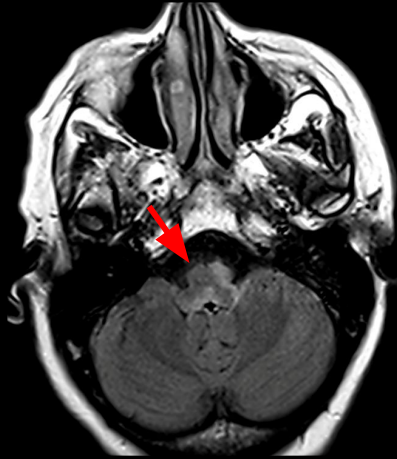
DWI (B) and ADC (C):

Central restricted diffusion, later confirmed like aseptic abscesses (**red arrows**).



Axial FLAIR (A) shows two rounded lesions with a high signal halo (**A-arrows**) that enhances smooth and regular (**D-arrows**), accompanied by peripheral hemosiderin (**E-arrows**).

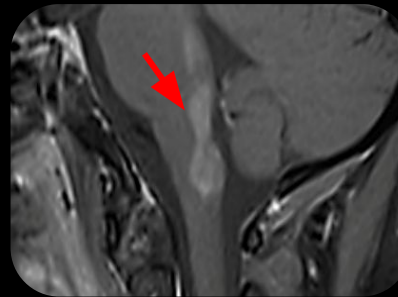
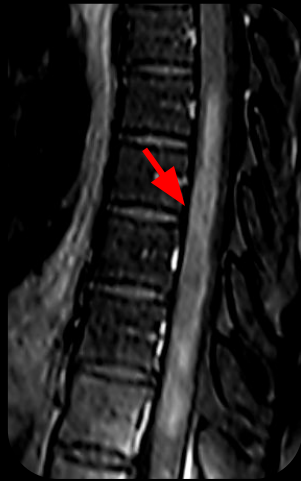
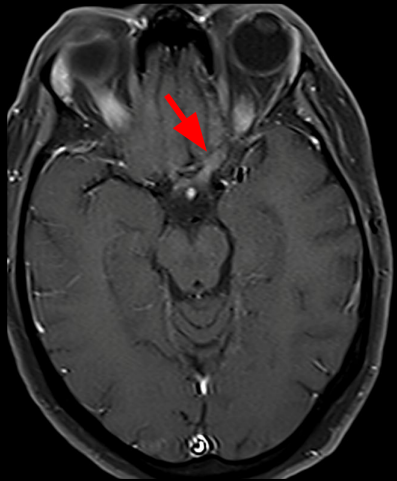
Training differential diagnosis by cases



Case 1: Make your bets!

A 44 years old healthy woman presenting with low left visual acuity, náusea and vomiting for 2 weeks and new progressive onset inferior limbs paraparesia about 5 days ago

CSF: Pleocytosis (lymphocytes) and high protein levels



A) Neuromyelitis optica

B) Bickerstaff encephalitis

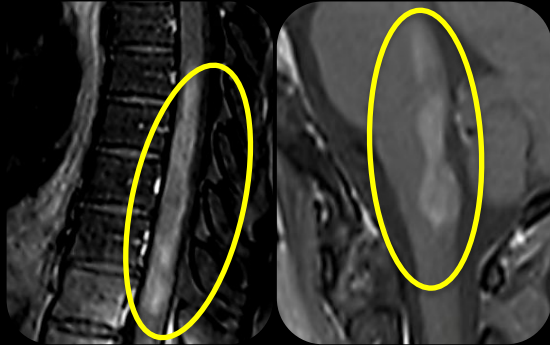
C) Multiple sclerosis

D) Listeria encephalitis

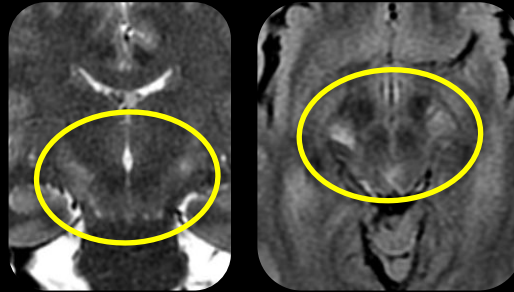
Training differential diagnosis by cases

NMO: *How to approach?*

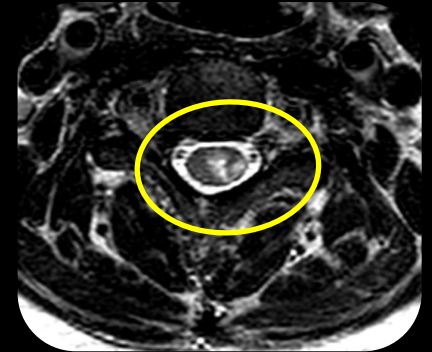
Autoimmune
Demyelinating
Anti-AQP5 IgG



Optic nerve (long segment)
Spinal cord (long segment)
Area postrema

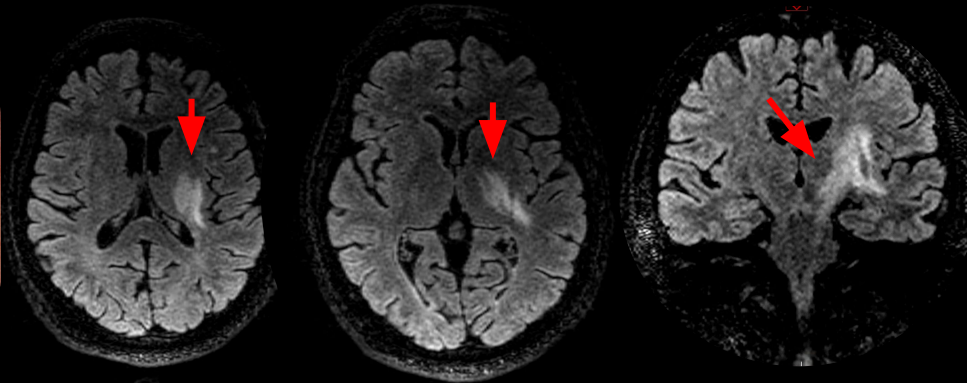


Periventricular lesions
Thalamus / Diencephalon
Brainstem tegmentum
Corticospinal tracts



T2 Bright Spotty lesions (>CSF)

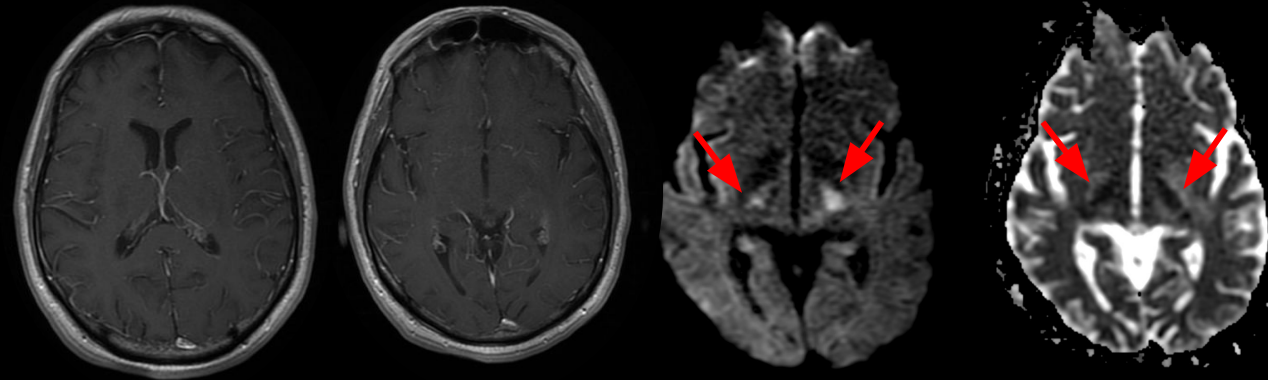
Training differential diagnosis by cases



Case 2: And what about now?

A 74 years old man presented with right hemiparesia about 1 week ago. He had fever, chills, odinofagia and headache. Recurrent episodes of non-identified fever

CSF: Pleocytocis (405 cells/ >90% neutrophils)



A) Neuro-Sweet

B) Bickerstaff encephalitis

C) Neuromyelitis optica

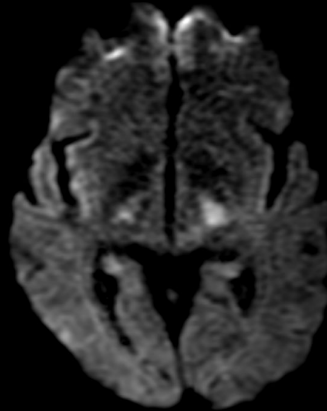
D) Listeria encephalitis

Training differential diagnosis by cases

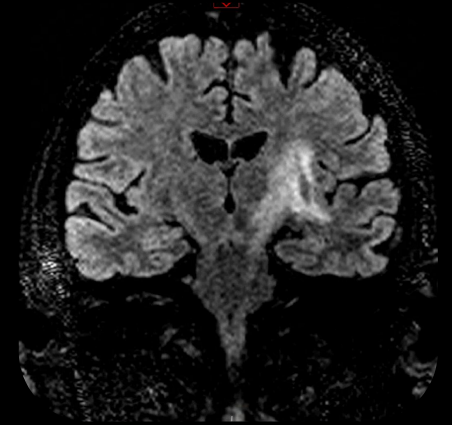
Neuro-Sweet: *How to approach?*



Neutrophilic dermatosis
Encephalitis
HLA B54 +

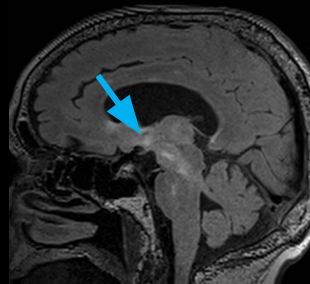
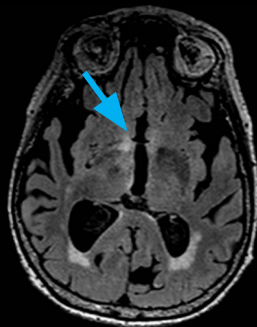
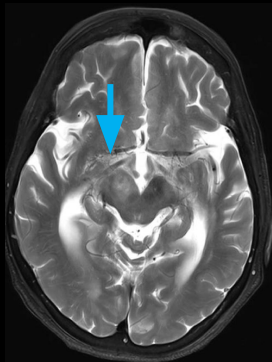


Brainstem
Basal ganglia
Cortex



Can be paraneoplastic

Training differential diagnosis by cases



Case 3: Are you still there?

Female, 90 years old, presenting with confusional state and hipoactivity for 2 days.

Rapidly progressive cognitive impairment during last month, with anterograde amnesia.

Left lower limb paraparesia

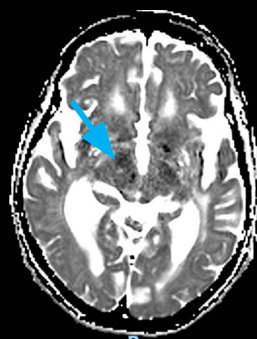
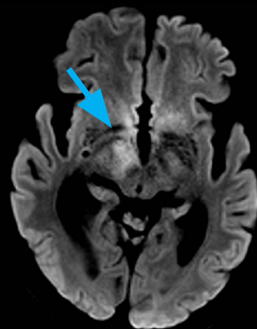
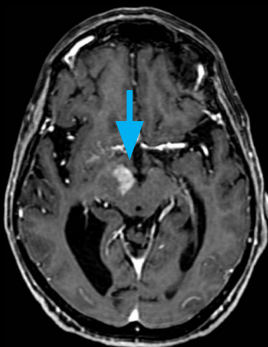
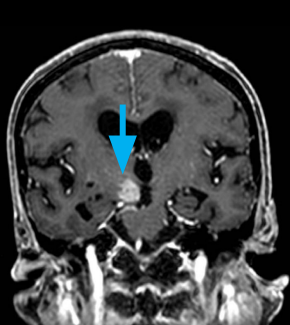
CSF: normal

A) Lymphoma

B) High-grade glioma

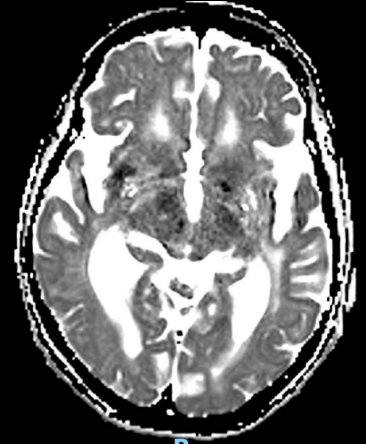
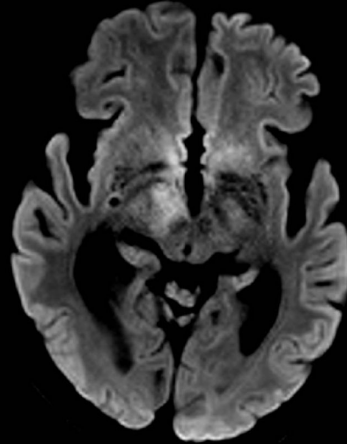
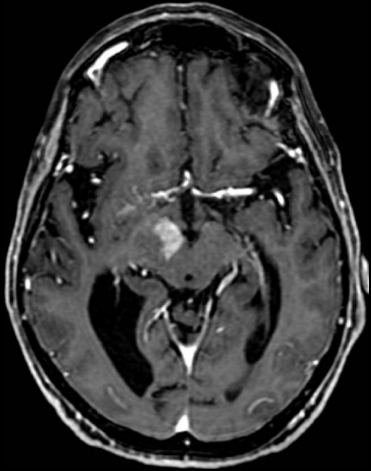
C) Metastasis

D) Listeria rombencephalitis



Training differential diagnosis by cases

Lymphoma: *How to approach?*

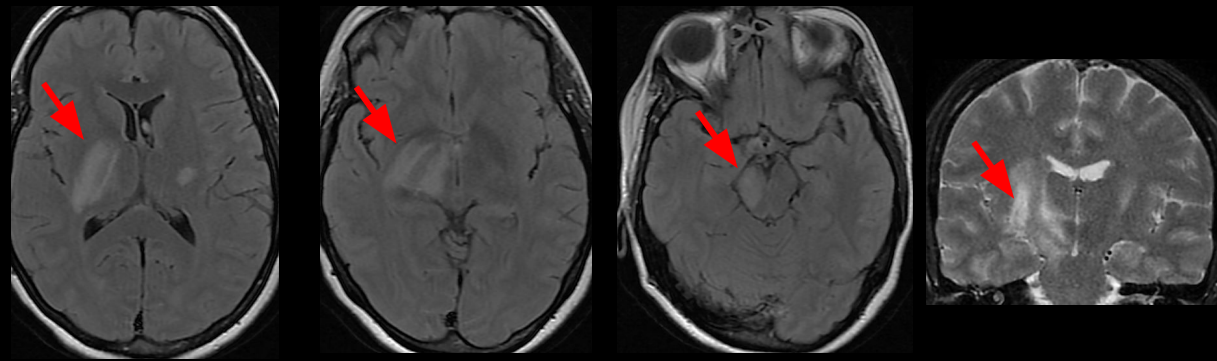


Spontaneous hyperdense
Restricted diffusion
Solid and homogeneous
enhancement

Central / Nucleocapsular
Meningeal

Periventricular
Basal ganglia
Brainstem

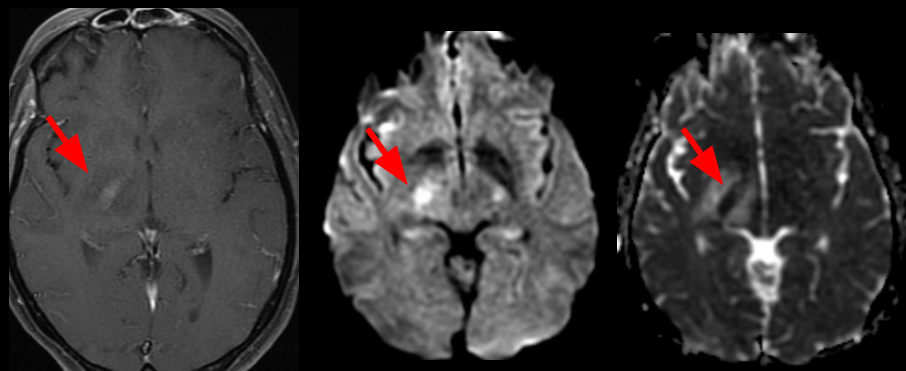
Training differential diagnosis by cases



Case 4: Almost there... Keep calm!

A 30 years-old men with left hemibody paresis for 2 days, holocranial moderate headache

CSF: Pleocytosis + High protein levels



A) Neuro-Behçet

B) Sarcoidosis

C) Lyme

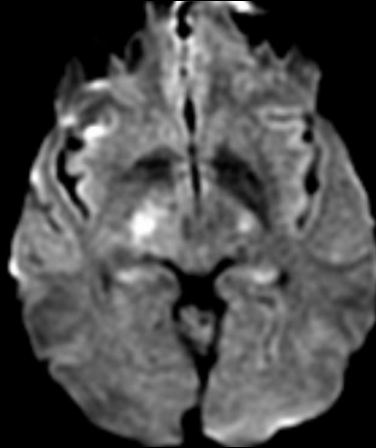
D) Syphilis

Training differential diagnosis by cases

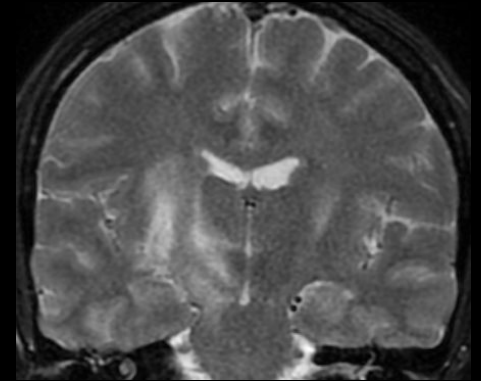
Neurosyphilis: *How to approach?*



Palmoplantar
lesions
(Secondary syphilis)

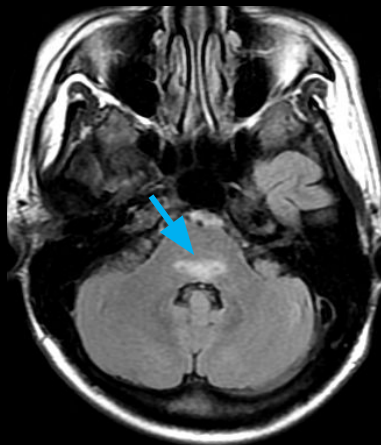


Early: Meningeal Meningovascular
Late: Tabes Dorsalis



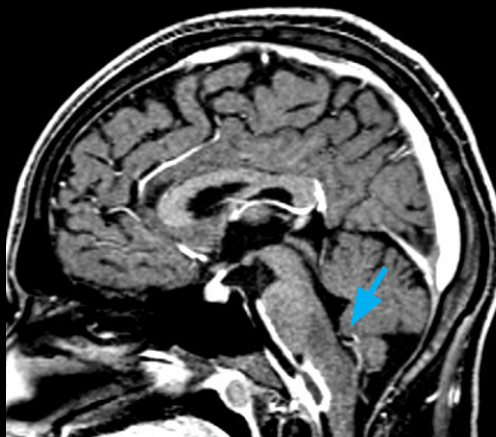
Infacts
Vasculitis
Cranial nerves
Meninges

Training differential diagnosis by cases



Case 5: Believe me!

A 17 years old man with non-Hodgkin's lymphoma, presenting with fever and headache



A) Listeria

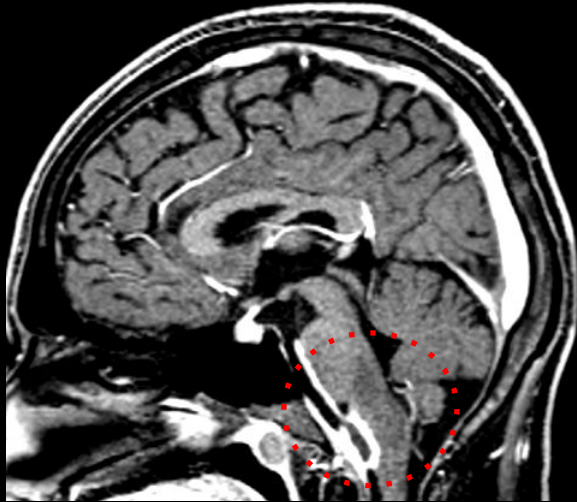
B) Lyme

C) Sarcoidosis

D) Schistosomiasis

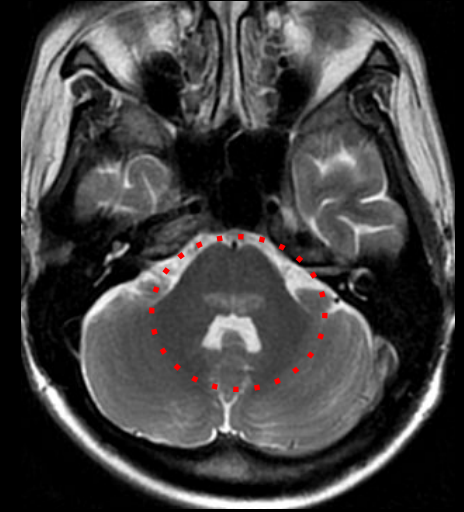
Training differential diagnosis by cases

Listeria: *How to approach?*



Site:

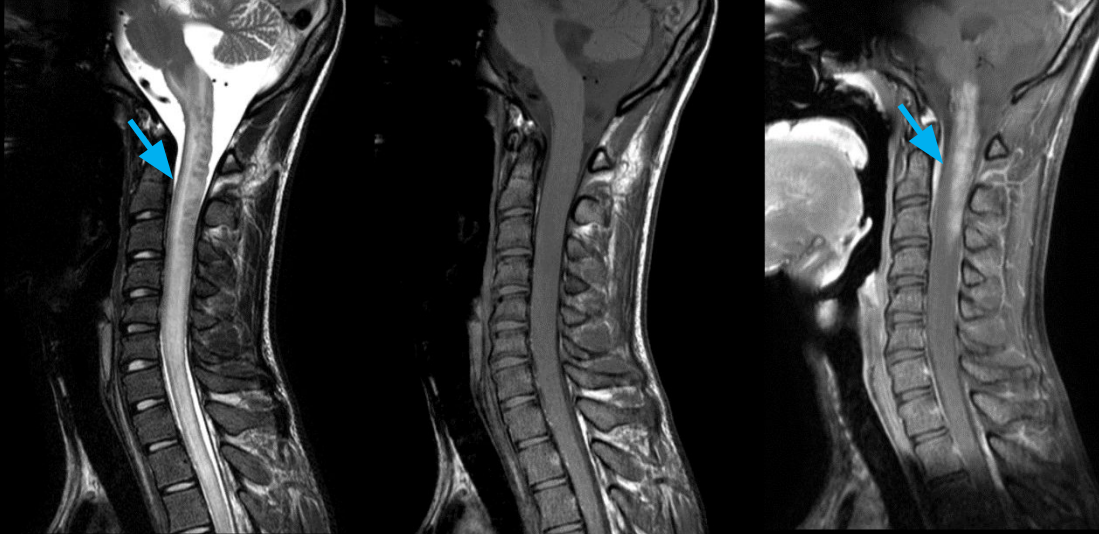
Diencephalic-mesencephalic
junction
(Behçet =
mesencephalonpontine)



Most common etiology for
rhombencephalitis

Elderly

Training differential diagnosis by cases

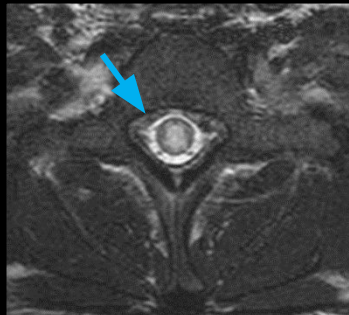


Case 6: Farewell!

A 19 years old man presenting with fever, weight loss, diarrhea and cervical pain.

Progressive and rapid progression to tetraparesia.

CSF: Pleocytosis + High protein levels + eosinophilia.



A) Listeria

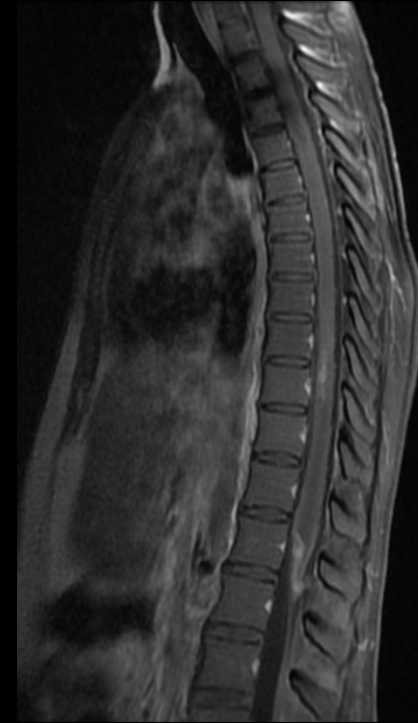
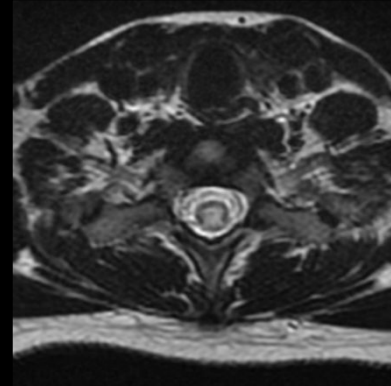
B) Lyme

C) Sarcoidosis

D) Schistosomiasis

Training differential diagnosis by cases

Schistosomiasis: *How to approach?*



Anamnesis + Neuroimaging + Laboratorial tests

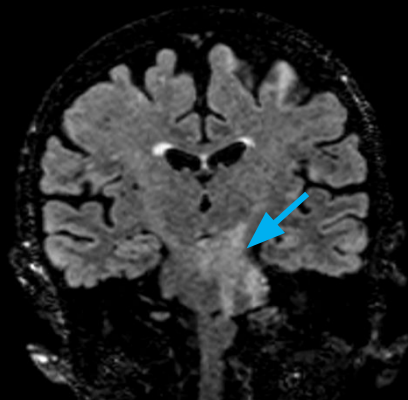
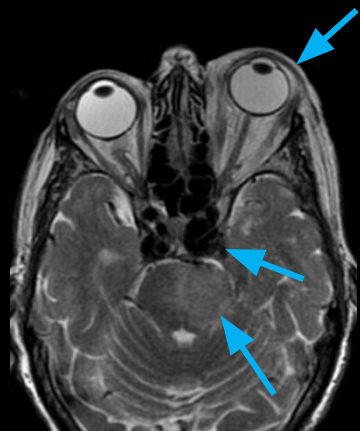
CSF: Eosinophilia (50%)

Urine culture / Coproculture: Eggs (50%)

Rectal Biopsy: Gold-Standard (95%)

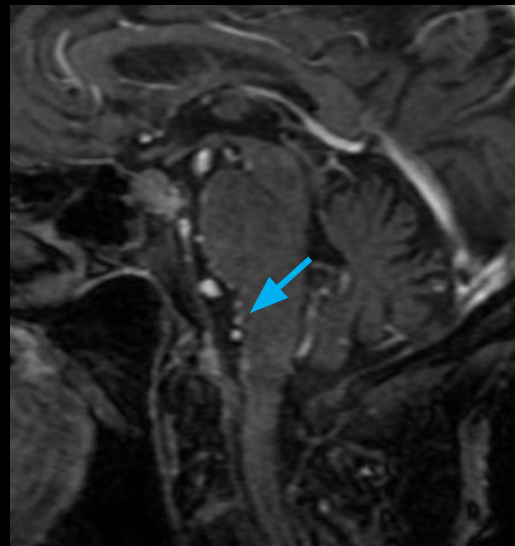
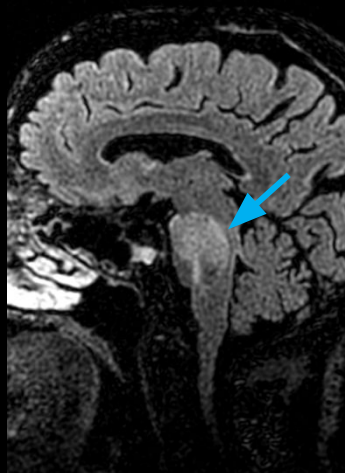
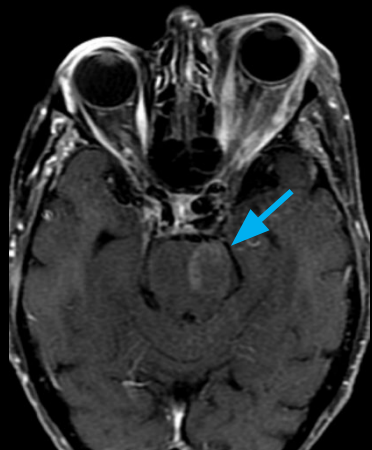
Hyperintense tumefactive T2
Gadolinium enhancement (multinodular,
leptomeningeal, cauda equina)

Training differential diagnosis by cases



Case 7: I'm back!

A 89 years old woman with previous history of ocular tuberculosis presenting with proptosis and lowered level of consciousness.



A) Tuberculosis

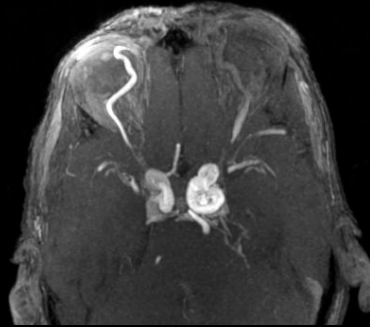
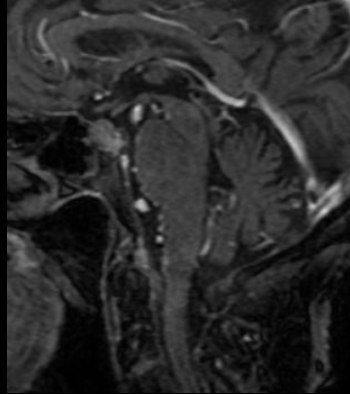
B) Listeria

C) Viral
rombencephalitis

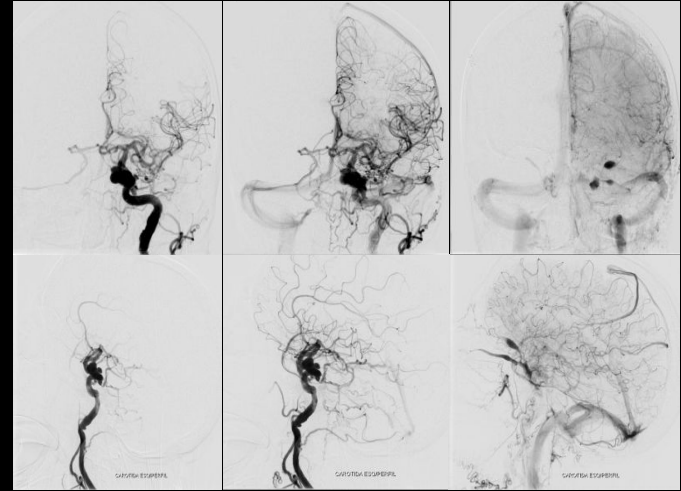
D) Carotid cavernous
fistula

Training differential diagnosis by cases

CCF: How to approach?



Arteriography of the case



Direct :

Traumatic (young male)
Acute symptoms

Proptosis / Retrobulbar fat edema
Cavernous sinus bulging + assymetric
enhancement, flow-voids,
hyperintensity on 3D-TOF
Proeminence of subarachnoid vessels
on prepontine cistern

Indirect :

Spontaneous
(postmenopausal female)
Insidious symptoms

Take home messages

- Behçet is a **multifaceted and multisystemic** disease and manifestations can arise in almost any tissue. Radiologic features can be challenging even for senior radiologists.
- Even though **Neuro-Behçet** is uncommon, radiologists usually can recognize some typical parenchymal and vascular impairment. Although, some atypical manifestations and differential diagnosis must be remembered.

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