Radiology Case Reports 18 (2023) 2987–2991

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Locked-in syndrome caused by extracranial and intracranial takayasu arteritis: A rare case report and radiological findings ☆

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ARTICLE INFO

Article history:
Received 10 May 2023
Accepted 13 May 2023

Keywords:
Takayasu's arteritis
Locked-in Syndrome
Stroke
Brainstem
Cardiovascular disease

ABSTRACT

Locked-in syndrome (LiS) is a condition of motor paralysis involving all the voluntary muscles of 4 extremities with retained consciousness and alertness. Meanwhile, Takayasu arteritis (TA) is a systemic inflammatory disease affecting large arteries, including the aorta and the branches. This disease often affects the extracranial arteries but rarely occurs in the intracranial arteries. An 18-year-old male presented with a sudden onset of decreased consciousness and bilateral weakness. The patient exhibited normal horizontal and vertical eye movements but presented with impairment of the trigeminal nerve, peripheral-type facial palsy, absence of gag reflex and tongue movement, and neck weakness. Physiological reflexes were increased bilaterally, while pathological reflexes were present on both sides. Autonomic function was impaired, and communication was impossible due to paralysis. Additionally, the patient displayed varying blood pressure readings between the right and left arms. The antinuclear antibody (ANA) and antismooth muscle (ASMA) antibody tests yielded negative results, while the electroencephalogram (EEG) showed normal readings. The cerebral angiogram indicated multiple internal and external stenoses, with total stenosis evident in the basilar artery. The patient was diagnosed with multiple extracranial and intracranial stenoses due to TA. Total stenosis of the basilar artery resulted in brainstem infarction, which led to the development of LiS. Meanwhile, the disturbance of the motor tracts in the ventral brainstem was the underlying cause of the LiS. This case report demonstrated a variety of atypical presentations of TA. The involvement of multiple extracranial and intracranial arterial was attributed to LiS.

Abbreviations: LiS, Locked-in syndrome; TA, Takayasu arteritis; MCA, Medial Cerebral Artery; PCA, Posterior Cerebral Artery; RVA, Right Vertebral Artery; LVA, Left Vertebral Artery; LSA, Left Subclavian Artery; LECA, Left External Carotis Artery.

☆ Competing Interests: None.

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https://doi.org/10.1016/j.radcr.2023.05.039

Introduction

Locked-in syndrome (LiS) is a medical condition characterized by complete motor paralysis that affects all four voluntary extremity muscles, including dysarthria, with retained consciousness and alertness [1]. Patients with LiS are capable of opening their eyes and exhibit normal cognitive function, but their communication is limited to vertical or lateral eye movements or blinking of the eyelids [2]. Similarly, Takayasu arteritis (TA) is an uncommon idiopathic systemic inflammatory disease affecting large arteries, such as the aorta and large vessels [3]. The diagnosis is challenging, especially in the early stages, as it may only present with nonspecific systemic inflammatory symptoms [4]. Therefore, the diagnostic criteria for TA rely on a combination of physical examination, laboratory, and imaging findings [5].

Due to the disease's rarity, TA has limited incidence data. People of any ethnicity can experience TA. It is worth noting that people of any ethnicity can experience TA, but it is generally more prevalent among Asian populations [6]. Even though extracranial vascular involvement is common, intracranial involvement in TA is rarely reported. This study aims to present a rare case of extracranial and intracranial involment in TA that caused LiS and to describe the radiological findings.

Case presentation

An 18-year-old male presented with an abrupt decrease in consciousness. The patient exhibited right-sided weakness initially, which progressed to involve both sides of the body within 24 hours. The level of consciousness improved on the third day of treatment and became fully awake. Subsequently, the blood pressure was different between the right (108/78 mm Hg) and left arms (87/60 mm Hg). Clinical examination showed trigeminal nerve impairment, peripheral-type facial palsy, absence of gag reflex and tongue movement, neck weakness, and double hemiparesis. Physiological reflexes were also found to be increased bilaterally, while pathological reflexes were present on both sides. Autonomic dysfunction was noted, and the patient was unable to communicate due to paralysis.

A normal cerebrospinal fluid analysis was obtained, and a blood examination showed mild leucocytosis. An electroencephalograph examination did not show any slowing or epileptiform activity. In the emergency room, a contrasting head CT scan was performed, which indicated leptomeningeal and gyral enhancement on the right and left front-parietal-temporal-occipital contrast regions accompanied by brain edema Fig. 1.

The result of the cerebrospinal fluid analysis indicated normalcy. The tests for antinuclear antibody (ANA) and anti-smooth muscle (ASMA) antibody showed negativity. Based on the Magnetic Resonance Imaging (MRI) findings, the patient had a chronic thrombotic infarction in the central region of the Pons. Additionally, the Magnetic Resonance Angiography revealed severe stenosis of the mid-bacillary artery, mild stenosis in the distal M1 segment of the left Medial Cerebral Artery (MCA), and stenosis of the P1 segment of the right Posterior Cerebral Artery (PCA) as depicted in Fig. 2.

Cerebral angiography was performed, reporting wall thickening of the aortic arch and proximal great vessels. It also in-

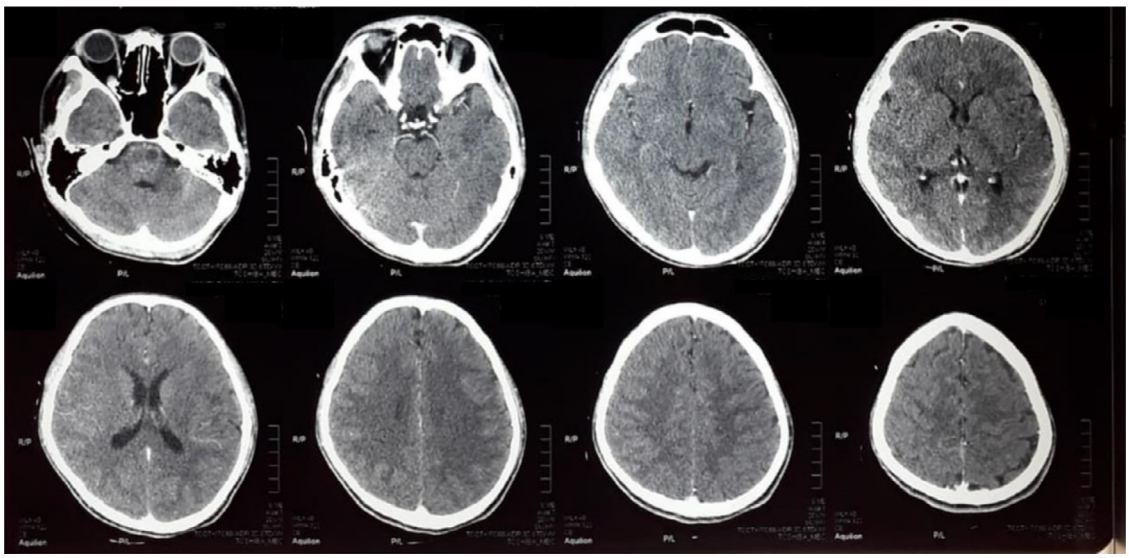

Fig. 1 – Contras head CT scan showed leptomeningeal and gyral enhancement on the administration of right and left front-parietal-temporal-occipital contrast regions accompanied by brain edema. CT Scan was not showing infarction in the brainstem area.

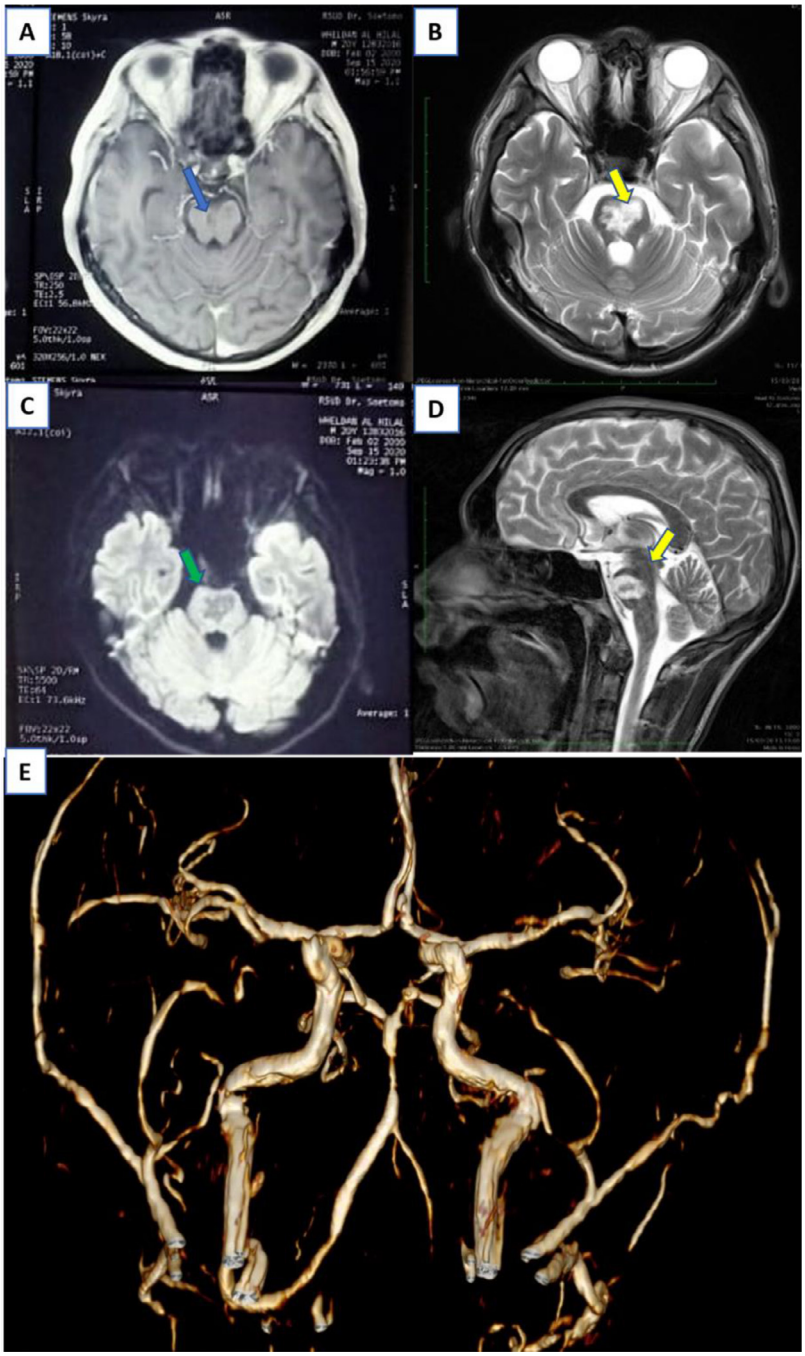

Fig. 2 – Head MRI revealed a central pons lesion. (A) Hypointese in T1-Weighted images (blue arrow) (B&D) Hyperintense in T2-Weighted images (yellow arrow) (C) Unrestricted diffusion area in Diffused Weighted Images (green arrow) (E) MR Angiography showed basilar artery stenosis and mild stenosis in distal segment M1 of left MCA.

dicated moderate stenosis on the proximal Right Subclavian Artery (RSA) and distal RSA. Contrast injection in the Right Vertebral Artery (RVA) showed mild to moderate stenosis on the proximal RVA and the blood flow to the Left Vertebral Artery (LVA). A proximal Left Subclavian Artery (LSA) occlusion was found, and flow to the LSA was from the thyrocervical artery. Furthermore, the Inferior Thyroid Artery was anastomosed with the contralateral artery. During contras injection on the Left External Carotis Artery (LECA), an anastomose from the occipital to the Left Vertebral Artery was found. In addition, cerebral angiography reported multiple intracranial and extracranial stenosis leading to TA Fig. 3.

This patient was treated with conservative medical management and received oral Methotrexate and planned physical rehabilitation. After1 month of treatment, the patient was discharged.

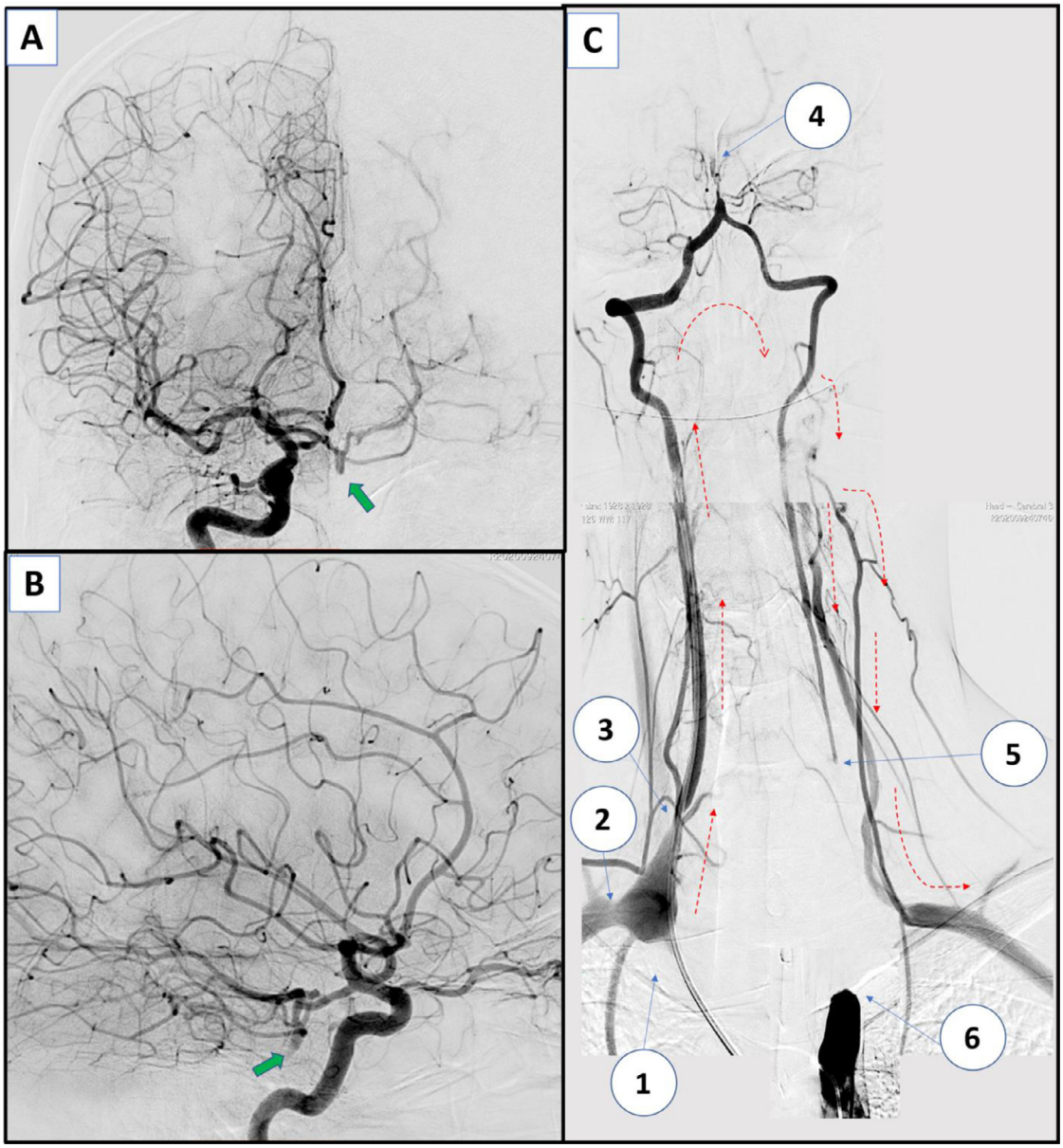

Fig. 3 – Although the MRA showed stenosis in the Basilar Artery, the cerebral angiography confirmed total Mid-basilar Artery occlusion. (A&B) Cerebral angiography showed flow from the right-anterior circulation (anteroposterior and lateral view) to the distal Basilar Artery (Green Arrow) through the Posterior Communicating Artery. (C) Cerebral angiography also revealed stenosis of the Brachiocephalic Trunk (arrow 1), proximal RSA (arrow 2), proximal RVA (arrow 3), total occlusion Mid-basilar Artery (arrow 4), total occlusion LVA originated from the ostium (arrow 5), and total occlusion LSA (arrow 6). The red dot arrow showed flow from the right to the left subclavian artery.

Discussion

Takayasu's arteritis (TA) involving intracranial vascularity is a rare occurrence [7], and one of the potential complications is stroke [7]. In cases where patients experience stroke due to TA, they may remain aware and alert of the environment but unable to speak or move their limbs. Instead, the patients may only be able to move the eyeballs and blink the eyes due to LiS. This condition arises as a disorder in the motor tracts located in the ventral brainstem, which is a secondary cause of the rare illness known as LIS [1]. It is essential to note that LiS is seen in approximately 14.4% of patients who experience basilar artery occlusion [8].

Cerebral Angiography is a safe diagnostic tool used to identify vascular malformations of the brain [9]. The examination shows stenosis of the extracranial and intracranial arteries. Flow from the anterior circulation appeared to fill the posterior part but did not reach the Basilar artery due to complete stenosis. As a result, there was no blood flow to the brainstem

and pons, leading to infarction in these areas, and death of several cranial nerve nuclei and motor pathways. In this patient, infarction occurred in the brainstem and pons area and caused the death of several cranial nerve nuclei and motor pathways. Infarction of cranial nerve nuclei can cause trigeminal nerve disorders, peripheral-type facial palsy, dysphagia, and vestibular disorders [10,11]. LiS was clinically observed, and the presence of mild to moderate LSA caused a reduction in blood flow to the right arm. Conversely, a contrast injection in the RVA caused flow-through of the RVA and counterflow to the LVA, which explains the difference in blood pressure of the patient's arms.

This case report highlights a rare occurrence of LiS in multiple intracranial and extracranial stenosis in TA. Meanwhile, intracranial involvement by the arthritic process is infrequent, stroke is a significant neurological consequence that carries a poor prognosis with persistent neurological impairment [12]. The treatment of TA aims to control inflammation with medications and prevent further blood vessel injury.

Conclusion

In conclusion, this case demonstrates a range of rare presentations of TA. Large extracranial arteries are most commonly involved, and this patient has multiple intracranial involvements. Furthermore, the associations between TA and LiS are also discussed.

Patient consent

A written, informed consent was obtained from the patient/legal representative for the publication of this case report (including all data and images).

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