Multiple little intracranian aneurysms as familial inheritance

Valentin Munteanu[1], Mircea R. Gorgan[2], Radu Stanescu[3], Vasile Ciubotaru[4]

[1]PhD Student in Neurosurgery, "Carol Davila" UMPh, Faculty of Medicine, Departament of Neurosurgery
[2]Professor of Neurosurgery, "Carol Davila" UMPh, "Arseni – Bagdasar" Hospital
[3]Resident in Neurosurgery, "Bagdasar Arseni" Hospital, Bucharest
[4]Senior Neurosurgeon, "Bagdasar Arseni" Hospital, Bucharest

Abstract

In this particular case, the authors have studied a family in wich two of the members on the same genetic line-mother and daughter have suffered intracranial bleeding from ruptured aneurysm. The congenital nature and the patterns of inheritance of the disease are discussed. The indications for elective investigation of the asymptomatic relatives and surgical prophylaxis on asymptomatic aneurysms are also discussed.

Material and method: Two cases, same hereditary line, same nosocomial pattern, rare case of multiple intracranial aneuysms. Surgical treatment of ruptured and unruptered aneurysm, same part, same time.

Results: Operated pacient done well without neurological deficits after one year.

Conclusions: Making good judgment based on complete investigations lead to a good aoutcome. Further investigations on family hereditary aneurismal lesions should be perform.

Keywords: Multiple intracranial aneurysms, familial aneurysms, asymptomatic aneurysms, surgery

Introduction

Familial intracranial aneurysms are uncommon. Multiple familial intracranial aneurysms are much more uncommon. According to our review of the literature, only few families are known to have more than one of their members with multiple aneurysms. Our report describes a family in which mother and daughter had multiple intracranial aneurysms.

Case Report

A 39-year-old woman was admitted in our neurosurgical department. The night prior to admission he awoke, complaining of severe headache accompanied by vomiting and followed by loss of consciousness lasting about ten minutes. On admission the patient was conscious but with severe headache and confused.

Neurological examination revealed only neck stiffness.

Blood pressure was 160/80 mmHg. Laboratory data were normal. Lumbar puncture show bloody spinal fluid under elevated pressure.

A CT scan was perform one hour after the onset of headache at the first hospital where she has been addressed to, and it was repeated three hours later, at the arrival in our department. On all CT scan images it was the same subarachnoidian hemorrhage picture, unchanged.

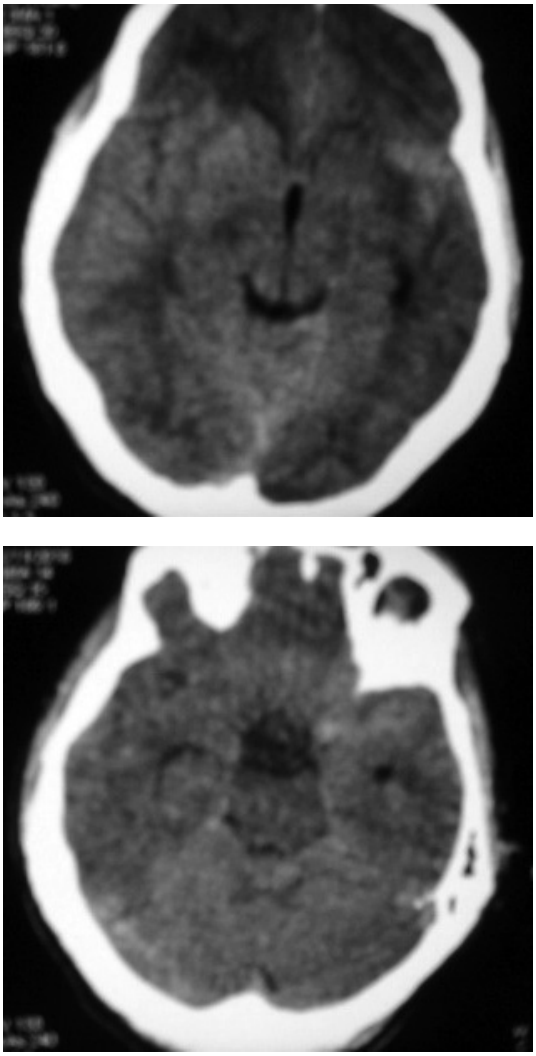

Figure 1 CT scan with minimal subarachnoid hemorrhage predominant on the left side

Four-vessel angiography disclosed multiple intracranial aneurysms, atotal of 6 aneurysms. On the right side 1,78 mm C6-carotid aneurism with 3,86 neck, under the PCoA origin, aneurismal dilatation also at the C6 – carotid level nearby the ophthalmic origin, aneurismal dilatation at the PCoA origin. On the left side aneurism with 2,2 maximal diameter and 3,45 mm neck at the ICA bifurcation, ruptured aneurism at the MCA bifurcation, with daughter sac 4,3-3,39 and 2,74 neck, aneurismal dilatation at the PCoA origin.

Also we perform angio-CT wich we believe not only disclosed the number and pattern of the aneurysms but, make the surgical plans more accurate. In this case particulary we have to find and choose the side where the bleeding was and wich aneurism has been bled, and base on all data we choose that the ICA bifurcation aneurism bled, being the only one ruptured, but we judged that also the MCA bifurcation aneurism is at high risk of rupture so we decide to operate on the left side.

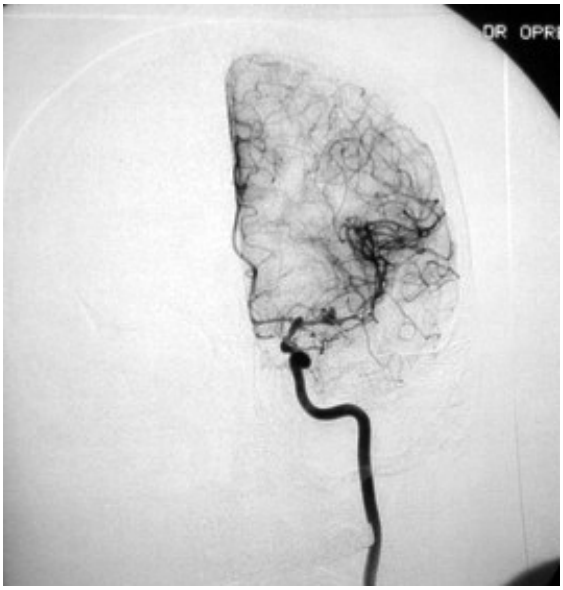

Figure 2 Four vessels cerebral angiography is routinely perform, but is very usefull in multiple aneurysms a 3D image of the lesions

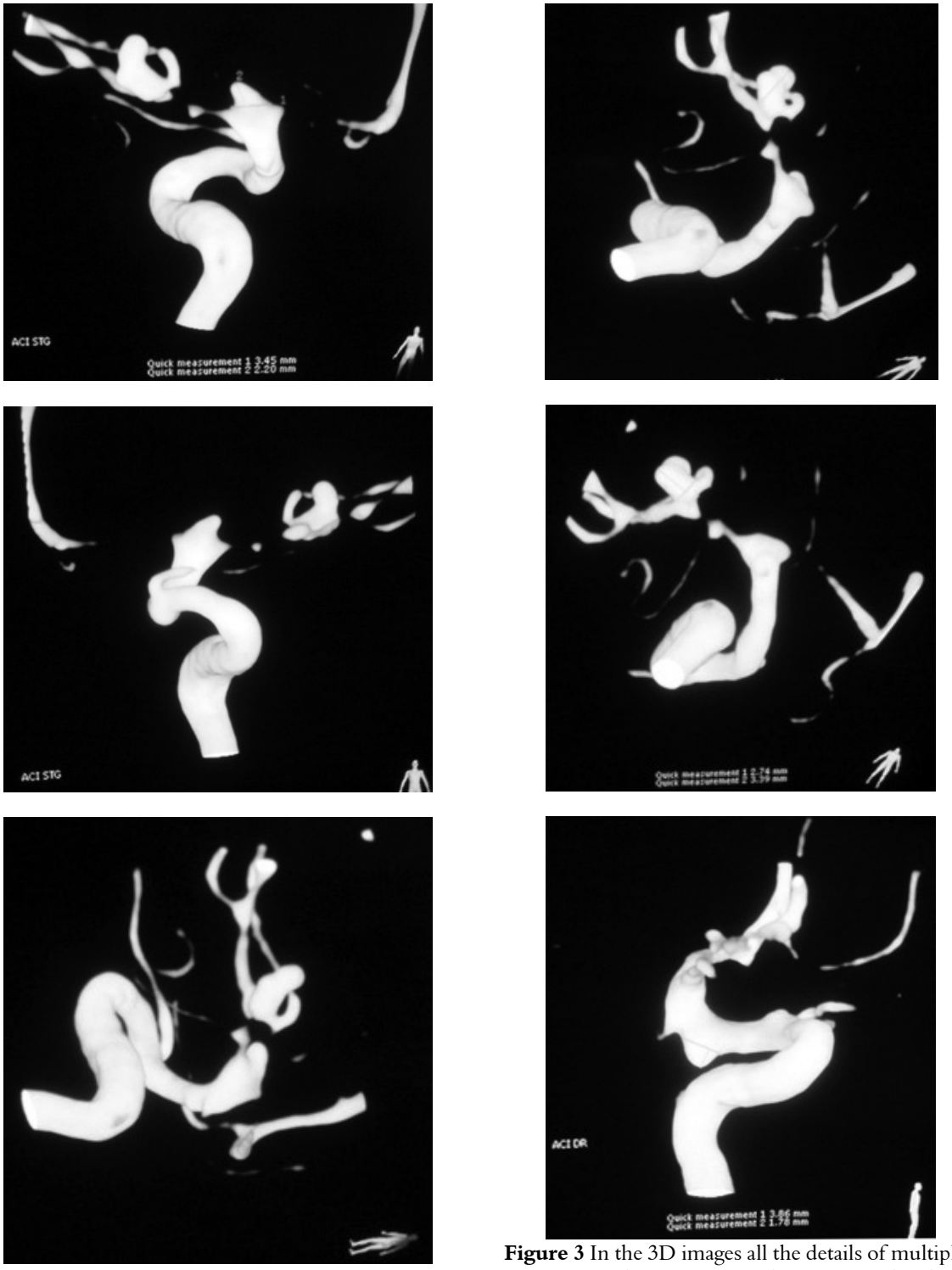

Figure 3 In the 3D images all the details of multiples intracranial aneurysms can be appreciated and a precise surgical approach can be done

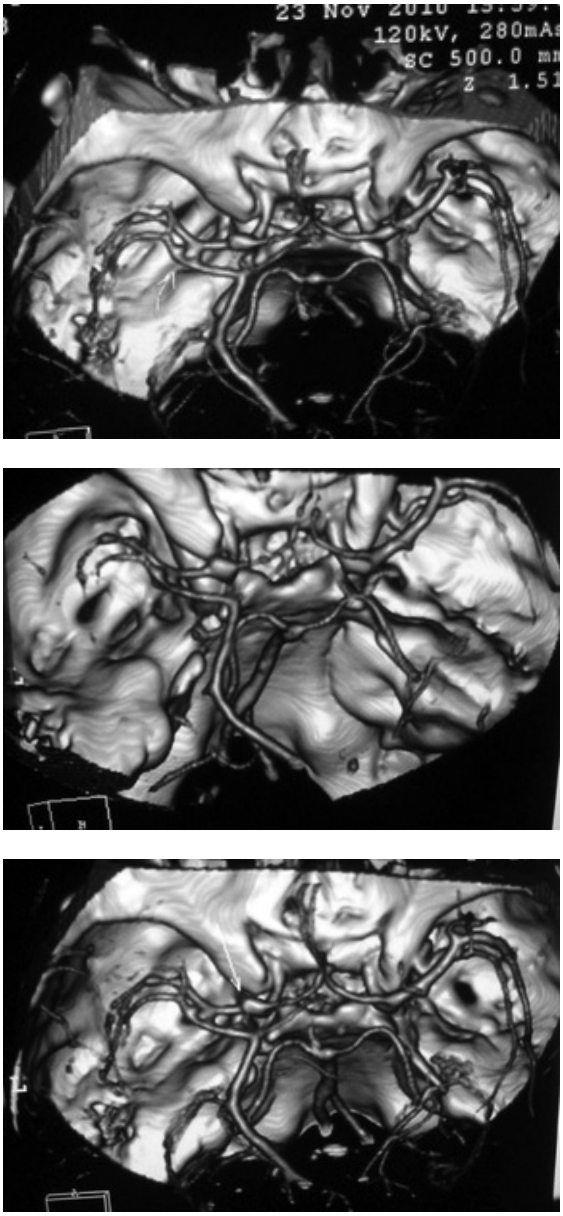

Figure 4 Angio CT with very clear aspect of the aneurysms size and location

The patient underwent surgery through a left fronto-pterional approach, and two of the aneurysms were successfully clipped the aneurism situated at the level of the ICA bifurcation wich was the one who bled, and the aneurism situated at the level of the MCA bifurcation. The postoperative course was of an improving neurological status but with motor aphasia lasting about three weeks, which recovered partially during the hospitalization. Overall now the patient is well, without any other neurological deficit.

For the mother case, the data has been received by family anamnestic interview wich disclosed severe intracranial hemorrhage with gcs 6 at presentation 23 years ago, operated after she has been diagnosed on angiography with multiple intracranial aneurisms but unfortunately died 4 days after one bled carotid aneurism has been clipped. Our patient has been operated one year ago but because of some personal, social issues, did not come back to allow us to make proper angiographic controls, only telephonic survey has been done.

Discussion

The congenital nature of intracranial aneurysms is generally assumed, and seems to be due to a maldevelopement of the embryonic vasculature resulting in a deficiency of the elastic layer at the bifurcation of a vessel (1, 3). Whether hereditary factors play a role in the origin of cerebral aneurysms is still uncertain. Intracranial aneurysms are not uncommonly associated with congenital malformations and other disorders, such as polycystic kidneys, arteriovenous malformations, coarctation of the aorta, Ehlers-Danlos syndrome, fibromuscolar hyperplasia of arteries, and possibly other connective tissue diseases (3).

A very interesting part of these pathology is because there are cases reported of familial aneurysms located in identical positions on the same side of the head. There are identical twins wich have been reported having multiple intracranial aneurysms; these findings suggest that

hereditary factors do have a specific role in some exemples of familial aneurysms,

An autosomal dominant inheritance, with variations in penetrance, seems to offer the best explanation and a detailed prospective study of the families with more than one member having intracranial aneurysms would be very helpful to be studied in the future (4).

There is still disagreement as to whether a relative of a patient affected by intracranial aneurysms has an increased risk of having an asymptomatic aneurysm. It has been stated that there is not any increased risk to a relative (2, 5, 7). However, review of the literature indicates that there are many reported cases in which the presence of an intracranial familial aneurysm, although strongly suspected, was not verified by angiography, surgery, or autopsy. Also, in some well-documented families, members are reported suddenly dead in whom the likely presence of ruptured intracranial aneurysms remained unexplored due to a lack of post-mortem examination. These findings suggest that the actual incidence of familial aneurysms is very likely to be higher than has been reported.

Another important dispute concerns the indication for cerebral angiography on asymptomatic members and surgical prophylaxis for asymptomatic aneurysms found by elective investigation. Asymptomatic familial aneurysms found by elective angiography have been reported and well documented (2, 6). In view of the good results of the surgical treatment of intact cerebral aneurysms, in japanese studies (5, 7) it has been suggested that elective investigation of the asymptomatic members should be considered when there are already two or more individuals affected in a family.

We consider that is not possible, at least yet to justify and perform cerebral angiography on all asymptomatic relatives. We think that families in wich a pacient with multiple aneurisms has been disclosed, all relatives on the direct hereditary line shoud be advise of being alert of any new symptoms that will possibly appear and notice their physician.

Elective investigation should be performed as soon as any warning signs, even headaches, appear. It should be done periodically neurological examinations of the asymptomatic members in order to prevent, if possible, bleeding by trying to disclose any early neurological warning signs.

References

1. Unruptured intracranial aneurysms--risk of rupture and risks of surgical intervention. International Study of Unruptured Intracranial Aneurysms Investigators. N Engl J Med 339:1725-1733, 1998.
2. Campi A, Ramzi N, Molyneux AJ, Summers PE, Kerr RS, Sneade M, Yarnold JA, Rischmiller J, Byrne JV: Retreatment of ruptured cerebral aneurysms in patients randomized by coiling or clipping in the International Subarachnoid Aneurysm Trial (ISAT). Stroke 38:1538-1544, 2007.
3. Harris PC, Ward CJ, Peral B, Hughes J: Polycystic kidney disease. 1: Identification and analysis of the primary defect. J Am Soc Nephrol 6:1125-1133, 1995.
4. Hassler O: Morphological studies on the large cerebral arteries, with reference to the aetiology of subarachnoid haemorrhage. Acta Psychiatr Scand Suppl 154:1-145, 1961.
5. Kaminogo M, Yonekura M, Shibata S: Incidence and outcome of multiple intracranial aneurysms in a defined population. Stroke 34:16-21, 2003.
6. Molyneux AJ, Kerr RS, Birks J, Ramzi N, Yarnold J, Sneade M, Rischmiller J: Risk of recurrent subarachnoid haemorrhage, death, or dependence and standardised mortality ratios after clipping or coiling of an intracranial aneurysm in the International Subarachnoid Aneurysm Trial (ISAT): long-term follow-up. Lancet Neurol 8:427-433, 2009.
7. Quinones-Hinojosa A, Gulati M, Singh V, Lawton MT: Spontaneous intracerebral hemorrhage due to coagulation disorders. Neurosurg Focus 15:E3, 2003.