Neuro-behçet’s disease (NBD) is an uncommon, serious presentation of behçet’s disease (BD) causing long-term morbidity and mortality. Cerebral aneurysms are rare in BD, with only a handful of cases reported worldwide. A 39-year-old female experienced slurring of speech, numbness, and reduced sensation in the left face, arm, and leg 90 minutes prior to presentation. She provided a history of recurrent oral and genital ulcers associated with intermittent joints pain and blurring of vision. Erythrocyte sedimentation rate was elevated, but all other autoimmune workup was negative. Neuro-behçet’s disease was diagnosed. Brain magnetic resonance imaging demonstrated acute right periventricular infarction. Magnetic resonance angiogram revealed M1 segment stenosis and right internal carotid artery saccular aneurysm. Catheter angiography confirmed its presence measuring 4.8 mm × 6.1 mm. She was stabilized after coiling of the aneurysm and was started on medical therapy. Brain imaging should be carried out in young patients with BD presenting with an ischemic event, and intervention may be lifesaving.

Disclosure. The authors declare no conflicting interests, support or funding from any drug company.
vision. There was no history suggestive of ophthalmic symptoms. Her family history was unremarkable. She was not a smoker. She was not taking any medications prior to presentation. She denied allergies to drugs or food.

On examination, her blood pressure was 188/100 mmHg with pulse rate of 76 bpm. She was alert and oriented to time, place, and person (language comprehensive and coherent). Her cranial nerves were intact. Her motor system was normal. She had decreased sensation to pain and temperature on the left side of body involving face, arm, and leg. Her cerebellar function and gait were normal. Her systemic examination did not reveal significant findings.

**Diagnostic assessment.** She was admitted for further workup and management. Her routine blood workup was within normal limits. Brain imaging with MRI was performed which demonstrated an acute infarction involving the right periventricular area. (Figure 1) Magnetic resonance angiogram revealed a stenosis of M1 segment of the right middle cerebral artery (MCA) and a saccular aneurysm of the cavernous portion of the right internal carotid artery (ICA). Due to aforementioned findings, catheter angiography was carried out for her, and it confirmed the presence of saccular aneurysm measuring 4.8 mm×6.1 mm projecting medially from the cavernous portion of right ICA. (Figure 2)

Her stroke workup showed cholesterol of 253 mg/dl,

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**Table 1 -** The patient’s progress summarized in a timely manner from day of admission until discharge.

| Dates     | Relevant past medical history and interventions                                                                 |
|-----------|---------------------------------------------------------------------------------------------------------------|
| 3rd Jan 2017 | 39-year-old female experienced slurring of speech, numbness, and reduced sensation in the left face, arm, and leg 90 minutes prior to presentation. She provided a history of recurrent oral and genital ulcers associated with intermittent joint pains and blurring of vision. Elevated inflammatory markers and negative autoimmune work up was exhibited. Neuro-behçet’s disease was diagnosed. Brain imaging demonstrated an acute stroke & MRA revealed M1 segment stenosis and right internal carotid artery saccular aneurysm. Catheter angiography confirmed its presence measuring 4.8 mm×6.1 mm. She was stabilized after coiling of the aneurysm and was started on medical therapy. |
| 8th Jan 2017 | MRI demonstrated acute right periventricular infarction.                                                      |
| 4th Jan 2017 | MRA revealed M1 segment stenosis and right internal carotid artery saccular aneurysm. Catheter angiography confirmed its presence measuring 4.8 mm×6.1 mm. |
| 3rd Jan 2017 | ESR was elevated, but all other autoimmune work up was negative.                                             |

| Dates     | Summaries from initial follow-up visits       | Diagnostic testing (including test dates)                  | Interventions                                        |
|-----------|-----------------------------------------------|----------------------------------------------------------|-----------------------------------------------------|
| 3rd Jan 2017 | Evaluated, but all other autoimmune work up was negative.                                                          | Started on aspirin and clopidogrel.                        |
| 4th Jan 2017 | MRI demonstrated acute right periventricular infarction.                                                      | No intervention.                                          |
| 4th Jan 2017 | MRA revealed M1 segment stenosis and right internal carotid artery saccular aneurysm. Catheter angiography confirmed its presence measuring 4.8 mm×6.1 mm. | No intervention.                                          |
| 8th Jan 2017 | Seen in the rheumatology clinic as an outpatient and was started on medical treatment.                         | Coiling of the right ICA aneurysm.                        |
| 4th Jan 2017 | MRI demonstrated acute right periventricular infarction.                                                      | Started on aspirin and clopidogrel.                        |
| 4th Jan 2017 | MRA revealed M1 segment stenosis and right internal carotid artery saccular aneurysm. Catheter angiography confirmed its presence measuring 4.8 mm×6.1 mm. | No intervention.                                          |
| 8th Jan 2017 | Seen in the rheumatology clinic as an outpatient and was started on medical treatment.                         | Coiling of the right ICA aneurysm.                        |
| 15th Jan 2017 | Patient did not report any symptoms recurrence or new neurologic symptoms on follow-up visits up to 8 months. | No intervention.                                          |

**NBD - Neuro-behçet's disease, MRA - Magnetic Resonance Angiography, ESR - Erythrocyte Sedimentation Rate, MRI - magnetic resonance imaging, ICA - internal carotid artery.**

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![Figure 1](image1.png) **Figure 1 -** Diffusion weighted imaging. A) showing hyperintense lesion involving right periventricular area with corresponding hypointense lesion on adjusted diffusion co-efficient sequence. B) suggestive of acute infarction of this area.

![Figure 2](image2.png) **Figure 2 -** Catheter angiogram of right ICA. A) pre-coiling showing aneurysm involving the cavernous portion of right ICA. B) post-coiling angiogram showing complete obliteration of right ICA aneurysm with coils visible inside aneurysm.
triglyceride 165 mg/dl, High-density lipoproteins (HDL) 42 mg/dl, and Low-density lipoprotein (LDL) 173 mg/dl. Her hemoglobin A1c was 6.2%. Her transthoracic as well as transesophageal echocardiogram were unremarkable. Her erythrocyte sedimentation rate (ESR) was 69 mm/hr, but all her autoimmune workup was negative. The computed tomography (CT) scan of her chest was unremarkable as well.

**Therapeutic intervention.** Thus, the diagnosis of NBD was presumed on the basis of her history. She was started on aspirin and clopidogrel, and her risk factors were controlled for secondary prevention of stroke. She underwent coiling of the right ICA aneurysm. She remained stable after the procedure, and she was discharged with advice to follow-up in clinic.

**Follow-up and outcomes.** She was started on azathioprine, and she did not report any symptoms recurrence or new neurologic symptoms on follow-up visits up to 8 months (Table 1).

**Discussion.** Our patient demonstrated stroke and cerebral aneurysm that might be the initial presentation of patients with BD, and imaging of brain vasculature should be considered as an essential workup for stroke in young people. The diagnosis of NBD is difficult in the absence of other criteria for BD. Thus, the diagnosis of NBD in our case was established based on the history of oral and genital ulcers (which were not present at the time of presentation), the presence of stroke-like episodes, the presence of a cerebral aneurysm, and the absence of other possible underlying causes.

Neuro-behçet's disease is one of the most serious presentations of BD and one of the most important

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Table 2 - Summary of 25 cases of neuro-behçet's disease with cerebral aneurysms.

| Author         | Year | Gender/Age | Ethnicity | Clinical manifestation       | Location                                      | Treatment                           |
|----------------|------|------------|-----------|------------------------------|----------------------------------------------|-------------------------------------|
| Katoh et al    | 1985 | M/29       | Japanese  | None                         | MCA                                          | Clipping                            |
| Buge et al     | 1987 | M/43       | Moroccan  | Hemiplegia                   | ACA, MCA, IC                                 | None                               |
| Kerr et al     | 1989 | M/12       | Caucasian | None                         | MCA, AC, PC                                  | Clipping                            |
| Tsuji et al    | 1990 | F/62       | Japanese  | Headache, decrease LOC, SAH  | Bilateral MCA, IC, ACommA                    | Clipping                            |
| Khodja et al   | 1991 | M/43       | Tunisian  | Lower limb weakness          | ACommA                                       | None                               |
| Dietl et al    | 1994 | F/47       | Turkish   | -                            | Bilateral IC                                 | Coiling                            |
| Ildan et al    | 1996 | M/28       | Turkish   | SAH                          | ACommA                                       | Clipping                            |
| Itoh et al     | 1996 | M/65       | Japanese  | Headache, ataxia             | Medullary infarction                         | -                                  |
| El Abbadi et al| 1999 | M/44       | Moroccan  | Stroke                       | MCA                                          | Clipping                            |
| Nakasu et al   | 2001 | M/57       | Japanese  | Decrease LOC, SAH, paresis   | Bilateral MCA                                | Clipping                            |
| Rosensting et al| 1990 |            | Caucasian | None                         | Superior cerebellar artery                   | Coiling                            |
| Kizilikic et al | 2003 | M/38       | Turkish   | SAH, headache                | Superior cerebellar artery, PICA             | Coiling                            |
| Kocak et al    | 2004 | M/37       | Turkish   | Headache, cerebral haematoma | MCA                                          | Clipping                            |
| Chi and Deruytter et al | 2005 | F/43       | Japanese  | Headache, seizure, SAH       | Superior cerebellar artery                    | Cranietomy                         |
| Kaku et al     | 2007 | F/19       | Japanese  | Headache, seizures, SAH, paresis | Bilateral MCA                              | Surgical treatment (excision and grafting) |
| Agrawal et al  | 2007 | F/36       | Indian    | SAH                          | IC                                           | Coiling                            |
| Aktas et al    | 2008 | M/38       | Turkish   | Headache, SAH                | Basilar artery                               | None                               |
| Ozveren et al  | 2009 | M/38       | Japanese  | Incidental, unruptured       | ICA                                          | Coiling                            |
| Bahar et al    | 2010 | M/36       | Turkish   | SAH                          | IC                                           | Coiling                            |
| Senel et al    | 2010 | M/45       | Turkish   | SAH                          | PCA                                          | None, spontaneously thrombosed     |
| Kurdi et al    | 2010 | M/26       | Saudi     | Headache, seizure, SAH       | MCA and PCA                                  | Onyx embolization                  |
| Younes et al   | 2013 | M/44       | Tunisian  | Hemiplegia                   | Multiple and bilateral cerebral arteries      | None                               |
| Ha et al       | 2016 | F/41       | Korean    | Headache, SAH                | Multiple bilateral MCA and ACA               | Clipping                            |
| Our case       | 2016 | F/39       | Saudi     | Paresthesia                  | MCA                                          | Coiling                            |

LOC - level of consciousness, SAH - subarachnoid hemorrhage, MCA - middle cerebral artery, ACA - anterior cerebral artery, IC - internal carotid, ACommA - anterior communicating artery, PCommA - posterior communicating artery, PICA - posterior inferior cerebellar artery, PCA - posterior cerebral artery.
causes of long-term morbidity and mortality. Shahien R et al., reported that the incidence of NBD is very variable, as it is to be 18% (range 4%-49%), but Türsen U et al., reported that it can also be as low as 1.3%. Cerebral aneurysms are very rare in BD, and its incidence and prevalence have not been well documented in the literature. Although the vasculitis is theorized as the typical pathogenesis in arterial involvement, its underlying mechanism in developing cerebral aneurysms is uncertain to this day. One study reported by Benamour et al., demonstrated only one case from 316 cases of BD that was identified as having a cerebral aneurysm. MRA is the most sensitive non-invasive method to screen for cerebral aneurysms, and cerebral angiogram and it remains as the gold standard.6

Table 2 summarizes 25 cases of NBD with cerebral aneurysms previously reported in the literature. Eighteen (75%) of the cases with NBD and cerebral aneurysm were related to those of the male gender, and the mean age of diagnosis of the aneurysm was 41.7 years of age, with ages ranging from 12-65. The majority of patients in these cases were reported to be of a Turkish descent with 8 patients, followed by Japan with 7 cases, then Tunisia and Morocco with 2 cases each. Patients of Saudi Arabian, Indian, Korean, Caucasian, and Armenian descent were reported with one case each. When it came to the clinical manifestation, the majority of the patients 11 (44%) presented with subarachnoid hemorrhage (SAH), and cerebrovascular disease was present in 4 (16%) of patients. Our case presented with with paresthesia alone. The aneurysm was unruptured in 3 (12%) of patients and were most commonly located on the MCA. In our patient, the aneurysm was in the cavernous portion of right ICA. The aneurysm treatment in these 25 patients was commonly located on the MCA. In our patient, the aneurysm was in the cavernous portion of right ICA. The aneurysm treatment in these 25 patients was commonly located on the MCA. In our patient, the aneurysm was in the cavernous portion of right ICA. The aneurysm treatment in these 25 patients was positively identified as having a cerebral aneurysm. MRA is the most sensitive non-invasive method to screen for cerebral aneurysms, and cerebral angiogram and it remains as the gold standard.

The use of adjuvant immunosuppressive therapy and steroids has also been reported, but there is a lack of consistency.6 The prognosis of ruptured intracranial aneurysms in patients with NBD remains unclear and is likely influenced by the severity of the hemorrhage and the course of the disease.

In conclusion, NBD presenting with a cerebral aneurysm is very rare and potentially life threatening. We believe such cases are often underdiagnosed and not reported. A high index of suspicion should be raised, and imaging of brain vasculature should be carried out in young patients with BD presenting with an ischemic event, and intervention may be potentially lifesaving in such patients.

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