Open access Journal of
Neurology & Neurosurgery
ISSN: 2476-0501

Juniper Publishers
key to the Researchers

Case Report
Volume 16 Issue 2 - October 2021
DOI: 10.19080/OAJNN.2021.16.555933

Open Access J Neurol Neurosurg

The Dual Catastrophe of Behçet's Disease: Visual Loss Followed by Acute Spinal Shock After Lumbar Drain Removal

Naim I Kajtazi[1]*, Mohammed Al Sheef[2], Ehtesham Khalid[3], Juman Al Ghamdi[4], Majed Al Hameed[1] and Mohammed Bafaquh[5]

[1]*Department of Neurology, National Neuroscience Institute, King Fahad Medical City, Riyadh, KSA*

[2]*Internal Medicine Department, Thrombosis Section, King Fahad Medical City, Riyadh, KSA*

[3]*Department of Neurology, Ideal Medicare Clinic, Multan, Pakistan*

[4]*Medical Imaging Administration, Intervention Neuroradiology, King Fahad Medical City, Riyadh, KSA*

[5]*Department of Neurosurgery, National Neuroscience Institute, King Fahad Medical City, Riyadh, KSA*

Submission: September 29, 2021; **Published:** October 18, 2021

***Corresponding author:** Naim I Kajtazi, National Neurosciences Institute, Department of Neurology King Fahad Medical City PO. Box 59046 11525 Riyadh, Kingdom of Saudi Arabia

Summary

We report a case of a 40-year-old-male with Behçet's disease and cerebral venous thrombosis, and other multiple comorbidities admitted with a four-day history of increasing headache and rapidly progressive visual loss bilaterally. The neurological examination was positive for bilateral papilledema of grade 3 with light perception on the left eye and counting fingers on the right eye. Brain imaging showed old findings of cerebral venous thrombosis without any intraparenchymal lesions to suggest a flare-up of Behçet's disease. The lumbar puncture followed by the lumbar drain insertion gave no benefit in headache or vision. However, he completely lost sight. The right optic nerve sheath fenestration did not result in vision improvement. The acute spinal shock complicated the lumbar drain removal due to epidural hematoma. An urgent lumbar laminectomy with hematoma evacuation undertook. Intra-operatively, the neurosurgeon noted suspicious abnormal vessels at conus medullaris with the possibility of an arteriovenous malformation. In few days following the spinal surgery, the patient vision started to improve. Further improvement was achieved after plasma exchange sessions followed by cyclophosphamide. In the recent follow-up in the clinic, he reported better vision, drove, and completed his Ph.D. studies. This patient's story is significant for a high disease burden and complicated hospital course by acute spinal shock due to spinal lumbar drain removal with a possible underlying spinal arteriovenous malformation.

Case presentation

A 40-year-old-male with Behçet's disease and Cerebral Venous Thrombosis (CVT), presented to the ER (emergency room) with a four-day history of worsening headache and rapidly progressive visual loss bilaterally. The headache was mild, generalized, and not associated with nausea or vomiting. He could see only to count fingers.

In a young man with a history of acute vision loss in the background of Behçet's disease and cerebral venous thrombosis, we were concerned about raised intracranial pressure (ICP) due to possible recurrence of thrombosis, intracerebral hemorrhage, vasculitis, acute decompensation of idiopathic intracranial hypertension (IIH) as a known complication of CVT, or local orbital causes.

At the age of 38 was diagnosed with Behçet's disease following a two-month history of severe headache, neck pain, recurrent oral and genital ulcers with recurrent superficial vein thrombophlebitis of lower extremities. In addition, brain MRI with MRV (magnetic resonance imaging with magnetic resonance venography) had shown cerebral venous thrombosis. He was given warfarin. Other past medical history was significant for hypertension, dyslipidemia, hyperhomocysteinemia, hyperuricemia, vitamin B12 deficiency. At age 39 was admitted to the cardiology floor with angina chest pain and was diagnosed with NSTEMI (non-ST segment elevated myocardial infarction). He underwent percutaneous coronary intervention with stenting for severe three-vessel disease with an aneurysm secondary to vasculitis. Thus, he was started on aspirin, clopidogrel, azathioprine, prednisolone, and cyclophosphamide treating coronary artery vasculitis.

We have a patient with a complex medical history, and all the factors have to be accounted for. Moreover, the progression is

rapid, and he has a very significant past medical history at risk of being confused with another differential diagnosis. Therefore, we realized the necessity of quick actions and decisions to preserve the patient's vision while looking for the exact cause.

The neurological examination was positive for bilateral papilledema of grade 3 (worsened from the last exam two months ago). Visual acuity was decreased; light perception on the left eye and counting fingers on the right eye. Brain CT was normal. Brain MRI with MRV documented a filling defect in the superior sagittal sinus and torcular herophili, suggesting chronic thrombosis with the thin flow within the superior sagittal sinus and good recanalization within the transverse sigmoid sinuses as well as internal jugular veins (Figures 1 & 2). No new lesions were suggestive of a flare-up of Behçet's disease. Prominent subarachnoid spaces along the optic nerves were worrisome for increased ICP. ESR (erythrocyte sedimentation rate) and CRP (C-reactive protein) were normal, and INR was therapeutic. An ophthalmologist's examination documented severe bilateral papilledema with no uveitis.

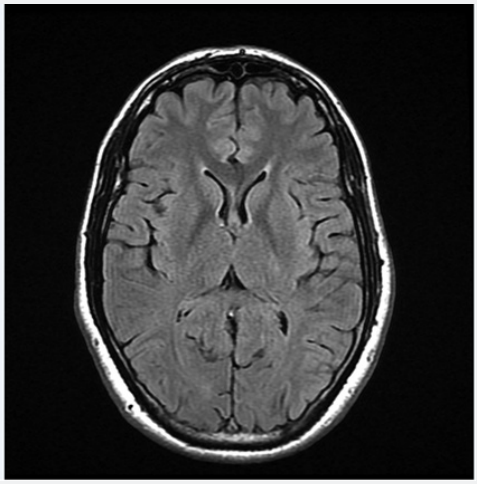

Figure 1: Brain MRI axial FLAIR (fluid attenuated inversion recovery) shows no abnormal signal intensity within the brain parenchyma.

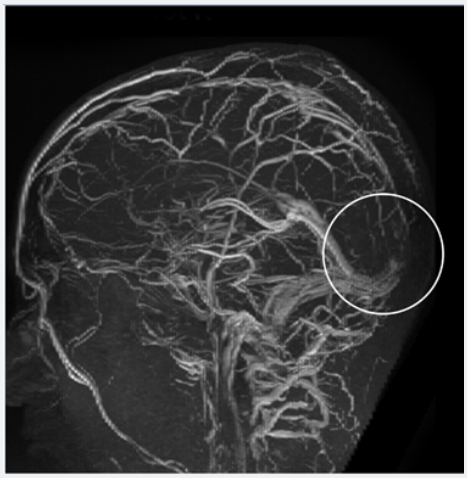

Figure 2: Brain MRV (magnetic resonance venography) shows a filling defect in the superior sagittal sinus and torcular herophili suggestive of chronic thrombosis with thin flow within the superior sagittal sinus and presence of collaterals".

We concluded this is not secondary to an acute thrombotic event because the brain MRV showed no new thrombosis and was unchanged from the previous imaging a few months ago when he was asymptomatic and the patient was fully anticoagulated. In addition, his ESR and CRP were normal, which argued against active vasculitis, and the patient was on immunosuppressive agents already. We needed a diagnostic lumbar puncture (LP) to measure the opening pressure and a therapeutic for an acute decompensated raise in the ICP. However, the patient was anticoagulated, and we needed to reverse that.

Acetazolamide that he had stopped unintentionally was restarted, and LP was done after reversing the INR to normal with fresh frozen plasma. The opening pressure was 46 cmH20, and a total of 29 ml of clear fluid were collected.

How to cite this article: Naim I K, Mohammed Al S, Ehtesham K, Juman Al G, Majed Al H, and Mohammed B. The Dual Catastrophe of Behçet's Disease: Visual Loss Followed by Acute Spinal Shock After Lumbar Drain Removal. Open Access J Neurol Neurosurg 2021; 16(2): 555933
DOI: 10.19080/OAJNN.2021.16.555933

CSF analysis showed normal cell count, glucose, and protein. However, following the procedure, the patient's vision and headache did not improve.

We have evidence of increased ICP likely causing visual loss and headache. Removing CSF usually improves the symptoms, but it was not the case in this patient. Could this be vasculitis, and does he require augmented immunosuppression, or did we not remove enough CSF to reduce the increased ICP?

We consulted the neurosurgery team for the raised ICP and possible VP (ventriculoperitoneal) shunt insertion. However, they suggested the patient initially have lumbar drain insertion to prove an improvement in headaches, papilledema, and visual loss. Unfortunately, in the same evening, the patient lost his vision completely in both eyes. His pupils were dilated and became unreactive to light. The patient had an urgent lumbar drain insertion after temporarily holding heparin infusion. No immediate complications occurred, and the rate of CSF drainage was set at 5 ml/hour and increased to 20 ml/hour. Despite this, the patient had no improvement in his vision. After consultation with the thrombosis and rheumatology teams, the anticoagulation was restarted with unfractionated heparin. We continued him on aspirin and clopidogrel for the significant cardiac condition. Azathioprine and prednisolone were discontinued. He was started on pulse intravenous steroids treating possible vasculitis, but the patient vision did not improve.

The patient has a persisting headache and lost vision, which is not improving either by CSF drainage through lumbar drain or medications, making him unlikely to respond to a VP shunt insertion. He was also treated for possible vasculitis and was fully anticoagulated and on two antiplatelets to treat possible thrombotic events. We were left with the option of optic nerve sheath fenestration, which is a modality used to relieve intracranial pressure. In addition, the option of starting plasma exchange for possible vasculitis was suggested.

The patient underwent right optic nerve sheath fenestration four days after admission. However, there was no immediate improvement in his vision. The lumbar drain was removed five days later due to severe low back pain and no vision or headache improvement. However, six to eight hours after removing the lumbar drain, the patient developed more headaches and severe back pain with urinary retention and weakness in the lower limbs.

The patient, unfortunately, presents with acute spinal cord syndrome, likely compression of the spinal cord secondary to hematoma formation following the drain's removal. The patient was still on aspirin, clopidogrel, and heparin infusion when the drain was removed. The symptoms developed during the night when access to MRI was limited, so he went for an urgent spine CT instead.

An urgent spine CT scan was done, which did not show any evidence of collection. Twenty-four hours after developing his spine symptoms, the patient had a spine MRI revealing heterogeneous abnormal signal intensity within the thecal sac and in the epidural space extending from T12 down to L1; measuring approximately 9 mm in the maximum thickness resulting in displacement and compression of the cauda equina nerve roots and the conus medullaris which exhibited abnormal intramedullary T2 hyperintensity (Figures 3-5). The overall appearance was highly suggestive of intraspinal hematoma with different stages of hemorrhagic degradation products.

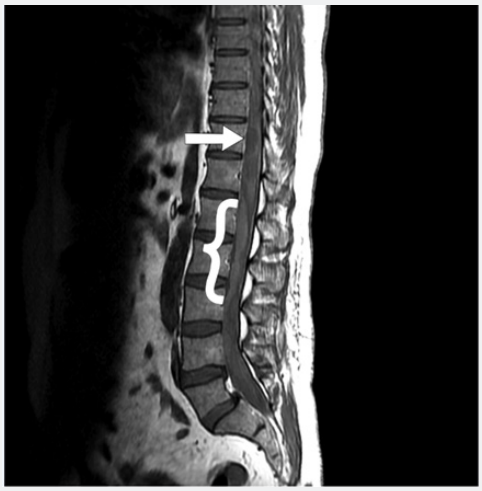

Figure 3: Lumbo-sacral spine MRI, sagittal T1 shows heterogeneous abnormal signal intensity with susceptibility artifact within the thecal sac and in the epidural/subdural space and the later one appears to be extending from T12 through L1 level and measuring approximately 9 mm in the maximum thickness.

How to cite this article: Naim I K, Mohammed Al S, Ehtesham K, Juman Al G, Majed Al H, and Mohammed B. The Dual Catastrophe of Behçet's Disease: Visual Loss Followed by Acute Spinal Shock After Lumbar Drain Removal. Open Access J Neurol Neurosurg 2021; 16(2): 555933
DOI: 10.19080/OAJNN.2021.16.555933

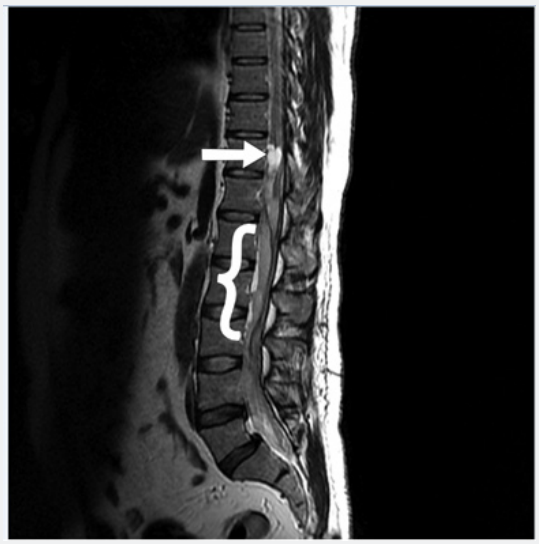

Figure 4: Lumbo-sacral spine MRI, sagittal T2 shows heterogeneous abnormal signal intensity with susceptibility artifact within the thecal sac and in the epidural/subdural space and the later one appears to be extending from T12 through L1 level and measuring approximately 9 mm in the maximum thickness resulting in displacement and compression of the cauda equina nerve roots and the conus medullaris which exhibit abnormal intramedullary T2 hyperintensity.

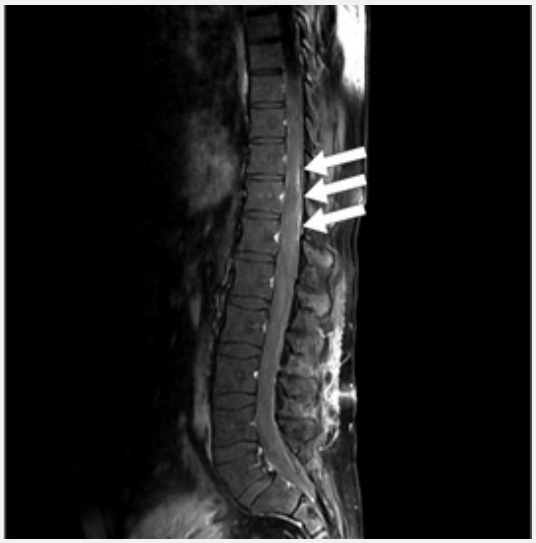

Figure 5: Lumbo-sacral spine MRI, sagittal T1 with contrast shows linear contrast signal enhancement seen along the surface of the conus medullaris but no perceptible signal contrast enhancement demonstrated within the hematoma area.

The patient developed iatrogenic complications related to lumbar drain removal while on aspirin, clopidogrel, and heparin infusion, although the procedure was not traumatic. Nevertheless, his initial symptoms on presentation did not improve and became completely blind and paralyzed. With this new emerging complication, the neurosurgical team was consulted for urgent hematoma evacuation. **The patient underwent an urgent lumbar laminectomy at T11, T12, and L1 with hematoma evacuation. Intra-operatively, the neurosurgeon noted suspicious abnormal vessels at conus medullaris with the possibility of an AV (arteriovenous) malformation, which may have contributed to the hematoma formation on the background of uninterrupted antiplatelet agents and anticoagulation. Therefore, the dura was kept open within the spinal space to prevent the hematoma's recollection, and the patient had wound drainage**. In few days following the spinal surgery, the patient vision started to improve. However, we remained unsure of the exact mechanism that improved the patient's vision, as he was fully anticoagulated on immunosuppressant agents and acetazolamide. Additionally, the dura was kept open within the spine, and he had optic nerve sheath fenestration. What remained as a question is whether he has CNS vasculitis and whether or not he needs a cerebral angiography to document that? This patient had coronary vasculitis in the past that caused acute coronary

How to cite this article: Naim I K, Mohammed Al S, Ehtesham K, Juman Al G, Majed Al H, and Mohammed B. The Dual Catastrophe of Behçet's Disease: Visual Loss Followed by Acute Spinal Shock After Lumbar Drain Removal. Open Access J Neurol Neurosurg 2021; 16(2): 555933
DOI: 10.19080/OAJNN.2021.16.555933

syndrome. Will finding CNS vasculitis change our management as the patient was planned to receive plasma exchange, but we delayed when he developed the acute spinal cord syndrome? We discussed the option of doing a cerebral angiography with the patient, but as he had an eventful and complicated hospital course, he did not want to undergo another procedure. So, the decision was not to do it, and he was planned for aggressive immunosuppression by the rheumatology team.

The patient received five plasma exchange sessions with improving vision and headache, and we transferred him to the Rehabilitation hospital where further improvement was achieved. Later he developed left testis epididymitis secondary to Behçet's disease, conservatively managed by the urology team. Upon discharge, he was independent in bed mobility, sitting to standing with good sitting balance and maintaining standing balance. With supervision, the patient could walk with a simple cane for more than 50 meters but required intermittent catheterization for urine retention. He had completed seven sessions of cyclophosphamide infusion of 1 gram. A follow-up visit in the clinic noted walking with a simple cane, driving, and having mild bilateral lower limb weakness with right hemiparetic gait and still required intermittent self-catheterization for urine retention. His INR had been stable on warfarin 2.5 mg, and the rheumatology team had started infliximab injection along with prednisolone 5 mg daily in addition to colchicine, aspirin, clopidogrel, acetazolamide and other medications. Later developed subdural hematoma related to combined anticoagulant and dual antiplatelet use, which resolved with discontinuation of clopidogrel and did not result in any neurological sequelae. In his recent follow-up in the clinic, he reported better vision, drove, and completed his Ph.D. studies.

The improvement of the patient's symptoms was a very relieving factor for the patient and his family, and the treating team. He especially had a high disease burden with significant past and current medical history presenting with headache and visual loss and complicated hospital course by acute spinal shock due to spinal lumbar drain removal with a possible underlying spinal arteriovenous malformation.

Discussion

Difficulties in clinical diagnosis with high number of clinical variants.

The clinical diagnosis of a disease can be difficult and goes through a multilevel of information gathering and investigations, which will require more time than usual in the case of the patient's complex medical history. These difficulties that the medical team can face are related to many factors, such as factors related to the disease, patients, intuitional factors, and that related the experience of the medical teams dealing with the case of interest. Medical illness can be very complex to understand and diagnose, particularly when its progression is rapid or has a long list of variants and variables that make it easy to be confused with another differential [1]. Even with the advancement of technology in the medical field, we still depend heavily on history, physical examination, and imaging in diagnosing a medical illness. Although this approach is essential, it is sometimes not enough to reach the appropriate diagnosis promptly. Two fundamental reasons can result in this: one is the molecular and genetic factors, which are still not well understood for many illnesses, and the other is the difficulties in finding information regarding exposure to environmental factors in many patients [2]. In one study and multiple surveys, the misdiagnosis was found to be as high as 35% [3]. However, in an autopsy study, Goldman found the rate of misdiagnosis to be 24%, with stable misdiagnosis over three decays. In the same study, the author found that misdiagnosis would not change the management of the patient 12% of the time [4]. In our case, the patient presented with complex medical history with multiple complex and challenging to diagnose disorders such as Behçet's disease, cardiac vasculitis associated with LAD aneurysm, CVT that became chronic, and epididymitis. He also presented with very rapid visual deteriorations that did not give a chance for the team to establish the diagnosis, gather information, and start a medical management attempt to save a vital function/organ of the patient (i.e., vision). Although initially did not show effectiveness, the management, however eventually and after the neurosurgeon had to deal with a complication of the lumbar drain by decompressing the spine has improved the patients' symptoms. Although this is not ideal, the team has felt the burden of saving patient vision and probably live in a very complex clinical illness with multifactor, including antiplatelet and anticoagulation.

Causes of visual loss in IIH secondary to vasculitis or and CVT, and in Behçet's disease.

Increased intracranial pressure and associated symptoms such as headache, papilledema, motor or sensory deficits, seizures, and conscious disturbance are well known in acute CVT [5]. However, visual loss is not commonly associated with this disease, except in the case of secondary IIH associated with it. Ophthalmological features such as diplopia, subnormal visual acuity, and visual field defect have been documented in the literature and related those manifestations to the increase in the ICP [6]. Zhao T and his colleagues report three cases of CVT and elevated ICP, without apparent symptoms of ICP other than visual impairment [7]. In this study, the first case visual acuity improved after six months with conservative management, the second case improved after being treated with stent placement to the superior sagittal sinus, and the third case was found to have stenosis of the transverse sinus and improved after stent placement. Important to notice that not all of their patients had a definitive diagnosis of CVT.

On the other hand, vision loss can be noted with Behçet's disease, and there is a 39% risk of visual loss, and severe visual

How to cite this article: Naim I K, Mohammed Al S, Ehtesham K, Juman Al G, Majed Al H, and Mohammed B. The Dual Catastrophe of Behçet's Disease: Visual Loss Followed by Acute Spinal Shock After Lumbar Drain Removal. Open Access J Neurol Neurosurg 2021; 16(2): 555933
DOI: 10.19080/OAJNN.2021.16.555933

loss is noted in 21% of cases, which is commonly due to uveitis and ischemic maculopathy [8]. However, there are various potential etiologies, including high ICP secondary to venous outflow obstruction or due to abnormal shunting [9]. High ICP could be due to compromise in CSF drainage at the level of arachnoid granulations or venous sinuses, and small changes may be missed on comparison of the scans as it focuses more on large sinuses and venous channels. In our case, later worsening of vision and complete vision loss could be due to abnormal shunting as noted in dural AV malformations with increased retrograde pressure. The latter point is also confirmed by the neurosurgeon's findings of abnormal blood vessels in the lumbosacral region. Cerebral angiography could have clarified the question but was deferred due to the high disease burden and repeated complications (spinal hematoma causing spinal shock). Based on workup, we believe that visual loss phenomena are secondary to blood flow rather than actual inflammatory etiology as the ophthalmology team confirms no evidence of uveitis or maculopathy.

Spinal shock

What complicated the clinical condition of this unfortunate patient is intrathecal hematoma development. The compression on the spine and the cauda equina in the background of antiplatelet and anticoagulation led to a picture of spinal shock. The typical presentation of spinal shock is usually described in patients with severe spinal cord injury associated with refractory hypotension. The decrease in the sympathetic tone complicated the clinical condition of our patient with deficits and weakness of spine-related reflexes. In addition, we noticed a transient increase in blood pressure, probably due to catecholamine release, which is a well-known initial presentation of sympathetic overdrive, followed later by hypotension and urinary retention.

Vasculo-Behçet's disease

Vasculo-Behçet's disease continues to pose a significant diagnostic and therapeutic challenge to physicians due to the heterogeneity of the clinical presentation and lack of diagnostic laboratory tests. Therefore, we must consider/have a low threshold for it in any patient with vascular thrombosis or aneurysm. Lower extremity vein thrombosis is the most frequent manifestation of vasculo-Behçet's disease, followed by vena cava thrombosis, pulmonary artery aneurysms (PAA), Budd-Chiari syndrome, peripheral artery aneurysms, CVT, and abdominal aorta aneurysms were the other vascular manifestations as listed in order of decreasing frequency [10]. Coronary artery vasculitis is a rare finding in vasculo-Behçet's disease, reported primarily on case reports [11]. Our patient developed coronary artery vasculitis despite adherence to the prescribed anticoagulant monotherapy. After combining immunosuppressive therapy with the pre-existing anticoagulant therapy, he had no thrombosis recurrence. Thus, combined immunosuppressive and anticoagulant therapy is essential in suppressing and preventing venous and arterial recurrence attacks.

Conclusion

The story of this unfortunate young patient is unique in his high disease burden and complicated hospital course. Visual loss in patients with Behçet's disease should always be anticipated and taken reasonable care of, ensuring that they receive well-combined immunosuppression with anticoagulation and agents to reduce intracranial pressure. This is essential in suppressing and preventing the venous and arterial recurrence attacks in addition to surgery where indicated surgical correction. We believe the patient mandated an earlier optic nerve sheath fenestration and VP shunt to reduce his high intracranial pressure and preserve his vision. Lumbar drain removal should not have been done in a setting of ongoing heparin infusion and two antiplatelets. A high suspicion of hematoma causing acute spinal shock should have had prompted an urgent spine MRI. We also emphasize multidisciplinary teamwork. Is there an underlying syndrome and not being identified? Did the patient have a complete and timely manner investigations? Can a patient be managed without a definitive diagnosis?

Acknowledgment

We are deeply grateful to all the nursing staff of the National Neurosciences Institute for their outstanding care provided to the patients and other teams involved in the care of this patient, especially thrombosis, rheumatology, neurosurgery, ophthalmology, physical therapy, and occupational therapy teams, and nevertheless, the ancillary services of Medical Imaging and Laboratory departments.

Authors' Contribution Statement

Naim I. Kajtazi has contributed with case report design, planning, scanned files review, writing patient history, investigations, treatment, follow up, interpretation of data, intellectual content of manuscript, coordinated work with other co-authors, and submission on behalf of all of them Mohammed Al Sheef has contributed with literature review, discussion part and intellectual content of manuscript. Ehtesham Khalid has contributed with discussion part and intellectual content of manuscript, Juman Al Ghamdi has contributed with selection of good quality images with annotations from PACS, Majed Al Hameed has contributed with intellectual content of manuscript, Mohammed Bafaquh has contributed with case report design, literature review, discussion part and intellectual content of manuscript.

References

1. Margo KL, Margo GM (1994) The problem of somatization in family practice. Am Fam Physician. 49(8): 1873-1879.
2. Debnath M, Prasad GBKS, Bisen PS (2009) Diagnosis of Complex Diseases. Molecular Diagnostics: Promises and Possibilities. 347-382.
3. Berner ES, Graber ML (2000) Overconfidence as a cause of diagnostic error in medicine. Am J Med. 121(5 Suppl): S2-23.

How to cite this article: Naim I K, Mohammed Al S, Ehtesham K, Juman Al G, Majed Al H, and Mohammed B. The Dual Catastrophe of Behçet's Disease: Visual Loss Followed by Acute Spinal Shock After Lumbar Drain Removal. Open Access J Neurol Neurosurg 2021; 16(2): 555933
DOI: 10.19080/OAJNN.2021.16.555933

4. Goldman L, Sayson R, Robbins S, Cohn LH, Bettmann M, et al. (1983) The value of the autopsy in three medical eras. N Engl J Med 308(17):1000-1005.

5. Ameri A, Bousser MG (1992) Cerebral venous thrombosis. Neurol Clin 10(1): 87-111.

6. Purvin VA, Trobe JD, Kosmorsky G (1995) Neuro-ophthalmic features of cerebral venous obstruction. Arch Neurol 52(9): 880-885.

7. Zhao T, Wang G, Dai J, Liu Y, Wang Y, et al. (2018) Cases of visual impairment caused by cerebral venous sinus occlusion-induced intracranial hypertension in the absence of headache. BMC Neurol 18(1):159.

8. Taylor S, Singh J, Menezo V, Denis Wakefield, Peter McCluskey, (2011) Behçet disease: visual prognosis and factors influencing the development of visual loss. Am J Ophthalmol 152(6):1059-1066.

9. Gandhi D, Chen J, Pearl M (2012) AJNR Am J Neuroradol. 33(6):1007-1013.

10. Seyahi E, Yurdakul S (2011) Behçet's Syndrome and Thrombosis. Mediterr J Hematol Infect Dis 3(1): e2011026.

11. Mogulkoc N, Burgess MI, Bishop PW (2000) Intracardiac thrombus in Behçet's disease: a systematic review. Chest 118(2):479-487.

DOI: 10.19080/OAJNN.2021.16.555933

How to cite this article: Naim I K, Mohammed Al S, Ehtesham K, Juman Al G, Majed Al H, and Mohammed B. The Dual Catastrophe of Behçet's Disease: Visual Loss Followed by Acute Spinal Shock After Lumbar Drain Removal. Open Access J Neurol Neurosurg 2021; 16(2): 555933
DOI: 10.19080/OAJNN.2021.16.555933