ISSN: 2836-5968

Research Article

Journal of Neuro and Spine

Spontaneous Upper Cervical Cord Infarction: A Cohort of 13 Cases and Review of the Literature

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Submitted: 2023, Aug 01; **Accepted:** 2023, Sep 02; **Published:** 2023, Sep 12

Citation: Dokponou, Y. C. H., Belghacham, H., Sahri, I. E., Slimani, S., Hamid, M., et al. (2023). Spontaneous Upper Cervical Cord Infarction: A Cohort of 13 Cases and Review of the Literature. *J Neuro Spine, 1*(3), 77-82.

Abstract
The purpose of this study was to evaluate the risk factors, etiologies, clinical features, and functional outcomes of 13 consecutive patients who were admitted to our institution with a spontaneous upper cervical cord infarction (SUCI), a rare and devastating condition. A literature review of variables relevant to our case series was conducted.

***Methods:** We revised all spinal cord stroke cases admitted between 2005 and 2020 to the Department of Neurology of the University Hospital Center "Mohammed V Military Teaching Hospital" of Rabat (Morocco). Data on the patient's demography, clinical features, and functional outcomes of the upper cervical cord infarction patients were analyzed in Jamovi software for descriptive analysis.*

***Results:** Abdominal aortic aneurysm repair n=5(38.5%) and aortic dissection secondary to giant cell arteritis n=5(38.5%) were the possible etiology found. Nadir's deficits were ASIA grade A in 4(30.8%) patients, grade B in 2(15.4%), C in 4(30.8%) and D in 3(23.1%). The Modified Rankin Scale at admission was mRS3 in 2 patients 15.4%, mRS4 in 7(53.8%), and mRS5 in 4(30.8%). At different stages of follow-up, three patients, 23.1% had died. In the other 10 patients, the residual deficits were ASIA grade B in (1/10) patients, D in (3/10), and E in (6/10). The recovery and degree of continued disability over time were mRS1 in (6/10) patients, and mRS2 in (4/10).*

***Conclusion:** The overall better outcome observed in this study can be related to the fact that more than half of the patients in our cohort had nadir ASIA C/D with mRS 3-4.*

Keywords: Upper Cervical Cord, Cervical Infarction, Risk Factors, Etiology, Predictors of Outcome.

Abbreviations
SUCI: Spontaneous Upper Cervical Cord Infarction
SCI: Spinal Cord Infarction
ASIA: American Spinal Injury Association Impairment Scale
MRS: Modified Rankin Scale
FCE: Fibrocartilaginous Embolism

1. Introduction
Spinal cord infarction (SCI) is often disabling, and the diagnosis can be challenging without an inciting event (e.g., aortic surgery). Onset is more protracted and radiologic distinction from competing diagnoses is more difficult than with cerebral infarction [1]. Thus, patients with spontaneous upper cervical cord infarction (SUCI) are often misdiagnosed as having transverse myelitis. Differential diagnosis of SCI should exclude diseases such as acute inflammatory myelitis, multiple sclerosis, intra or extramedullary tumor, infectious conditions, and hematoma, which are characterized by features of spinal cord disorders; brain lesions and peripheral nerve lesions should be excluded as well [2,3].

Aortic disease is the most common cause, most probably due to the vulnerability of the thoracolumbar spinal cord to hypoperfusion [4,5]. Other etiologies include atherosclerosis, adjacent spinal degenerative disease, fibrocartilaginous embolism (FCE), vertebral dissection, systemic hypotension, cardiac embolism, coagulopathies, vasculitis, surfing-related myelopathy, and decompression sickness. Idiopathic cases are seen at a frequency of 20–40% [6,7]. A recent clinical study examining cervical cord infarctions in patients with and without vessel dissection showed that lesions were frequently located in the upper cervical cord among patients with dissection. In contrast, patients without dissection presented with an older mean age of onset and the lesions tended to be located in the lower cervical regions [8–10].

Spinal cord infarction may present diverse etiologies, with the cause remaining undetermined in many patients. Furthermore, the treatment of spinal cord infarction continues to be controversial, and no clinical practice guidelines have been issued on the topic. Factors associated with poor prognosis should be identified in order to prevent futile treatment. Long-term functional prognosis is poor and depends on baseline characteristics and clinical presentation [11–13].

Due to the rarity of spinal cord infarction, variables like risk factors, etiologies, and outcomes associated with spontaneous upper cervical cord infarction (SUCI) have seldom been discussed. In this study, we performed the description of these variables among the study cohort.

2. Methods

We report the risk factors, etiology, clinical presentation, and functional outcomes of thirteen consecutive SUCI patients admitted to the Department of Neurology of the University Hospital Center "Mohammed V Military Teaching Hospital" of Rabat (Morocco), between 2005 and 2020. The diagnosis was determined by the treating neurologist for all patients and consensus agreement by the co-authors of the study. Patients were assessed at nadir (time of maximal severity of signs), Modified Rankin Scale (mRS) (Table 1B), and last follow-up according to the American Spinal Injury Association Impairment (ASIA) scale (Table 1A). The ASIA scale is a widely used scoring system of motor and sensory function following spinal injury (including SCI) where a score of A indicates a complete loss of motor and sensory function and E indicates normal function. The Modified Rankin Scale (mRS) assesses disability in patients who have suffered a stroke and is compared over time to check for recovery and degree of continued disability. A score of 0 is no disability, 5 is a disability requiring constant care for all needs; 6 is death. Medline (1980–2022), EMBASE (1980–2022), and PubMed were used to identify 8 studies of spontaneous upper cervical cord infarction (SUCI) reporting prognosis and outcome and formed the basis of our review. Jamovi 2.3.18 was used for statistical analysis.

Grade	Description
A	**Complete:** No sensory or motor function is present at the most caudal sacral segments (S4-S5) as measured by perianal light touch or pinprick sensation
B	**Incomplete sensory:** Sensory but not motor function is preserved below the neurological level including sensory sacral segment S4-S5 sparing, on either side of the body
C	**Incomplete motor:** Motor function is preserved below the neurological level, and more than half of key muscles below the neurological level have a muscle grade of less than 3 strength
D	**Incomplete motor:** motor function is preserved below the neurological level, and at least half of key muscles below the neurological level have a muscle grade of 3 or more strength
E	**Normal:** Normal motor and sensory function

Table 1A: American Spinal Injury Association Impairment Scale

Grade	Description
0	No symptoms
1	No significant disability despite symptoms; able to carry out all usual duties and activities
2	Slight disability; unable to carry out all previous activities, but able to look after own affairs without assistance
3	Moderate disability; requiring some help, but able to walk without assistance
4	Moderately severe disability; unable to walk and attend to bodily needs without assistance
5	Severe disability; bedridden, incontinent, and requiring constant nursing care and attention
6	Dead

Table 1B: Modified Rankin Scale for Neurologic Disability (mRS)

3. Results

3.1. A Cohort of 13 Patients

We identified thirteen patients, 8 men (61.5%) and 5 women (38.5%) who were treated at our institution during a 15-year period (Table 2). Their mean age was 57.9±8.28 years. As risk factors, 11 patients 84.6% were followed up for arterial hypertension, 6(46.2%) for diabetes, 9(69.2%) were active smokers at the time of stroke, 7(53.8%) with ischemic heart disease, and 6(46.2%) with hyperlipidemia. Abdominal aortic aneurysm repair n=5(38.5%) and aortic dissection secondary to giant cell arteritis n=5(38.5%) were the possible etiology found. At the admission, three patients 23.1% presented with acute onset of mild cervical pain and tetraplegia. Ten patients 76.9% presented with paraplegia; among them, 6(46.2%) had severe cervical pain radiating to the lower limbs and 4(30.8%) had sudden onset of left upper limb pain. The latter, presented with sensory impairment and bowel dysfunction 4(30.8%) (Table 2). The clinical neurological level, defined by the most caudal spinal cord segment with normal sensory and motor function on both sides, was thoracic in nine patients and cervical in four patients. Six patients had no underlying vascular risk factors raising suspicion of fibrocartilaginous embolism. Overall reflexes were abolished while plantar responses were upgoing in eight, equivocal in three, and downgoing in two patients. The time to nadir was undetermined in all patients. Nadir's deficits were ASIA grade A in 4(30.8%) patients, grade B in 2(15.4%), C in 4(30.8%) and D in 3(23.1%) (Table 2). The Modified Rankin Scale at admission was mRS3 in 2 patients 15.4%, mRS4 in 7(53.8%), and mRS5 in 4(30.8%). All patients were unable to mobilize and were wheelchair dependent. Spinal MRI showed abnormal signal change compatible with ischemia (Figure 1). The initial MRI was abnormal in 9 patients, while the repeat MRI became abnormal in the other 4 patients on day 3 of the onset of the symptoms. The level of the lesions was C1 in 6 patients 46.2%, C2 in 3(23.1%), and C3 in 4(30.8%). All thirteen patients in this cohort benefited from symptomatic treatment and physiotherapy. The outcomes were assessed every six months. At different stages of follow-up, three patients, 23.1% had died. In the other 10 patients, the residual deficits were ASIA grade B in (1/10) patients, D in (3/10), and E in (6/10). The recovery and degree of continued disability over time were mRS1 in (6/10) patients, and mRS2 in (4/10).

Patient	Sex	Age (years)	Risk factors	Etiology	ASIA at nadir	FU/ASIA	mRS at nadir	FU/mRS	FU Duration
1	F	62	DT2, Smoking	Abdominal aortic aneurysm repair	C	D	4	2	24 months
2	M	60	Smoking	Aortic dissection secondary to giant cell arteritis	C	MD	4	MD	10 months
3	F	58	AHT, IHD, Smoking, Hyperlipidemia	Abdominal aortic aneurysm repair	B	D	4	1	24 months
4	M	69	AHT, IHD, Hyperlipidemia	Aortic dissection secondary to giant cell arteritis	A	MD	5	MD	6 months
5	F	61	DT2, AHT	Undetermined	A	MD	5	MD	6 months
6	M	51	DT2, AHT, Smoking, Hyperlipidemia	Aortic dissection secondary to giant cell arteritis	D	E	3	1	24 months
7	M	54	AHT, IHD	Abdominal aortic aneurysm repair	D	E	4	1	24 months
8	M	47	DT2, AHT, IHD, Smoking, Hyperlipidemia	Undetermined	C	E	3	2	24 months
9	F	69	AHT, IHD, Smoking, Hyperlipidemia	Aortic dissection secondary to giant cell arteritis	A	D	5	1	24 months
10	F	63	DT2, AHT, Smoking	Aortic dissection secondary to giant cell arteritis	D	E	4	2	12 months
11	M	41	DT2, AHT, Hyperlipidemia	Undetermined	A	E	5	2	24 months
12	M	64	AHT, IHD, Smoking	Abdominal aortic aneurysm repair	B	B	4	1	24 months
13	M	54	AHT, IHD, Smoking	Abdominal aortic aneurysm repair	C	E	4	1	24 months

DT2 = Type 2 diabetes, AHT = Arterial Hypertension, IHD = Ischemic Heart Disease, FU = Follow-up, nadir = time of maximal severity of signs, MD = Missing Data

Table 2: Cohort of 13 Patients Admitted to Our Institution Between 2005 And 2020 With Spontaneous Upper Cervical Cord Infarction

4. Literature Review of Prognostic Factors of SUCI

A summary of patient demographics, etiologies of SUCI, risk factors, initial and follow-up motor function, and mortality rates from 7 reported papers in the literature are outlined in Table 3.

5. Discussion

5.1. Key Findings

The three most striking findings from this study are: (i) As risk factors, arterial hypertension counted for 11 patients 84.6%, diabetes for 6(46.2%), smoking status for 9(69.2%), ischemic heart disease for 7(53.8%) and hyperlipidemia for 6 patients 46.2%. (ii) Abdominal aortic aneurysm repair n=5(38.5%) and aortic dissection secondary to giant cell arteritis n=5(38.5%) were the possible etiology found. (iii) At different stages of follow-up, three patients, 23.1% had died. In the other 10 patients, the residual deficits were ASIA grade B in (1/10) patients, D in (3/10), and E in (6/10). The recovery and degree of continued disability over time were mRS1 in (6/10) patients, and mRS2 in (4/10).

6. Implications

The risk factors of SUCI are as numerous as those already described in the literature for another topographical spinal infarction. Most patients presented vascular risk factors 75.6%, with the most frequent being arterial hypertension 53.6%. Smoking, hyperlipidemia, ischemic heart disease, and diabetes mellitus are described in various proportions [1]. Co-occurring vascular disorders included coronary artery disease, peripheral vascular disease, atrial fibrillation, and previous cerebral ischemia [11,13–15]. These reported findings confirmed those in our current study. But, aortic dissection, atherosclerosis demonstrated in vascular studies, fibrocartilaginous embolism, surgery, and hypotension were also cited as risk factors by another author [16]. Spinal cord infarction was attributed to atrial fibrillation and a state of hypercoagulability due to resistance to activated protein C and a prothrombin gene mutation. Nevertheless, the two most common etiologies of the USCI i.e. abdominal aortic aneurysm repair n=5(38.5%) and aortic dissection secondary to giant cell arteritis n=5(38.5%) we reported in our study are also the most common found in our literature review (Table 3).

Four of the patients in our series were Nadir ASIA grade A. Three of them responded well to the physiotherapy and became ASIA grade D after 12 months of follow-up. Moreover, 6 patients improved their neurological status from ASIA grade B/C to ASIA grade E with proprioceptive involvement. At 24 months of follow-up, 60% (6/10) of the patients had regained full walking ability. In contrast, our literature review of Table 3 highlighted only two patients being reported to be improved from ASIA grade B/D to ASIA grade E. In some other studies, clinical improvement was less frequent in ASIA grade A (14%) than in ASIA grades B (50%), C (40%), and D (59%). At follow-up, 41% of the patients had regained full walking ability, 30% were able to walk with aids, and 20% were wheelchair-bound. The best clinical outcome was found in patients with ASIA grade D: 68% were able to walk without aids, 23% walked with aids, and only 4.5% used wheelchairs. ASIA grade A patients had the worst clinical outcome with regard to walking ability: 2 of 7 were able to walk with aids, and 4 of 7 were wheelchair-bound [4,17,18].

The majority of the patients in our series, (n=11, 84.6%) scored 4/5 on the Modified Rankin Scale at admission. After 12 months of follow-up, 60% (6/10) recovered from disabilities and scored 1 of the mRS3, whereas 40% (4/10) improved to the mRS2. Our literature review of Table 3 has the same findings, with 43% (3/7) of the patients recovering from disabilities and scored from mRS 5/4 to 1. Finally, the mortality rate in our series was 23.1% while it was higher in some reported cases. Forty patients (53%) had died at the last follow-up, 8 of which were temporally related to complications from the procedure and/or SCI within a median of 1 month (range 0–4 months) [19,24].

Study Year(n)	Age (Years)	M:F	Risk factors	Etiology	ASIA at nadir	FU/ASIA	mRS at nadir	FU/mRS	FU Duration
Ogawa et al. [21], 2018(1)	86	0:1	AHT, DT2, Dyslipidemia	Undetermined	D	E	4	2	6 months
Lu et al. [10], 2019(1)	54	1:0	AHT, DT2	stenosis VA	A	D	5	1	03 months
Elzamly et al. [22], 2018	70	1:0	AHT, Dyslipidemia, Strokes	stenosis VA	B	E	2	1	03 months
Wada et al. [9], 2020(2)	84 74	1:1	AHT, DT2 AHT, prostate cancer, cholecystectomy	atherosclerosis occlusion RVA	D E	D E	1 1	1 0	4 months MD
Lei et al. [23], 2020(1)	44	1:0	-	stenosis BA+RVA	A	D	3	1	12 months

Zheng-Zhang et al. [16], 2022(1)	53	1:0	AHT, smoking, alcoholic	atherosclerosis FTP variation	C	C	5	1	12 months
Kim et al. [24], 2011(142)	MD	MD	DT2, AHT, Smoking, Hyperlipidemia, Previous stroke history, PCSE	Large-artery atherosclerosis, cardioembolisms, VA dissection, antiphospholipid antibody syndrome,	MD	MD	MD	MD	MD

n = Number of patients included, DT2 = Type 2 diabetes, AHT = Arterial Hypertension, IHD = Ischemic Heart Disease, FU = Follow-up, nadir = time of maximal severity of signs, MD = Missing Data, PCSE: Potential cardiac sources of embolism, FTP : fetal type posterior cerebral artery, VA = Vertebral Artery, BA = Basilar Artery, RVA = Right Vertebral Artery.

Table 3: Summary of Spontaneous Upper Cervical Cord Infarction Cases in The Literature

7. Conclusion

Spontaneous upper cervical cord infarction is a devastating neurological condition with challenging diagnosis and management, and with high mortality rate. The overall better outcome observed in this study can be related to the fact that more than half of the patients in our cohort had nadir ASIA C/D with mRS 3-4 at admission.

8. Limitations

There were several limitations of the study. This is a retrospective study that included patient's outcomes based on their clinical evolution with various follow-up lengths. There was no magnetic resonance imaging consideration during the follow-up. The small number of cases prevented the obtaining of certain generalizations. However, it should be mentioned that spontaneous upper cervical cord infarction is a quite rare pathology and that is why even the studies on small groups of patients are significant.

Declarations

Ethics Approval and Consent to Participate: N/A

Consent for Publication: N/A

Availability of Data and Material: All data generated or analysed during this study are included in this published article.

Competing Interests: The authors declare that they have no competing interests.

Funding: No funding was received for this research.

Acknowledgements: N/A

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