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CLINICAL RESEARCH

A Case of Intracranial Aneurysm with a Stent Implantation - In A 9-Year-Old Girl with Tubersclerosis

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ABSTRACT

BACKGROUND

Tuberous sclerosis is a rare genetic multisystem disease with an autosomal dominant inheritance pattern. Till now, very much little is known about the incidence, prevalence and types, and other characteristics of intracranial aneurysms in the tuberous sclerosis complex. This paper presents a girl diagnosed with a tuberous sclerosis complex with an intracranial aneurysm who had a new onset of headache, ptosis, and blurry vision.

CASE SUMMARY

A nine-years old girl with a diagnosis of TSC was admitted to the pediatric neurology polyclinics with the complaint of a new onset of headache, ptosis, and blurry vision of the left eye. A Cerebral MRI conducted at the age of two displayed 6.1 mm mild ectasia of the cavernous segment of the left internal carotid artery. Time of flair magnetic resonance images, angiography, and digital subtraction angiography were performed. An aneurysm with fusiform configuration on the internal carotid artery was detected. A stent was implanted to prevent any intracranial bleeding.

CONCLUSION

The pathogenesis of intracranial aneurysms in tuberous sclerosis complex is still unclear. Any presentation as headache and ptosis in patient with tuberous sclerosis, intracranial aneurysms should be kept in mind. Time of flair magnetic resonance images screening tests should be performed in the routine to detect even the smallest aneurysms.

KEYWORDS

Tuberous sclerosis; Intracranial aneurysms; Headache; Ptosis

BACKGROUND

Tuberous Sclerosis Complex (TSC) is a rare genetic multisystem disease with an autosomal dominant inheritance that belongs to a group of phacomatoses. TSC occurs as a result of mutations in TSC1 (9q34) and TSC2 (16p13.3) genes, respectively [1,2].

One of the most stunning features of the TSC is white, colorless spots on the skin that are unclear, not puffy. Acne-like lesions that do not heal on the skin (angiofibromas). Arterial wall developmental disorders, such as aneurysms, in association with TSC, have been reported for extracranial vasculature. The intracranial aneurysm is rare but can be seen in these patients [3-5].

CASE PRESENTATION

A nine-year-old girl with a diagnosis of TSC was admitted to the pediatric neurology polyclinics with the complaint of a new onset of headache, ptosis, and blurry vision of the left eye. She was the second child of consanguineous parents. The elder sibling of the patients was normal. The mother's pregnancy was eventful due to gestational diabetes mellitus, and the child was born at a gestational age of 34 weeks and admitted to the newborn intensive care unit and incubated for ten days. Episodes of seizures started in late infancy, and the manifestation of psychomotor retardation was seen, and the diagnosis of TSC was made at the age of one. EEG showed spike waves at the left temporal region. Oxycarbamezepine, levetiracetam, and clobazam were used to control the seizures Magnetic resonance images (MRI) of the brain previously showed tuberous and hazardous lesions with leukomalacia lesions with bilateral subependymal nodules and calcifications. A Cerebral MRI conducted at the age of two displayed 6.1 mm mild ectasia of the cavernous segment of the left internal carotid artery (ICA). Due to the development of ptosis at the age of 9, MRI was conducted. It showed a 13.2 mm expansion of the ectasia (Figure 1).

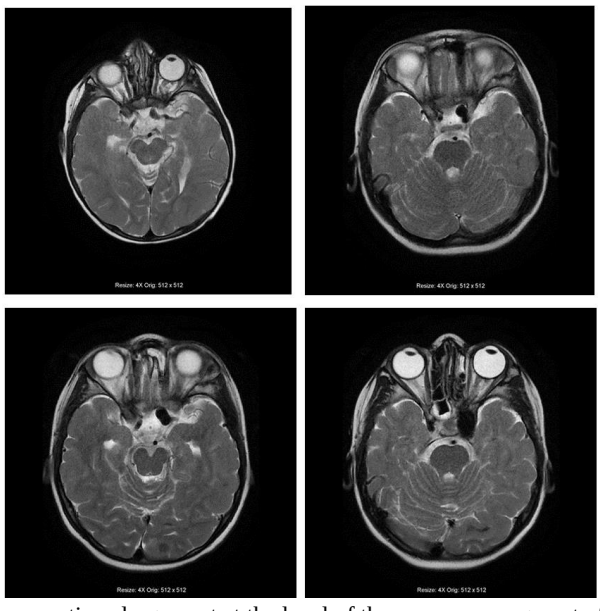

Figure 1: Progression of aneurysmatic enlargement at the level of the cavernous segment of the left internal carotid artery in axial images in T2-weighted sequence in the cerebral MR examination of the patient taken 1 year apart.

Diagnostic digital subtraction angiography (DSA) under sedation was conducted, fusiform aneurysmatic dilatation was detected in the cavernous segment of the left ICA to the end of the terminal segment of the posterior communicating artery. An endovascular treatment decision was taken at the multidisciplinary pediatric council to prevent thromboembolic complications. A binary antiaggregant treatment (asetilsalsalik acid tablet 81 mg/a day and clopidogrel tablet 75 mg per day) was initiated five days before starting the endovascular treatment. Consent of the parents was obtained. The endovascular treatment was applied under general anesthesia, 2 mg intravenous dexamethasone was administered during angiography. 2000 IU intravenous heparin was given as a bolus dose. After that, a maintenance dose of 250 IU intravenous heparin to bring activated coagulation time (ACT) up to 2 times the norm. Two surpass evolved current converter stent implantation with sizes of 4 mm × 30 mm and 4.5 mm × 40 mm, was done in telescopic configuration to include the aneurysmatic expansion (Figure 2).

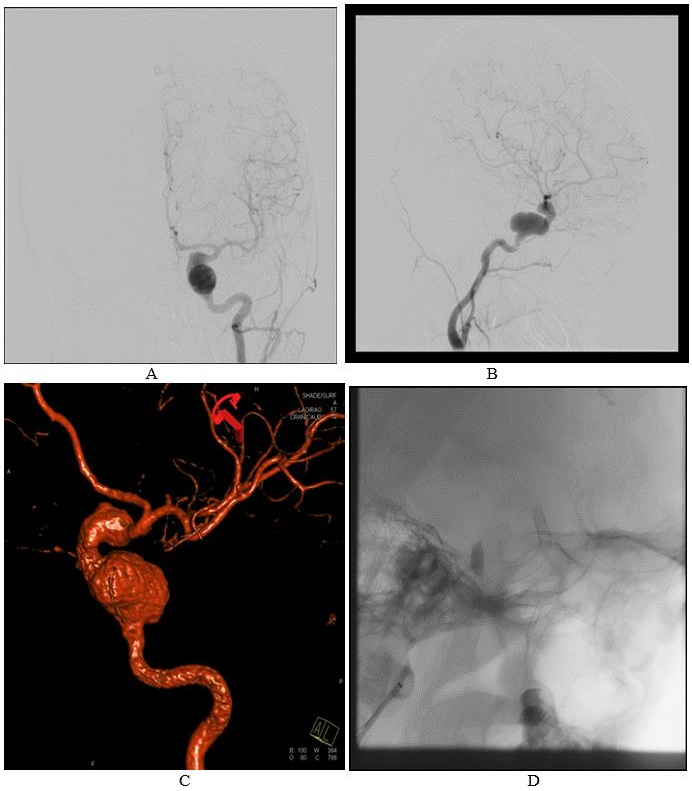

Figure 2: Images of the patient's fusiform aneurysm on digital subtraction angiography. A) DSA town projection image of aneurysmatic enlargement in the left ICA; B) DSA lateral projection image; C) 3D view; D) Stagnation image in the aneurysm after the implantation of the current converter device in the lateral projection of the DSA.

The patient was awakened without any problems. Intravenous heparin infusion was continued 24 hours in the pediatric intensive care unit. Diffusion MRI is conducted the following day. No acute ischemia was seen.

Even though dual antiaggregant were given to be used for six months, the family used dual antiaggregant for only one month, then stopped clopidogrel and continued with aspirin only. The patient came a few months after the treatment with seizures. Cerebral MRI was performed. It was seen that the ICA was occluded at the location of the flow converting stent. The patient had no new neurological symptoms. The DSA was performed to see the collateral circulation. It was noticed that the ICA (the anterior communicating) was occluded and the occluded side was bloodied, and there was venous drainage. Although ICA occlusion was an option during the first treatment, it was aimed to protect the ICA with a stent by having a council consultation decision because the

patient was young. The patient did not develop any neurological symptoms other than seizures because of the slow-developing occlusion rather than the acute occlusion (Figure 3).

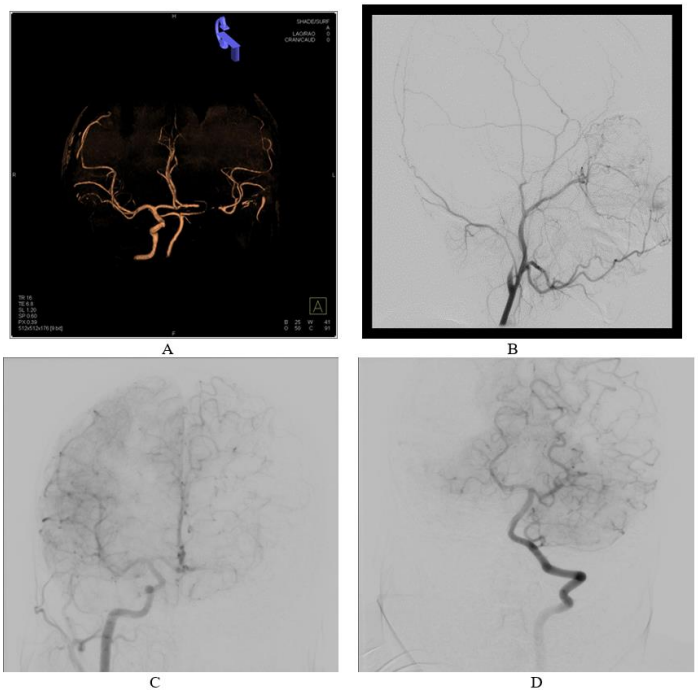

Figure 3: Follow-up imaging of the patient. A) No flow is observed in the left ICA in the MR angiography 3D time of flight (3D TOF) sequence; B) Occluded view of left ICA from proximal in lateral projection in DSA; C) The image in which the left cerebral hemisphere is vascularized through the anterior communicating artery in the right carotid injection in the town projection in DSA; D) View of retrograde collateral arteries seen towards the left cerebral hemisphere with left vertebral artery injection in town projection in DSA.

DISCUSSION

Cerebral aneurysms, also known as Intracranial Aneurysms (IA), are generally saccular or fusiform focal dilations in the wall of big arteries in the circle of Willis that grow and carry a certain risk of rupture. To date, arterial wall anomalies in TSC, especially in aneurysms, were only described in the extracranial vasculature, such as aortic aneurysms or renal aneurysms that were considered as the result of a congenital defect. Recently, there are sporadic cases that reported the coexistence of IA in TSC and hypothesized their congenital origin. In this paper, we present a rare case of a fusiform aneurysm in a nine-year-old girl with TSC [6,7].

The reported cases of IA in TSC showed a specific demographic pattern. The male/female ratio was 1.9:1, and 66.7% of the patients were under 18; among them, 36.4% were two years of age or younger. Incidental diagnosis of IA in patients with TSC was (36.4%) and (21.2%) due to the new onset of a neurological deficit. Rupture of IA was seen in only 7.1% of the cases. The most frequent IA location was the anterior circulation (85.7%) in favor of the ICA (61.9%), where aneurysms originated remotely from branching zones. Of the 42 IA, 57.1% were large (size: 10 mm - 24 mm) or giant (size: ≥25 mm) and 45.2% had a fusiform configuration. Multiple aneurysms were seen only in 21.2% of the cases, and rapid growth was described and documented only in 2 patients (6%). Our case was a female who was nine years old with a single big fusiform aneurysm of the cavernosıs segment of the left ICA to the end of the terminal segment of the posterior communicating artery. Two surpass evolved current converter stent implantation was implanted, which to the best of our knowledge, is the first TSC patient with IA that has been implanted a stent under DSA at the age of nine [1,8].

As no screening tests have been performed yet, the incidence of IA in TSC could be higher than reported. Besides, according to the recommendations of the 2012 TSC Consensus Conference, the MRI at diagnosis and every 1 years - 3 years until the age of 25 years do not involve a special sequence for the vascular system, so-called time-of-flair MR angiography (TOF-MRA) which could detect even the smallest aneurysms. In our case, the aneurysm was detected after the new onset of headache, ptosis, and blurred vision. There are no aneurysm screening tests are used in the routine. So maybe many aneurysms might be missed because of the non-established routine screening tests [8].

CONCLUSION

The pathogenesis of intracranial aneurysms in tuberous sclerosis complex is still unclear. Any presentation as headache and ptosis in patient with tuberous sclerosis, intracranial aneurysms should be kept in mind. Time of flair magnetic resonance images screening tests should be performed in the routine to detect even the smallest aneurysms.

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Both SB and GGM authors have participated sufficiently in the submission and take public responsibility for its content. SB: writing the manuscript. HBO: selecting the images and corresponding with the journal. DY, OH and SA authors read and approved the final manuscript.

AVAILABILITY OF DATA AND MATERIALS

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study (as this is a case report).

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

The manuscript obtained ethical review exemption from the ethical review committee (ERC) of the authors' institution as case reports are exempt from review according to the institutional ethical review committee's policy. Written consent was obtained from the participants for publishing the case.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patients' legal guardians for publication of this case report and any accompanying images.

COMPETING INTERESTS

The authors declare that they have no competing interests. The authors declare no conflict of interest in preparing this article.

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