

Cerebellar Tonsillar Herniation Following Acute Ischemic Stroke in Meningovascular Neurosyphilis: A Case Report

Prakash Gupta

Virgen Milagrosa University Foundation

Linh Nguyen

Saint James school of medicine

Raman Goit

goitraman@gmail.com

Kathmandu University

Prateeti Bekoju

Richmond University Medical Center

Manoj Argariya

East Lancashire Hospital Trust

Srivatsa Srirama Reddy

Mandya Institute of Medical Sciences

Parikshit Prasai

Kathmandu Medical College

Daniel Nguyen

University of California

Case Report

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Abstract

Introduction: Meningovascular neurosyphilis (MVNS) represents an uncommon manifestation of *Treponema pallidum* infection, potentially leading to stroke, cerebral edema, and elevated intracranial pressure. Cerebellar tonsillar herniation represents a rare yet critical complication with potentially life-threatening consequences. Timely diagnosis and intervention are critical to avert lethal consequences.

Case Presentation: A 20-year-old female presented with headache, dizziness, and altered sensorium, which were preceded by a self-limiting genital lesion eight weeks prior. She exhibited disorientation without any focal neurological deficits. The MRI indicated a left thalamic infarct, while cerebrospinal fluid analysis validated the presence of meningovascular neurosyphilis. Intravenous ceftriaxone and corticosteroids were administered for her treatment. On the 14th day of hospitalization, the patient experienced a generalized seizure accompanied by a rapid deterioration in consciousness. Subsequent imaging revealed widespread cerebral edema and herniation of the cerebellar tonsils. Despite receiving supportive care, she ultimately died due to complications arising from brainstem compression and increased intracranial pressure.

Conclusion: This case illustrates the significant consequences of MVNS resulting in infarction, cerebral edema, and tonsillar herniation in a young woman without a previous STD diagnosis. Early neuroimaging, cerebrospinal fluid analysis, and timely antimicrobial therapy are essential. The provision of IV penicillin and prompt surgical decompression are essential, particularly in resource-limited settings.

Key Clinical Message: This case shows how a thalamic stroke caused by meningovascular neurosyphilis can quickly lead to cerebral edema and a tonsillar herniation. Neurological problems need to be diagnosed, treated, and decompressed surgically as soon as possible to avoid permanent damage.

1. Introduction

Syphilis, caused by *Treponema pallidum*, can progress to neurosyphilis at any stage. It is more prevalent in high-risk groups, including men who have sex with men and individuals with human immunodeficiency virus (HIV) ¹. Early neurosyphilis may involve the meninges, cerebrospinal fluid (CSF), and vasculature ²; syphilitic meningitis and meningovascular forms are more common, and timely treatment along with earlier diagnosis can help prevent late progression to conditions such as tabes dorsalis ³.

While most cases resolve spontaneously and remain asymptomatic, approximately 5% progress to meningitis, which may present with seizures, aphasia, hemiplegia, or confusion. Meningovascular neurosyphilis (MVNS), a sub-type, typically occurs as early as six months to seven years after primary infection ⁴. It causes vascular inflammation, ischemic infarction, cerebral edema, and increased intracranial pressure (ICP) ⁵.

Cerebellar tonsillar herniation, an exceptionally rare but life-threatening consequence of elevated ICP, can result in brainstem compression, hydrocephalus, and fetal cardio-respiratory failure ⁶. Timely recognition and neurosurgical decompression are essential to prevent irreversible injuries ⁷.

We present a rare case of a 20-year-old female with neuro-psychiatric symptoms and altered sensorium, ultimately diagnosed with MVNS complicated by thalamic infarction, which progressed to diffuse cerebral edema and ultimately cerebellar tonsillar herniation. This case underscores the critical importance of early neuroimaging, CSF analysis, and prompt decompression to prevent life-threatening complications.

2. Case History and Examination

A 20-year-old female presented to our hospital with a sudden onset of a dull headache localized to the parietal region. The patient reported dizziness, nausea, vomiting, and orbital pain. The patient presented with a history of multiple episodes of watery vomiting, devoid of bile or blood. The patient reported no fever, visual disturbances, or prior history of trauma, diabetes, or hypertension. The patient's mother reported that eight months ago, the patient developed a painless, itchy lesion on her genitalia, which resolved spontaneously after two months without treatment. Two weeks ago, she had visited the emergency room with similar symptoms, which painkillers alleviated. The patient exhibited altered sensorium characterized by confusion, disorientation, amnesia, nervousness, personality changes, hostility, aggressive behaviors, hallucinations, and illusions. The patient's temperature measured 98.9°F, with a pulse rate of 100 beats per minute, a respiratory rate of 20 breaths per minute, blood pressure recorded at 110/90 mm Hg, and oxygen saturation at 94% in room air. On physical examination, pupils were equal, measuring 4 mm in diameter, and exhibited reactivity to light. She experienced disorientation regarding time, place, and person. Motor examinations reveal that muscle bulk, tone, and power in both upper and lower limbs are within normal limits. The plantar reflex, along with other reflexes, was intact. We did not conduct the sensory examination. Aside from the neurological examination, the examination of other systems revealed no significant findings.

3. Differential Diagnosis

At this point, given the patient's presentation with altered mental status, headache, visual hallucinations, and a prior painless genital lesion, the most probable differential diagnoses included viral encephalitis, bacterial or tubercular meningoencephalitis, autoimmune encephalitis and cerebral vasculitis.

4. Diagnosis, Investigation, and Treatment

Routine labs (CBC, RFT, LFT, BMP) were normal. TSH was elevated (16.28 mU/L), and Vitamin B12 was low (143 ng/L). Serology for HBsAg, HCV, and HIV was negative. RPR and TPHA tests confirmed syphilis. CSF analysis revealed 79 cells/ μ L (100% lymphocytes), high protein levels, low glucose levels, and numerous pus cells. Gram stain and culture results were negative; the CSF VDRL test was positive. AFB stain and HSV-1/2 PCR were negative. Findings are summarized in [Table 1].

An MRI of the brain revealed diffusion restriction in the postero-lateral region of the left thalamus, suggestive of an acute infarct [Figure 1]. The patient was admitted to the intensive care unit (ICU) with a provisional diagnosis of acute ischemic stroke with possible meningoencephalitis. Empirical antimicrobial therapy was initiated on the day of admission, including intravenous ceftriaxone, vancomycin, and acyclovir. On the following day, ampicillin was added to broaden coverage. In light of the ischemic infarct, she was started on aspirin and atorvastatin. Concurrently, levothyroxine 75 mcg daily was administered for hypothyroidism, and intramuscular methylcobalamin 1000 mcg was given to correct her vitamin B12 deficiency.

Following the cerebrospinal fluid (CSF) analysis results, acyclovir, vancomycin, and ampicillin were discontinued, and ceftriaxone was continued as monotherapy. Given the patient's young age and ischemic stroke, further investigations were conducted to rule out autoimmune and hypercoagulable states, including rheumatoid factor, antinuclear antibodies (ANA), serologies for HIV, hepatitis B, and C, as well as tests for natural anticoagulant levels and vasculitis markers. Based on positive VDRL and TPHA findings in both serum and CSF, the patient was diagnosed with neurosyphilis, specifically the meningovascular form.

Despite our best efforts, intravenous crystalline penicillin could not be administered due to its unavailability. The patient initially showed mild clinical improvement with ceftriaxone and corticosteroid therapy. However, on the 14th day of hospitalization, her condition deteriorated abruptly. She developed a generalized tonic-clonic seizure, after which her Glasgow Coma Scale (GCS) score declined to E2V3M2. The seizure was managed with intravenous midazolam, and she was immediately intubated and placed on supportive care.

A repeat MRI could not be performed on the same day of clinical deterioration. However, an MRI brain with MRA done the following day revealed significant findings, including tonsillar herniation, diffuse cerebral edema, and effacement of the visualized sulci and cisterns [Figure 2]. Neurological examination showed bilateral pupillary dilation to 4.5 mm and a positive Babinski sign, indicating severe brainstem involvement and raised ICP. Unfortunately, her neurological status continued to decline despite ongoing management, ultimately leading to her demise.

5. Conclusion and Results

Meningovascular neurosyphilis (MVNS), particularly in its early and aggressive form, may result in severe complications including ischemic infarction, diffuse cerebral edema, and cerebellar tonsillar herniation if not diagnosed and treated promptly. This case emphasizes the need to consider neurosyphilis and perform early interventions among young patients who present with altered sensorium and a history of genital lesions. Prompt CSF analysis and neuroimaging are crucial for assessing CNS involvement. Early intervention with appropriate antimicrobial therapy and management of complications, such as cerebellar herniation, often requires decompression surgery and is vital, involving a multidisciplinary approach. This approach can significantly improve outcomes, especially in resource-limited settings. The patient initially showed mild improvement but later developed seizures and a drop in GCS. Follow-up MRI revealed diffuse cerebral edema and cerebellar tonsillar herniation, leading to her death.

6. Discussion

Our case highlights a rare and severe complication post-neurosyphilis, a cerebellar tonsillar herniation secondary to diffuse cerebral edema and ICP, following an ischemic stroke in a young female with no prior diagnosis of a sexually transmitted infection (STD). Syphilis is an important but often overlooked cause of ischemic stroke, especially in young patients. According to a nationwide retrospective cohort study by Chang et al. (2022), it showed a 35% increased risk of stroke in patients with syphilis compared to matched controls. This supports the association between syphilis and cerebrovascular events⁸. Similarly, a Brazilian tertiary center found 13% of stroke patients had positive syphilis serology, and 4.7% met criteria for neurosyphilis, despite most having negative CSF-VDRL⁹.

Ischemic strokes are increasingly affecting individuals under the age of 50. Rare disorders collectively account for 22% of cases¹³ [Table 2]. MVNS, which affects 15% to 30% of neurosyphilis cases and usually manifests 7 years after infection, can cause ischemic stroke¹⁰. *Treponema pallidum* penetrates the central nervous system (CNS) hematogenously shortly after inoculation and stays latent¹¹. Later, it results in inflammation of small vessels (Nissl endarteritis) and medium to large arteries (Heubner endarteritis), which primarily affects the anterior circulation. The natural development of untreated neurosyphilis over time, including its different neurological symptoms like meningitis, MVNS, general paresis, and tabes dorsalis, is shown in [Figure 3], just like in our patient who did not get treatment on time. Prodromal symptoms like headache and malaise often precede onset¹². Prior to making the diagnosis, serologic treponemal tests such as the fluorescence treponemal antibody test, absorption test, or EIA are used to confirm the existence of a syphilis infection. The RPR and TPHA tests that were conducted on our patient produced positive findings. The next step should be to undergo a lumbar puncture. Neurosyphilis is consistent with pleocytosis, elevated protein content (>45 mg/dL), or a positive CSF VDRL test¹², as conducted in our patient. The vascular involvement may have been exacerbated by a prolonged, undetected infection.

Although cerebellar tonsillar herniation due to stroke is a well-documented phenomenon, secondary to stroke caused by MVNS, it has been rarely reported. Tonsillar herniation is a severe outcome of elevated intracranial pressure, typically arising from traumatic brain injury (TBI), which impacts around 69 million people worldwide annually¹⁴. Additional causes encompass intracerebral and subarachnoid hemorrhages. The precise incidence of tonsillar herniation remains undetermined, as it serves as a physiological endpoint for multiple underlying diseases. The Monroe-Kellie concept posits that the cranial cavity contains fixed amounts of brain tissue (1400 mL), cerebrospinal fluid (150 mL), and blood (150 mL)¹⁵. As one component rises, others must diminish to preserve equilibrium. Initially, cerebrospinal fluid and venous blood are displaced for compensation. Once these processes are depleted—termed loss of intracranial compliance—cerebral tissue may herniate through the foramen magnum. This results in medullary compression, which may lead to respiratory arrest, cardiac failure, coma, and death¹⁶. Therefore, herniation occurs when the cerebellar tonsils shift down through the foramen magnum, putting pressure on the medulla against the clivus or odontoid. This “coning” effect triggers Cushing’s reflex and can rapidly progress to death¹⁷. This could be a possible

cause of death of our patient. Tonsillar herniation generally represents a final phase of significant intracranial pressure, particularly with brainstem compression and respiratory failure¹⁶. Nonetheless, prompt identification and timely surgical decompression may enhance results and avert mortality, as described by Cushing⁷. Our patient, a young woman with no traditional risk factors, was diagnosed with MVNS based on CSF findings, resulting in thalamic infarct ultimately progressing to diffuse cerebral edema and tonsillar herniation.

The tragic outcome of the patient's condition highlights the critical nature of timely intervention in cases of escalating intracranial pressure. The shift from a thalamic infarct to widespread brain swelling and then to tonsillar herniation shows the serious dangers and difficulties associated with MVNS. This case serves as a reminder that prompt identification and surgical decompression are not merely procedural steps but vital actions that can significantly alter the course of patient outcomes. The unfortunate demise due to cerebellar tonsillar herniation illustrates the urgent need for awareness and swift response in managing neurological emergencies, emphasizing that every moment counts in preserving life when faced with such severe complications.

Declarations

Consent: During admission, the patient and her father gave consent for all the procedures and data collection. The authors have obtained written informed consent from the patient's legal guardian for publication.

Conflict of Interest:

No

Disclosure:

A preliminary version of this case was previously published in Dhulikhel Medical Update, concentrating on the diagnosis of meningovascular neurosyphilis. This report, submitted to Wiley Clinical Case Reports, offers a novel and essential perspective on cerebellar tonsillar herniation as a rare, life-threatening complication that was not addressed in the previous publication.

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Author Contribution

1-Prakash Gupta: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Project administration, Supervision, Validation, Visualization, Writing – original draft, Writing – review & editing
2-Linh Nguyen: Conceptualization, Investigation, Methodology, Software, Validation, Visualization, Writing – original draft, Writing – review & editing
3-Raman Goit: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Software, Validation, Writing – original draft
4-Prateeti Bekoju: Conceptualization, Formal analysis, Investigation, Supervision, Validation, Writing – original draft
5-Manoj Argariya: Writing – original draft
6-Karan Rana: Writing – original draft
7-Srivatsa Srirama Reddy: Writing – original draft
8-Parikshit Prasai: Writing – original draft
9-Daniel Nguyen: Writing – original draft

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Tables

Table 1: Comprehensive Cerebrospinal Fluid (CSF) Analysis, WBC: White Blood Cell, ADA: Adenosine Deaminase, AFB: Acid Fast Bacilli, VDRL: Venereal Disease Research Laboratory, HSV: Herpes Simplex Virus, DNA: Deoxyribonucleic Acid, PCR: Polymerase Chain Reaction

Parameter	Result	Reference Range
Volume	1 ml	
Color	Watery	
Transparency	Clear	
Total WBC Count	76 cells/ μ L	0-5 cells/ μ L (normal)
Differential Count		
Neutrophils	0%	0–6%
Lymphocytes	100%	40–80%
Monocytes	0%	15–45%
Eosinophils	0%	0%
Basophils	0%	0%
Protein, CSF	155 mg/dL	Adults <60 yrs: 15–45; >60 yrs: 30–60 mg/dL
Sugar, CSF	10 mg/dL	40–80 mg/dL
ADA	9 U/L	CSF: <10.0 U/L
Gram Stain	Pus cells present; no microorganisms	
Culture and Sensitivity	No growth after 48 hours at 37°C	
AFB Stain	No acid-fast bacilli seen	
VDRL (CSF)	Reactive	
HSV-1 and HSV-2 DNA PCR (CSF)	Negative	

Table 2. Etiological Classification of Stroke in Young Adults, HIV: Human Immunodeficiency Virus, COL4A1: Collagen Type IV Alpha 1 chain

Medications and substance use	Cardiac and structural associations	Important nonstructural associations	Genetic and acquired thrombophilia	Inflammatory vasculopathies	Noninflammatory vasculopathies	Infectious disorders	Monogenic disorders
Oral contraceptives	Patent foramen ovale	Migraine with aura	Factor V Leiden mutation	Primary central nervous system vasculitis	Cervical artery dissection	Infective endocarditis	Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy
Alcohol use	Congenital heart disease	Pregnancy	Prothrombin G20210A mutation	Systemic vasculitis	Intracranial artery dissection	Meningoencephalitis	Cerebral autosomal recessive arteriopathy with subcortical infarcts and leukoencephalopathy
Cannabis	Valvulopathies	Malignancy	Protein C deficiency		Radiation-induced vasculopathy	HIV	COL4A1 disorders
Cocaine	Intracardiac tumors		Protein S deficiency		Reversible cerebral vasoconstriction syndrome	Syphilis	Fabry disease
Amphetamines			Antithrombin III deficiency		Moyamoya disease or syndrome	Tuberculosis	Hereditary hemorrhagic telangiectasia
Opiates			Antiphospholipid syndrome				Homocystinuria
							Marfan syndrome
							Sickle cell disease

Figures

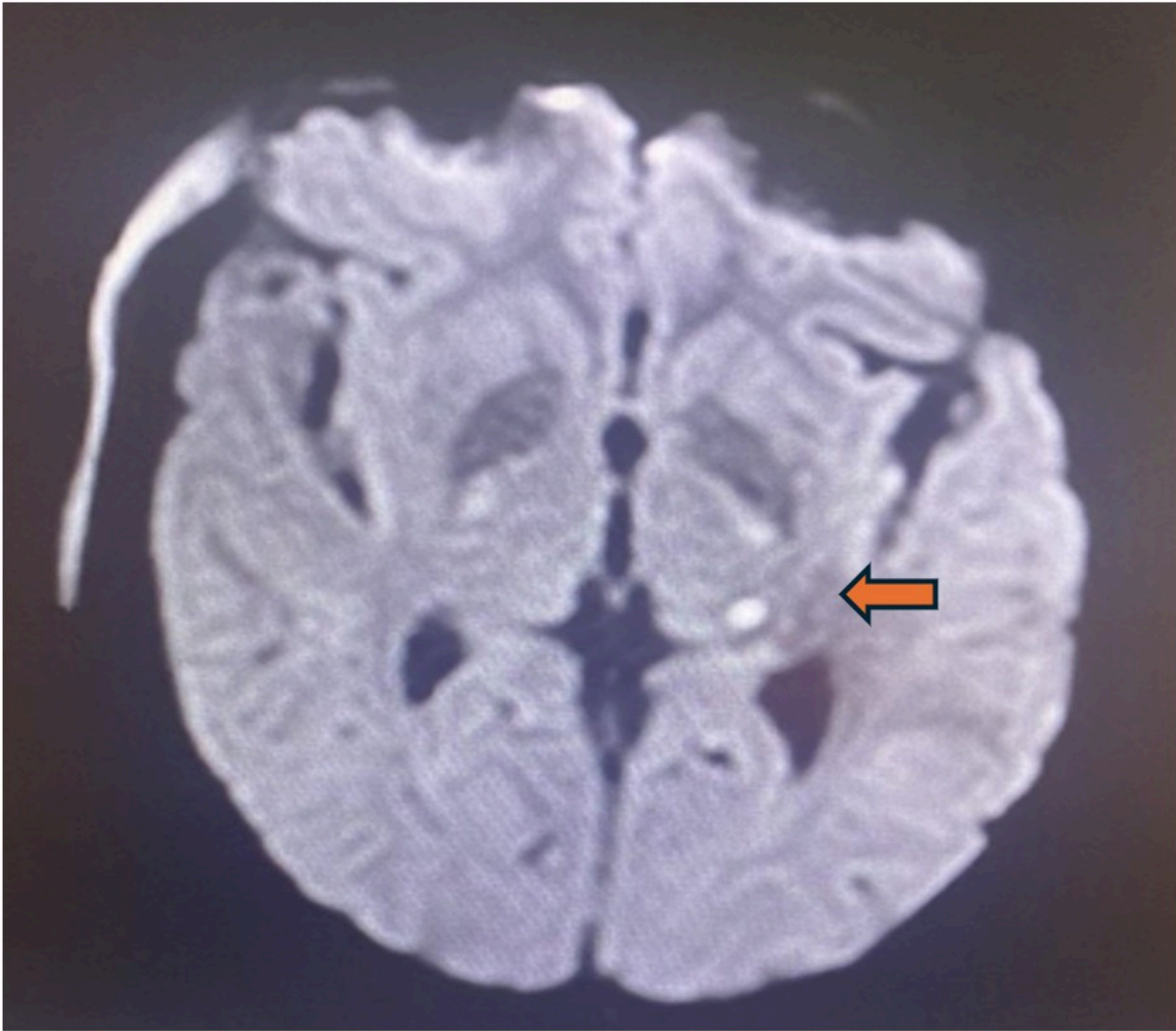


Figure 1

Axial DWI image shows diffusion restriction at posterolateral aspect of left thalamus s/o infarct (red arrow).

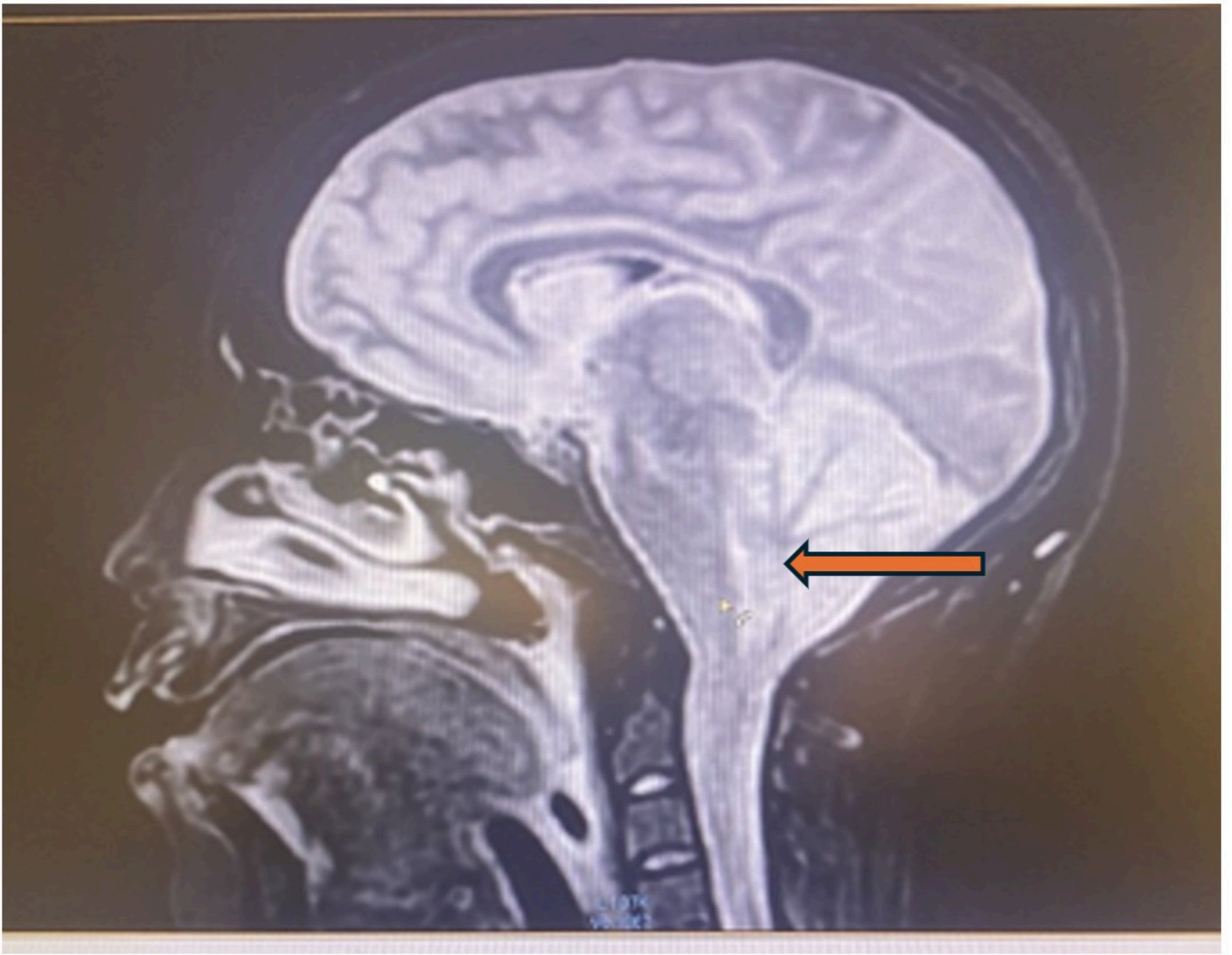


Figure 2

Sagittal FLAIR image shows tonsillar herniation, effacement of visualized sulci and cisterns and diffuse high signal intensity in cerebral and cerebellar cortex consistent with diffuse brain edema (red arrow).

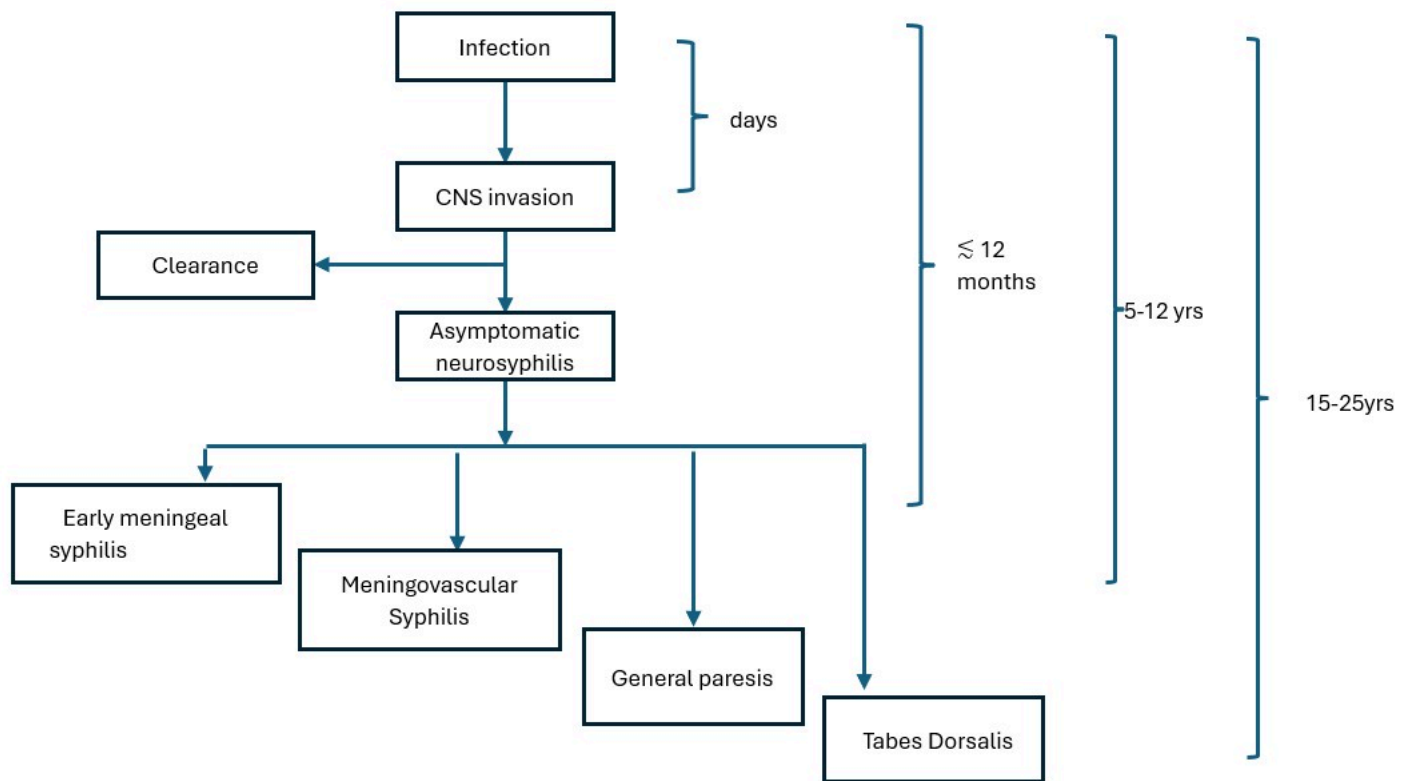


Figure 3

Neurological complications of syphilis with estimated timeframes indicating the average duration to progression in the absence of medical treatment