A case of „Spectacular Shrinking Deficit“ – case report and short review of elusive clinical phenomena

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Abstract:

Spectacular Shrinking Deficit (SSD) is a term attributed by J.P. Mohr to a rare cerebrovascular event defined by a rapid and dramatic improvement of major hemispheric stroke syndrome. It is presumed to be caused by the migration of initially embolic occlusion of an internal carotid artery or middle cerebral artery to its distal branches. It is only reported in several case reports, and case series with differing criteria of what defies an SSD meaning that its presumed prevalence rate of 7-14% of major hemispheric stroke syndromes could be an overestimation. It is usually associated with the cardioembolic cause of stroke, and it has a higher prevalence rate in younger patients, males, and nondiabetics. Our case is a 58 – year old male who presented to our ER with a major hemispheric stroke syndrome (deviation of head and eyes on the right side, central facioparesis on the left, left hemiplegia, and left hemineglect, NIHSS 16) 30 minutes after symptom onset. He was aggressive, insisting nothing was wrong with him. His initial brain CT showed acute ischemic changes in the right temporooccipital region with an inadequate filling of distal branches of the right MCA shown on CT brain angiography. His symptoms spectacularly improved in our ICU (NIHSS 0) 51 minutes from symptom onset negating the need for thrombolysis. Except for one positive Beta – 2 GPI test his detailed laboratory tests, 24h Holter ECG, TTE, carotid, and vertebral artery ultrasound were noneventful. He had no cognitive or neurological deficit. He denied the possibility of performing TEE and prolonged cardiac monitoring. Control brain MRI 4 days and 4 months later confirmed moderate ischemic changes of the right insular, temporal, and occipital cortex. Studies report that spontaneous recanalization usually happens in 17% of patients but does not correlate with TIA. This is the first report of an SSD with moderate ischemic stroke and no leftover neurological deficit. Other case series report moderate ischemic stroke SSD with a small residual neurological deficit (NIHSS 2-4). Perhaps mood changes can be associated with selective neuronal loss found in animals and patients suffering transient occlusion of the brain artery. The cardioembolic cause can in certainty be excluded with TEE and prolonged cardiac monitoring.

Keywords: Stroke, Spectacular Shrinking Deficit, TIA

Sažetak:

Slučaj „spektakularnog smanjujućeg deficita“ – prikaz slučaja i kratki pregled nedostižnih kliničkih pojava.

Spektakularno smanjujući deficit (SSD) je izraz koji J.P. Mohr pripisuje rijetkom cerebrovaskularnom događaju definiranom brzim i dramatičnim poboljšanjem velikog hemisfernog moždanog udara. Pretpostavlja se da je uzrokovan migracijom početne embolijske okluzije unutarnje karotidne arterije ili srednje moždane arterije u njezine distalne grane. Prijavljen je u samo nekoliko izvještaja o slučajevima

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i serijama slučajeva s različitim kriterijima onoga što definira SSD, što znači da bi njegova pretpostavljena stopa prevalencije od 7-14% velikih hemisferalnih moždanih udara mogla biti precijenjena. Obično je povezan s kardioembolijskim uzrokom moždanog udara, a ima veću stopu prevalencije u mlađih pacijenata, muškaraca i nedijabetičara. Naš slučaj je 58-godišnji muškarac koji se javio u hitnu neurološku ambulantu sa sindromom velikog hemisfernog moždanog udara (devijacija glave i očiju prema desnoj strani, centralna facioparеza s lijeve strane, lijeva hemiplegija te osjetno zanemarivanje lijeve polovice tijela, NIHSS 16) 30 minuta nakon pojava simptoma. Pacijent je bio agresivan, inzistirajući da s je s njim sve u redu. Njegov početni CT mozga pokazao je akutne ishemijske promjene u desnoj temporookcipitalnoj regiji s neadekvatnim punjenjem distalnih grana desne MCA prikazane na CT angiografiji mozga. Njegovi simptomi su se spektakularno poboljšali u našem JIL-u (NIHSS 0) 51 minutu od pojave simptoma negirajući potrebu za trombolizom. Osim jednog pozitivnog Beta – 2 GPI testa, njegovi detaljni laboratorijski testovi, 24h Holter EKG, TTE, ultrazvuk karotidnih i vertebralnih arterija su bili uredni. Pacijent nije imao kognitivni niti neurološki deficit. Negirao je mogućnost izvođenja TEE-a i produženog praćenja srčanog ritma. Kontrolni MR mozga 4 dana i 4 mjeseca kasnije potvrdio je umjerene ishemijske promjene desnog inzularnog, temporalnog i okcipitalnog korteksa. Istraživanja pokazuju da se spontana rekanalizacija obično događa u 17% pacijenata, ali ne rezultira nužno s TIA-om. Ovo je prvi prikaz slučaja o SSD-u s umjerenim ishemijskim moždanim udarom i bez zaostalog neurološkog deficita. Drugi prikazi slučajeva pokazuju SSD s umjerenim ishemijski moždani udarom i s blagim zaostalim neurološkim deficitom (NIHSS 2-4). Možda promjene raspoloženja mogu biti povezane sa selektivnim gubitkom neurona dokazanim u životinja i pacijenata koji su bili izloženi prolaznoj okluziji velikih moždanih arterija. Kardioembolijski uzrok može se sa sigurnošću isključiti samo kompletnom obradom koja uključuje TEE i produljenim praćenjem srčanog ritma.

Ključne riječi: Moždani udar, Spektakularno smanjujući deficit, TIA

1. Introduction

Spectacular Shrinking Deficit (SSD) is a relatively rare phenomenon in the field of cerebrovascular diseases. The term was first coined by J.P. Mohr in the mid-1980s with a typical definition of SSD being one of major hemispheric stroke syndrome followed by a rapid and dramatic spontaneous improvement of symptoms within a couple of hours from stroke onset with discrete or no clinical residual deficit (1). Major hemispheric stroke syndrome encompasses major neurological deficits such as aphasia, severe hemiparesis/hemiplegia with or without sensory symptoms, and cortical signs (gaze deviation, aphasia, neglect) with or without disturbed consciousness.

It is presumed that SSD comes about because of a migration of initially embolic occlusion of an internal carotid artery or middle cerebral artery to its distal branches (2). Clinical improvement is then possible thanks to the acute reperfusion of ischemic brain tissue in contrast to delayed reperfusion which gives rise to potential intracerebral hematoma (3). Reports about SSD in humans are scarce and are limited to several case reports, case series, and retrospective cross-sectional studies. Time constraint from stroke onset to the improvement of deficit necessary to fit in a definition of SSD is different from study to study (one study puts a time constraint necessary for diagnosing SSD to 6h from stroke onset, one requires it to be in an initial couple of hours, while another one acknowledges it to happen in a matter of days). Besides time a recovery level of neurological deficit is a source of debate with some studies defining SSD as a reduction in NIHSS score by 8 points, and others claiming a necessity of control NIHSS being ≤ 4 points. Therefore the true prevalence of SSD can only be presumed with estimates that it happens in about 7-14% of major hemispheric strokes. A study by Minematsu et al (2) had 14 SSD patients in 5 years, V.H. Lee et al. (4) had 9 (7%) of patients with SSD in 2 and a half years, Fujioka et al (5) reported 4 such patients in a 2 and a half year period, and 14 (13,5%) patients met the study criteria of SSD in a study by Kraemer et al. (6) (in that study all patients received IV thrombolysis which collides with „spontaneous“ part of SSD definition). It is more prevalent in younger patients (<60 years.), male sex, without previous history of diabetes mellitus, and with a potential cardiac source of emboli. Some studies think of SSD as a definite sign of cardiogenic brain embolism, while a study by V.H. Lee et al. (4) had only 2 out of 8 patients with certain cardioembolic stroke (4 had large artery stenosis/occlusion, and 2 had unknown stroke cause). SSD patients have a favorable long-term clinical outcome (4). SSD is an interesting phenomenon in human stroke pathophysiology with its rare clinical picture in a state of limbo between clinical TIA, and certain ischemic strokes with a clinical deficit.

We present a case report of a spectacular shrinking deficit which happened 51 minutes after the stroke onset and left a moderate cortical ischemic stroke visible on brain MRI without clinical deficit.

2. Case report

A 58- year-old right-handed man was reported to our ED with severe hemispheric stroke. By anamnestic accounts from his wife, the symptoms of our patient started around half an hour before arriving in our ED. The patient first experienced a sudden onset of severe headache after which his wife saw that he had profound weakness in his left extremities, and he became confused and agitated. He supposedly moved his left extremities with extreme awkwardness, unaware of his deficit, and attempted to drive a vehicle that alarmed his partner and prompted her to call an ambulance. The patient was aggressive insisting that there was nothing wrong with him. On arrival in our ED emergency neurologist was paged and in initial examination witnessed a deviation of head and eyes on the right side, central facioparesis on the left, left hemiplegia, and left hemineglect with an overall NIHSS score of 16. Initial CT (45 minutes from symptom onset) of the brain showed acute ischemic changes in the right temporoocipital region while CT angiography of the brain did not show adequate filling of distal branches of right MCA corresponding to ischemic areas (Figures 1. and 2.). Emergency laboratory tests and chest X-ray were of no significance. The patient fulfilled all criteria for thrombolysis treatment. While transporting the patient to our neurologic ICU and preparing him for an administration of IV thrombolysis the patient had a spectacular improvement of neurological deficits. All of his deficit had resolved with a control NIHSS score of zero. His neurological deficit was present during the 30 minutes before coming to ED, 15 minutes during the door to CT further 6 minutes from CT to ICU with a total of 51 minutes of large hemispheric syndrome deficit. While hospitalized the patient was agitated being convinced he did not have a stroke, and it was all a misunderstanding on behalf of his wife and all of the medical staff. Anamnestically from his medical history we found that patient had no chronic illnesses but had one episode of unspecified arterial thrombosis in his left leg 25 years before the incident. He said his father supposedly had thrombophilia but was unaware of serious neurological illnesses in the family. We excluded potential cardiac sources of emboli in our patient with nonsignificant findings in transthoracic echocardiography, while the patient declined to perform transesophageal echocardiography. 24- hour Holter ECG did not report aberrations of heart rhythm, with patient refusing to do a prolonged cardiac monitoring after hospitalization. We excluded potential carotid or vertebral artery stenosis with Doppler ultrasound of the aforementioned arteries and with the before-mentioned CT angiography. Detailed laboratory tests were done searching for potential risk factors for ischemic stroke including standard biochemical tests (glucose, lipids, hormones), inflammatory, autoimmune, hematological, and neoplastic tests, all of which came normal. Genetic testing was noneventful except for Beta 2 – GPI which was positive in one testing sample. Aditional brain MRI (5 days from symptom onset) showed supratentorial right temporal, parietal, caudal insular, and lateral occipital voluminous cortex which was hypointense on T1, hyperintense on T2, and FLAIR sequences with DWI, and ADC sequences showing restriction of diffusion (Figure 3. and 4.). The imaging correlated to ischemic changes in an early subacute stage. Head of right n. caudatus had also had an increased signal on DWI sequence, discreetly hypointense signal on ADC sequence corroborating acute ischemia. Right putamen had also shown discreetly higher signal on T2, and FLAIR sequences, and discreetly hypointense signal on T1 sequence with a slightly higher signal on DWI sequence attributed to acute ischemic changes. The patient was categorized as an unknown cause of ischemic stroke (possible antiphospholipid syndrome, at least 2 tests 12 weeks apart were needed to confirm the diagnosis), and was released from hospitalization with no neurological deficit (mRS 0, Barthel Index 100), and with dual anti-aggregation therapy prescribed. He was to conduct a control MRI of the brain four months after the hospitalization, control Beta 2 – GPI test 12 weeks after the hospitalization with referral to a rheumatologist, and afterward report to a cerebrovascular infirmary. The patient did a control MRI of the brain 4 months after the hospitalization which showed chronic gliotic, and malacial postischemic changes on the right insular, temporal, and occipital cortex with extensive zones of postischemic laminar cortical necrosis in the mentioned area. N. caudatus dex. had normal signal intensity and volume without the signs of chronic postischemic changes (Figure 5.-6.). The patient was without any neurological deficit. At the time of writing this report, the patient did not do a control Beta 2 GPI test.

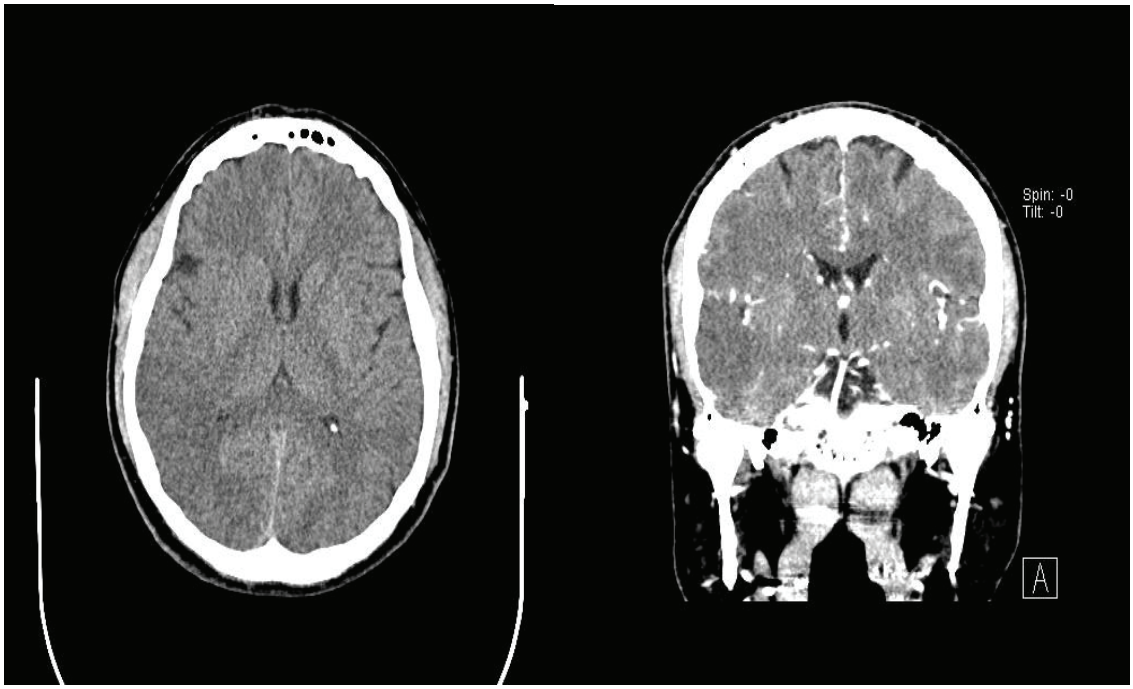


Figure 1. Initial emergancy brain CT (45 minutes after the onset of symptoms).
Figure 2. Emergancy CT angiography (45 minutes after the onset of symptoms).

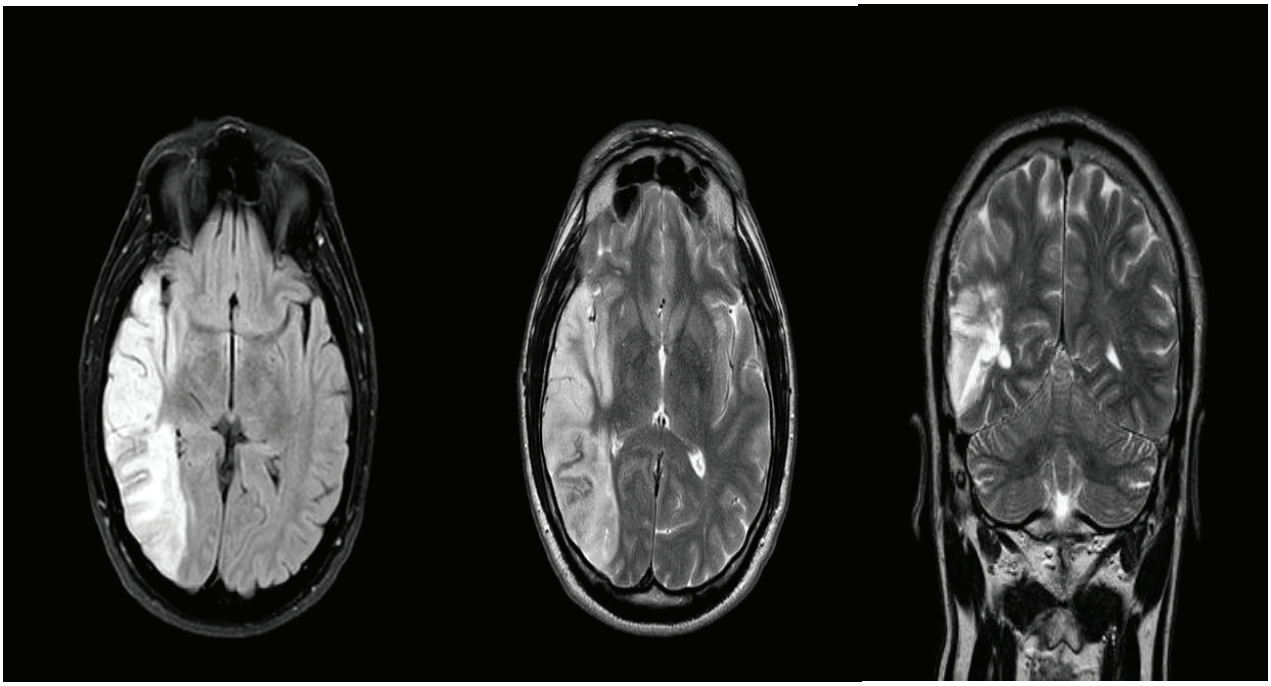
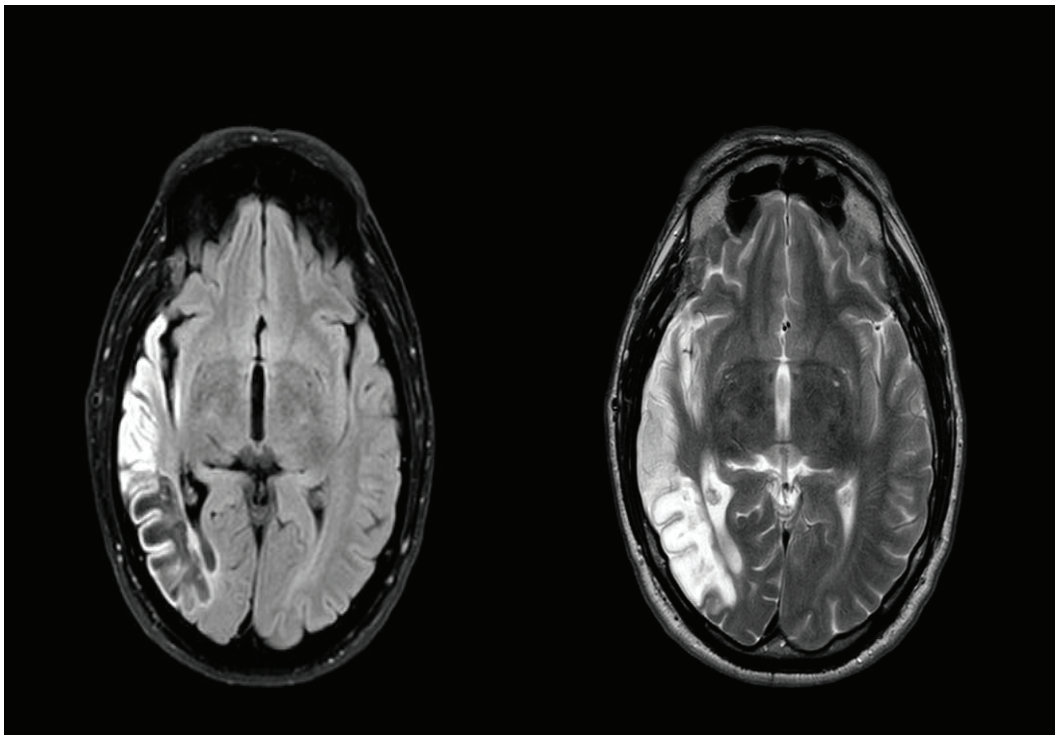


Figure 3. (T1W), 4. (T2W), and 5. (T2W) Show brain MRI 4 days after the onset of symptoms.



Figures 6. (T1W) and 7. (T2W) show MRI 4 months after the onset of symptoms with gliotic and malacial changes in right posterior insular gyrus, inferior, medial, and superior temporal gyrus, and lateral occipital cortex. Right nucleus caudatus showed no postischemic changes.

3. Discussion

Our patient satisfied even the most rigorous definitions of SSD with a spontaneous drop in NIHSS score from 16 to 0 in less than an hour. The evolution of his clinical picture would be following a theory of spontaneous breaking of occluding embolus which initially occluded right MCA and then spontaneously recanalized with fragments of the emboly clogging distal branches of right MCA. A review by H.K. Moussa finds that spontaneous recanalization in the first 6 to 8 hours happens in approximately 17% of patients but spontaneous recanalization did not necessarily correlate with clinical TIA (7). What differentiates this from „plain old" TIA is the fact that the ischemic changes of brain parenchyma were without the doubt present in all neuroimaging studies (let alone the fact that the ischemic changes were present on MSCT 40 minutes from stroke onset, and preserved on control MRI studies 4 months later). Ischemic changes being confirmed by follow-up MRI 4 months after stroke were present in the right posterior insular gyrus, middle, inferior, and superior temporal gyrus, and lateral occipital cortex which constitutes moderate ischemic stroke. According to the affected brain parenchyma our patient should have by all logic exhibited at least some of the symptoms such as inability to judge spatial relationships, agnosia for sounds, emotional and behavioral changes, delirium, and disturbances of time perception, left homonymous hemianopia, and loss of visual orientation (8,9,10) That being said our patient was oriented in time and space, emotionally stable, used his smartphone to listen to music without difficulties, and walked on our Department with excellent spatial orientation.

Being satisfied with our patient's complete reversal of deficit, and witnessing his overall normal cognitive functioning on subjective examiners terms we did not test our patient with some of the more profound neuropsychological tests (8). Our patient quite possibly had an arterial thrombosis caused by an antiphospholipid syndrome, but being that two independent tests of Beta 2 GPI 12 weeks apart are needed to confirm the diagnosis we couldn't make a final diagnosis (11). 24 h ECG and transthoracic echocardiography were uneventful meaning we couldn't make a usual association between SSD and cardioembolic stroke. Previous studies of embolic stroke of unknown source have found that prolonged heart monitoring reveals paroxysmal atrial fibrillation in 16,1% of patients compared to 3,2% in control group when other causes are excluded (12). Of 14 reported cases of SSD in a

study by Minematsu et al. only 2 patients had moderate ischemic stroke visible on brain CT, all the others had punctiform ischemia in basal ganglia or cortex. Of patients with a moderate ischemic stroke, one of them had mRS 0 upon release from hospital (recovery after 4 hrs), while the other one had mRS 3 (recovery after 8 hrs). Only 3 of 14 patients were women, and only one patient did not have a definitive cardioembolic cause of stroke (2). V.H. Lee et al. reported 8 eligible SSD patients (7 men, 1 woman), with a mean age of 62 years, a mean time to dramatic recovery of 3.4 hrs. 5 patients met the criteria for TIA while the other 3 patients were diagnosed with minor ischemic stroke (NIHSS score ranging from 2-4). Of those 5 patients (control NIHSS score 0) reported only one had moderate subcortical ischemia on MRI DWI while the others had punctiform subcortical ischemia. All 5 had initially moderate perfusion cortical lesion on CTP which subsided with time. Fujioka et al. reported 4 SSD patients (recovery of symptoms began within 60 minutes) with no abnormal findings on initial CT, and only 1 patient having left frontal cortical and external capsular infarct on initial MRI, with others having normal MRI obtained on day 2-3 after stroke. Further serial MRI studies reported delayed lesions with hyperintensity on T1W and hypointensity on T2W MRI in the caudoputamen of all patients, and cerebral cortex in 2 patients. These lesions appeared clearly between 1 and 3 weeks after stroke, and gradually faded with atrophy of affected structures. Termed „delayed ischemic hyperintensity on T1W MRI“ (DIH) it is speculated that it perhaps represents a selective neuronal loss or biochemical changes due to incomplete ischemia (5). Previous studies have found that transient occlusion of brain arteries both in experimental animals and in human patients causes selective neuronal loss (SNL) with most of the glial cells preserved together with some of the neurons leading to tissue atrophy without cavitation. Recanalization causes resolution of obvious clinical symptoms while SNL is supposedly connected to subtle behavioral and mood impairments (13,14,15,16). F.A.A. Gondim et al. report shows the width of this phenomenon with a patient whose major hemispheric stroke both clinically and neuroradiologically is recovered in 10 days to minimal residual ischemia with minor symptoms (17). Y. Terasawa et al. report a patient with SSD (NIHSS 15 to 0 in 24 hrs) whose DWI lesion in corona radiata resolved on follow-up MRI (2.5 hrs from onset) with persistent MCA occlusion. His follow-up MRI 24 hrs after onset revealed recanalization of MCA and multiple DWI hyperintense lesions in the MCA area without clinical deficit presuming that immediate opening of the leptomeningeal anastomosis with still occluded MCA caused initial saving of tissue (18). Even though reversal of symptoms may seem like a guarantee of a good outcome a study by V. Rajajee et al. bears a warning. Their study has shown that 1 in 10 patients with rapid improvement of motor symptoms and without treatment deteriorate in the next 48 hours, and 1 in 5 patients is discharged with poor functional outcomes. They found that a persisting large vessel occlusion is a major predictor of early neurologic deterioration, but those patients without one can safely be managed conservatively (19).

In conclusion, a lot can still be learned about the pathophysiology of ischemic stroke. Being that major hemispheric stroke has usually dismal outcomes SSD still amazes clinicians with its major or complete reversal of all deficits. Ischemic stroke and TIA are usually clearly defined opposites of a spectrum in between which there are still less explored territories. To our knowledge, this is the first presentation of an SSD patient with clear moderate ischemic stroke and ischemic necrosis visible on the control MRI without neurological deficit. Our patient failed to grasp that he had a serious incident despite our explanation showing that these situations need to be presented to both clinicians and patients. Thorough testing needs to be done to exclude the cardioembolic cause of SSD.

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