Case Report

Acute spinal cord ischemia in a patient with Situs Inversus Totalis

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ABSTRACT

Situs inversus totalis (SIT) is a rare developmental abnormality where the organs throughout both the thoracic cavity and abdomen are a mirror image of normal anatomy, often occurring concomitantly with other genetic and developmental defects. Acute spinal cord ischemia is diagnosed based on the clinical presentation along with consistent imaging, but since clinical manifestations of acute spinal cord ischemia—rapidly progressive motor, sensory, and autonomic dysfunction—overlap with a wide spectrum of myelopathies, a thorough diagnostic workup with consideration of inflammatory, infectious, compressive and nutritional etiologies is required to establish the diagnosis. In this report, we present the case of an 18-year-old female patient who was admitted with acute onset of severe lower back pain, progressive weakness, paralysis, loss of sensation in both lower limbs and void-
Introduction

The human body appears symmetrical from the outside; if a line is drawn down the center of the body, each side appears similar. The internal anatomy, on the other hand, is not like that. One heart, eg, is located in the left chest, 1 liver is located in the right abdomen, and 1 stomach is located in the left abdomen. The term situs refers to an organ's position or location in relation to the body's midline, specifically the atria and abdominal viscera [1,2]. The term "Situs Inversus Totalis" refers to the reversal of the natural location of the internal organs in a mirror image [1]. Situs Inversus Totalis (SIT) is estimated to occur in about 1 in every 12,019 pregnancies or 0.83 per 10,000 pregnancies [3]. SIT may appear as an isolated finding with no apparent symptoms or as part of a complex syndrome or associated with other diseases. SIT occurs in association with different syndromes like Kartagener's syndrome, duodenal atresia, different types of tumors, etc. Neurological anomalies associated with situs inversus totalis are very rare.

Spinal cord ischemia is a rare neurological condition, accounting for 0.3% - 1% of strokes and approximately 5% - 8% of myelopathies [4,5]. Its prevalence appears to be the same among men and women and common risk factors associated with strokes, such as hypertension, elevated blood glucose, diabetes mellitus and atherosclerosis are present in the majority of patients [4,5]. It arises from an acute obstruction in the normal blood supply to the spinal cord, constituted by the anterior spinal artery (ASA) and the 2 posterior spinal arteries (PSA) [4,5].

The clinical presentation is variable, consistent with the topography of the affected segment of the spinal cord; however, the classic symptoms include abrupt onset of painless paraplegia and urinary retention [4-6]. Paresthesia and dysesthesia may also be present [4-6]. When spinal cord injury occurs in the context of generalized hypoperfusion, clinical manifestations include flaccid paralysis, followed by hyperreflexia and spasticity [4-6].

Clinical presentation and imaging modalities, with MRI being the modality of choice, establish the definitive diagnosis. The differential diagnosis includes compressive (extramedullary tumors, abscess, hematoma), infectious (HIV, HTLV-1, Varicella), inflammatory (transverse myelitis, multiple sclerosis) and nutritional myelopathies (B12 deficiency) [4].

Guidelines on the management of acute spinal cord ischemia have yet to be established, however in current clinical practice, treatment is focused on the primary etiology of the injury [4]. Given that a number of patients are affected following aortic surgery, CFS drainage, CFS and epidural pothermia, as well as monitoring motor and somatosensory functions and maintaining an adequate mean arterial pressure are often employed to improve cord perfusion and prevent postoperative neurological complications [4].

Case report

An 18-year-old female patient is referred to the Neurology Department with acute onset of severe lower back pain, progressive weakness, paralysis and loss of sensation in both lower limbs and voiding difficulties. The symptoms developed within 24 hours, on the day of admission.

On physical examination, the patient was alert, oriented to person, place and time. Examination of the pulmonary, cardiovascular, musculoskeletal systems was unremarkable, with no significant findings. The abdomen was soft, nontender and nondistended, with no presence of rebound or guarding.

The neurological examination revealed a GCS of 15 points, vision acuity was normal with no visual field defects, bilaterally. Both pupils were round, equal and reactive to light. Cerebellar examination was normal. Motor and sensory functions, as well as reflexes of the upper extremities were all within normal limits. Examination of the lower extremities revealed bilateral weakness and hypotonia, loss of motor strength 0/5, bilateral tactile hyperesthesia up to the T6-T8 level, thermal hypoesthesia and perineal and perianal hypoesthesia. Areflexia, with no pathological reflexes was evident. No signs of a meningeal syndrome or focal neurological deficits were present.

Routine laboratory examinations were all within normal limits. Infectious diseases serology for Borrelia Burgdorferi, HSV-2, West Nile Virus, Toxoplasma gondii and Covid-19 were all negative. Serologic examination for Rubella and Cytomegalovirus were positive. Personal and family medical history were unremarkable.

Additionally, a whole spine MRI was ordered. It showed a disc herniation at the T6-T7 level with compression of the spinal cord, extraforaminal disc protrusion at the left L2-L3 level, asymmetrical disc bulging at the L1-L2 level. Schmorl nodes were evident in the T1, L1-L3 vertebrae.

MRI of the cervical and thoracic spine (Fig. 1) revealed an increase in T2 and STIR signal intensity in the spinal cord. Diffusion weighted MRI (DW-MRI) showed restricted diffusion in the anterior segment of the spinal cord from T3-T10 and slightly above the medullary cone, consistent with acute ischemia. Partial fusion of the posterior segments of C5 and C6 vertebral bodies was present. A contrast-enhancing lesion in
the level of T11-T12 and the intervertebral disc, suggestive of inflammatory changes were evident. Hyperintense spinal cord signals in the level of T11-L1, on axial T2/T2FS images were present as well. No signs of expansive lesions, traumatic lesions or fractures, metastasis, piriformis syndrome or sacroiliitis were visible.

Additionally, a CT angiogram (Fig. 3 and Fig. 4) was performed in the same visit, revealing the presence of Situs Inversus Totalis with dextrocardia and transposition of the great vessels, the aorta and the aortic arch located on the right side of the chest, as well as mirror-image location of the abdominal viscera.

Subsequently, a second MRI eight months later (Fig. 2) was ordered and revealed the changes in cervical segment and trunk which were lesion-free (partial fusion of C5 and C6 in segmented dorsal). From T3 to T10/T11 emphasized atrophic changes in the previous segment of spinalis medulla, with gliosis in the distal segment and areas of syringomyelia. Lesions are sequels of the vascular accident, very possible post COVID-19.

Fig. 4.

At first, a diagnosis of transverse myelitis was suspected. However, on account of the clinical presentation and the imaging findings, a diagnosis of acute spinal cord ischemia with Situs Inversus Totalis was established. The patient was managed conservatively with high-dose corticosteroid therapy.

### Discussion

In our patient, the diagnosis of situs inversus totalis (SIT) was established incidentally, due to the acute spinal cord ischemia. To the best of our knowledge, this is the first report of an association of SIT with acute spinal cord ischemia.

An asymmetry noted on X-ray imaging of this patient led the clinicians to perform a CT-angiogram. Situs inversus was diagnosed after visualization of the aortic arch on the contralateral side. This inversion occurs during embryogenesis, as a result of mutations in genes on chromosome 12 [7-10].

SIT patients are phenotypically unaffected, unaware of their medical condition as it does not impact their quality of life or lifespan. Nevertheless, some cases have been linked with cardiac and vascular anomalies, associated with amplified risk of heart, spleen and hepatobiliary malformations [11].
Fig. 2 – T2 sagittal plane of medulla spinalis eight months later. (A). The cervical segment and trunk are lesion-free (partial fusion of C5 and C6 in segmented dorsal) (a, yellow arrow). (B and C). From T3 to T10/T11 emphasized atrophic changes in the previous segment of spinalis medulla, (A and B, green arrows) gliosis in the distal segment and areas of syringomyelia. Lesions are sequels of the vascular accident, very possible post COVID19 (Color version of figure is available online).

Fig. 3 – Thoracoabdominal pelvic computed tomography CT scans (A, B) reveal situs inversus with multiple mediastinal and abdominal adenopathies. Dextrocardia and right sided aorta (red arrow) are demonstrated. The stomach bubble is seen in the right upper abdominal region (blue arrow), liver on the left (white arrow), and spleen on the right (yellow arrow) (Color version of figure is available online)
Spinal cord ischemia is caused by an acute disruption of the normal cord vascular irrigation. The spinal cord supply has been divided into 3 different sections, the first 1 extends from C1 to T3 and derives its blood supply from the vertebral arteries and at the level of C6 and C7 from the cervical ascending arteries [4–6]. The second section extends from T3 to T7 and is supplied by a left-sided intercostal artery. [4–6] The third section from T8 to the conus receives its supply mainly from the Adamkiewicz artery and sometimes, a cone artery originating from the internal iliac artery at the L2 or L5 level [4–6].

Studies show there are two patterns of spinal cord ischemia: radicular artery territory ischemia associated with bilateral anterior or posterior spinal artery infarcts and unilateral infarcts, frequently occurring after a mechanical triggering event and extensive spinal cord hypoperfusion, associated with central and transverse infarcts, in the event of prolonged hypotension [6].

Acute spinal cord ischemia is usually spontaneous, owing to atherosclerosis, cardioembolism, small-vessel occlusion, aortic pathology, degenerative spine disease and idiopathic etiology, or iatrogenic following aortic surgery, orthopedic lumbar surgery and on rare occasions, percutaneous procedures [4–6].

Some patients have a biphasic clinical presentation, initially manifesting transient sensory deficits and subsequent deterioration [4,5].

MRI revealing a hyperintense signal on T2-weighted images, a gadolinium-enhancing cord lesion or a characteristic high signal in diffusion-weighted imaging (DWI-MRI) is the diagnostic modality of choice [4].

Studies reveal that prognosis of spinal cord ischemia is better than previously suggested [12]. Risk factors such as atherosclerosis, diabetes mellitus, hypertension and male sex are associated with worse outcomes [4,5,12]. Spinal cord ischemia is more prevalent in younger females, compared to cerebral ischemia, as was the case of our patient [5].

In our case report, the patient was 18 years old and presented with hallmark features of acute spinal cord ischemia including lower back pain, rapidly progressive bilateral weakness, paralysis and loss of sensation in lower limbs and voiding difficulties. Given that situs inversus totalis does not impact the quality of life and the patient's lifespan, prognosis is determined by the neurological deficits and sequelae associated with spinal cord ischemia, along with any other possible concomitant comorbidities.

Patient Consent Statement
We obtained written, informed consent for publication from the patient.

Conclusion
Acute spinal cord ischemia is a rare neurological entity, caused by an acute disruption of the normal cord vascular irrigation. Situs Inversus is a rotational anomaly where the abdominal viscera are situated in a mirrored position of its normal, and when it furthermore involves the thoracic organs it is called Situs Inversus Totalis (SIT). In general, it is an autosomal recessive condition, but it can be X-linked. Most importantly it can be accompanied by different pathologies. There have been previous case reports of Situs Inversus Totalis (SIT) with coexisting pathological conditions.

Nevertheless, to the best of our knowledge, our case report represents the first report of acute spinal cord ischemia with concomitant situs inversus totalis. It underscores the heterogeneity of the pathological conditions that may be associated with situs inversus totalis and the need for a thorough and meticulous clinical and radiological evaluation of these patients, as establishing even common diagnosis may be challenging in this population.

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