

## A Foix's syndrome revealing "mirror" giant intracavernous aneurysms An illustrative case report of a therapeutic dilemma

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*Key words* : Intracranial aneurysm ; cavernous sinus ; ophthalmoplegia.

### Introduction

Intracavernous carotid artery aneurysms (ICAA) may generate a myriad of symptoms according as they enlarge or they rupture (1-3). In this latter situation, these aneurysms don't produce subarachnoid hemorrhage, but usually cause a carotid-cavernous sinus fistula (4) or rarely an epistaxis. More classically, ICAA can display a variety of neurological deficits, secondary to their growth within the cavernous sinus, including diplopia, oculomotor nerve pareses, decreased visual acuity, facial anesthesia and facial pain (5, 6). "Mirror" ICAA remain rare (7, 8). Their bilateralism is the starting-point of a therapeutic dilemma. The treatment options include selective embolization, detachable balloons, microneurosurgical exclusion, and carotid artery ligation, but the life-threatening risk of stroke has pushed some authors to recommend no treatment at all (3, 9).

The authors present the case of a 52 year-old woman in whom a Foix's syndrome revealed mirror ICAA and an associated other asymptomatic cervical carotid aneurysm, leading to a dilemma concerning the best therapeutic attitude.

### Case report

A 52 year-old woman, without any specific medical history, was admitted in the department of neurosurgery because of a right ptosis associated with an intense right facial pain outlining the ophthalmic territory of the fifth cranial nerve. The patient was perfectly awake, without signs of meningeal irritation. The cranial nerves examination revealed a complete right ophthalmoplegia, a right areflectic mydriasis without blindness and an ophthalmic and corneal hypoesthesia. There were no similar cases in the patient's family or systemic signs that could be linked to a connective tissue disease (Marfan's syndrome, Ehler-Danlos disease, polycystic kidney

disease). The CT-scan disclosed bilateral quite symmetric expanding processes within the two cavernous sinuses, with an homogeneous and intense contrast enhancing on the left side, and a partial enhancing on the right side, evoking bilateral intracavernous aneurysms of which the right one was partially thrombosed. MRI and MRA (Figs. 1, 2, 3) confirmed the diagnosis of multiple intra and extracranial aneurysms including two "mirror" ICAA and a third cervical carotid artery aneurysm. MRI confirmed that the right ICAA was partially thrombosed. A conventional angiography (Fig. 4) done 4 days later, while the clinical state of the patient began to improve, has showed a complete spontaneous thrombosis of the right internal carotid artery including the ICAA. The patient refused any clinical balloon test occlusion or preventive treatment of the remaining asymptomatic aneurysms. Fifteen days after the onset, a spontaneous improvement of the right Foix's syndrome was noticed : An ophthalmic pain resolution under 400 mg/day of Carbamazepin and a residual right tiny ophthalmoparesis, with the disappearance of the ptosis and the pupillary anomalies. At three months of follow-up, the Carbamazepin was progressively decreased, the right Foix's syndrome completely resolved and the patient refused any further preventive treatment concerning the remaining aneurysms.

### Discussion

The first description of an injury of the cavernous sinus external wall is imputable to Foix who described in 1922 a clinical syndrome involving the third, the fourth and the sixth cranial nerves but also the ophthalmic and the maxillary branches of the trigeminal nerve, resulting in a unilateral ophthalmoplegia and a trigeminal neuralgia (10). This syndrome may be produced by a wide variety of diseases such as cavernous sinus tumors, aneurysms of the intracavernous carotid artery, and even inflammatory processes (6). Besides this Foix syndrome, bulky ICAA can also cause a visual impairment by compressive optic neuropathy (5).

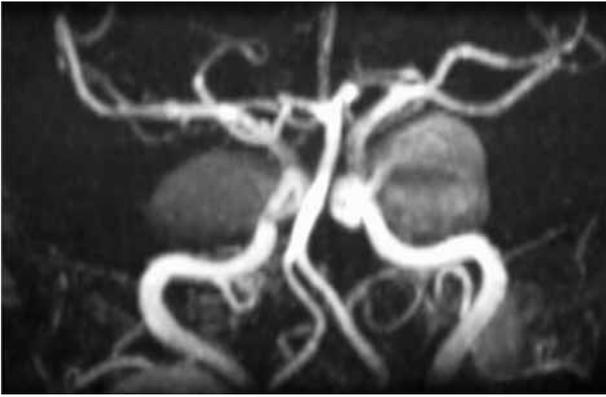


FIG. 1. — Frontal MR Angiogram showing two mirror aneurysms of the cavernous portion of both internal carotid arteries.

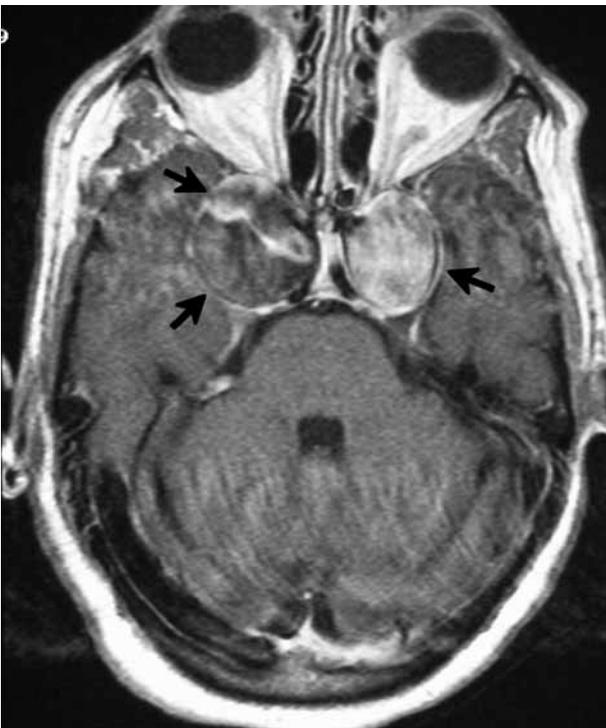


FIG. 2. — Axial T1 MRI after gadolinium displaying the aneurismal pouch within the two cavernous sinuses (arrows) partially thrombosed on the right side.

About one third of patients presenting with subarachnoid hemorrhage have multiple intracranial aneurysms (11). The bilateral symmetrical disposition of multiple intracranial aneurysms led to the denomination of “mirror aneurysms”. These latter may arise from a bilateral embryological developmental anomaly of the vessels wall (11). Mirror aneurysms represent about 11% of many ICAA clinical series (3, 9). Several papers dealt with the identification of risk factors for multiple intracranial aneurysms (11, 12). Among these factors, we can quote regular cigarette smoking, female sex, arterial hypertension, and postmenopausal state (11). Otherwise, several hereditary connective tissue disorders have been identified as major risk

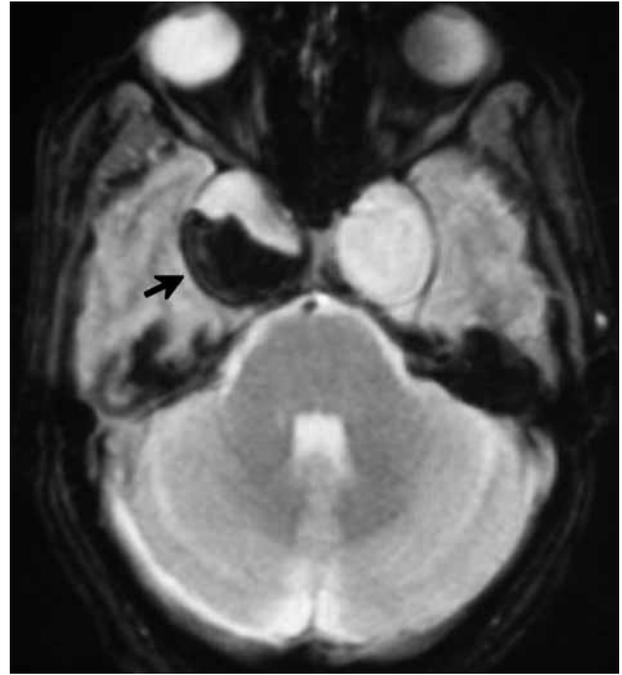


FIG. 3. — Axial proton density sequences (MRI) confirming the partial thrombosis of the right aneurismal sac (arrow).

factors for multiple intracranial aneurysms such as Marfan syndrome, Ehlers-Danlos syndrome, polycystic kidney disease and pseudoxanthoma elasticum. Finally, the existence of arteriosclerosis or fibromuscular dysplasia may generate ICAA, predisposing to arterial dissection episodes and even spontaneous thrombosis, as hypothesized in our case.

When ICAA rupture, they usually give birth to a carotid-cavernous sinus fistula (4). In this particular case, the treatment is well codified, using current endovascular techniques with a low morbidity (4). Contrary to their ruptured counterparts, the treatment of the unruptured ICAA remains problematic (9, 16), especially in the case of bilateral ICAA as in our patient. The therapeutic approach encloses abstention, endovascular selective embolization, microsurgical exclusion, and carotid artery ligation (7, 8). Some authors advocated the treatment of both aneurysms in the specific case of “mirror” ICAA (7, 8). Slaba *et al.* reported the sequential treatment of both mirror aneurysms, using a selective embolization on one side and detachable balloons on the other side, achieving a good result (7). Faria *et al.* described a disconcerting successful bilateral common carotid ligation in a patient with bilateral giant ICAA, similar to our case, with a good recovery (8).

As ICAA are rarely associated with life-threatening complications, therapeutic indications should be discussed in each singular case. Many ICAA have a “benign” natural history, leading to a spontaneous clinical improvement as noticed in our patient (3, 9). Among the 70 untreated ICAA



FIG. 4. — A frontal digital subtraction angiography 4 days after the onset, showing thrombosis of the right internal carotid including the right ICAA (black arrow), while the left ICAA and the right cervical carotid aneurysm are well injected.

patients of Stiebel-Kalish *et al.* who presented with diplopia, 56% of them spontaneously improved (3). The same rate of improvement was reported concerning the pain in the untreated group.

Nevertheless, the development of the endovascular techniques urged many authors to be more interventionist in front of ICAA (5, 17, 18). Among these techniques, the internal carotid occlusion is the most important and definitive treatment of complex giant ICAA (18). Trapping of the aneurysm was preferred to proximal occlusion when a retrograde filling of the aneurysm was angiographically demonstrated during the balloon test occlusion (BTO) (17).

The therapeutic internal carotid occlusion requires clinical BTO (13, 14, 17-19). This test is mandatory but seems insufficient (19). Internal carotid occlusion for ICAA can lead to life-threatening ischemic complications despite normal clinical BTO (17-19). Many pretherapeutic protocols were developed, including cerebral blood flow analysis with single photon emission computerized tomography (SPECT) during the balloon test occlusion (17-19), and angiographic test occlusion that seems to be a safe and easy procedure (20). Field *et al.* (19) classified their patients having ICAA planned for carotid occlusion according to the cerebral blood flow analysis in low risk (cortical CBF greater than 30 ml/100 g/min) and moderate risk (cortical CBF below 30 ml/100 g/min) for developing post-occlusion infarction. This classification permitted to indicate carotid occlusion alone

in low risk patients, while the moderate risk patients should undergo cerebral revascularization prior to carotid occlusion (19). Among fifteen patients with giant paraclinoidal aneurysms and ICAA, Do Souto (18) reported three patients who needed an extracranial-intracranial saphenous vein bypass before internal carotid occlusion, based on the analysis of the BTO associated with the data of the SPECT. Other authors reported their experience in the ICAA treatment with the use of extracranial-intracranial bypasses associated with the aneurysm trapping, leading to an 87,5% rate of excellent outcomes (21).

Alternatively, protection balloons and self-expandable stents are developed to assist the coiling of complex and giant ICAA (15). These techniques allow remodelling of the parent vessel, without compromising the cerebral blood flow and avoiding the coil bulging within the parent artery in cases of wide neck aneurysms (15, 22). However, long-term outcome of stent-assisted coiling of giant wide neck aneurysms is still to be evaluated.

## Conclusion

Since their natural history has been said to be benign, many clinicians have recommended that the ICAA should not be treated at all. Treatment should be rigorously debated in patients with intractable facial or retro-orbital pain not controlled with medication, progressive visual loss, carotid cavernous fistula or cerebral embolus from the aneurysm. Parent vessel occlusion is the best therapeutic option with well established good long term results. If balloon test occlusion is not well tolerated, balloon or stent assisted coiling might be a valuable option.

## REFERENCES

1. LINSKEY M. E., SEKHAR I. N., HIRSCH W. *et al.* Aneurysms of the intracavernous carotid artery : clinical presentation, radiographic features, and pathogenesis. *Neurosurgery*, 1990, **26** : 71-79.
2. LINSKEY M. E., SEKHAR I. N., HIRSCH W. *et al.* Aneurysms of the intracavernous carotid artery : natural history and indications for treatment. *Neurosurgery*, 1990, **26** : 933-8.
3. STIEBEL-KALISH H., KALISH Y., BAR-ON R. H., SETTON A., NIIMI Y., BERENSTEIN A., KUPERSMITH M. J. Presentation, natural history, and management of carotid cavernous aneurysms. *Neurosurgery*, 2005, **57** : 850-857.
4. MILLER N. R. Carotid-cavernous sinus fistula. In : MILLER N. R., NEWMAN N. (eds.). *Welsh & Hoyt's clinical neuro-ophthalmology*. Baltimore : Williams and Wilkins, 1997 : 3263-322.
5. GOLDENBERG-COHEN N., CURRY C., MILLER N. R. *et al.* Long term visual and neurological prognosis in patients with treated and untreated cavernous sinus aneurysms. *J. Neurol. Neurosurg. Psychiatry*, 2004, **75** : 863-867.

6. TRABE J. D., GLASER J. S., POST J. D. Meningiomas and aneurysms of the cavernous sinus. Neuro-ophthalmologic features. *Arch. Ophthalmol.*, 1978, **96** : 457-67.
7. SLABA S., SMAYRA T., MOHASSEB G. *et al.* "Mirror" intracavernous aneurysms. Therapeutic dilemma apropos of a case with review of the literature. *Neurochirurgie*, 1997, **43** (5) : 325-9.
8. FARIA M. A. Jr, FLEISCHER A. S., SPECTOR R. H. Bilateral giant intracavernous carotid aneurysms treated by bilateral carotid ligation. *Surg. Neurol.*, 1980, **14** (3) : 207-10.
9. KUPERSMITH M. J., HURST R., BERENSTEIN A. *et al.* The benign course of cavernous carotid artery aneurysms. *J. Neurosurg.*, 1992, **77** (5) : 690-3.
10. FOIX C. Syndrome de la paroi externe du sinus caveux (ophtalmoplégie unilatérale à marche rapidement progressive. Algie du territoire de l'ophtalmique). Amélioration considérable par le traitement radiothérapique. *Revue neurologique*, 1922, **38** : 827-832.
11. ELLAMUSHI H. E., GRIEVE J. P., JÄGER R., KITCHEN N. D. Risk factors for the formation of multiple intracranial aneurysms. *J. Neurosurg.*, 2001, **94** : 728-732.
12. JUVELA S. Risk factors for multiple intracranial aneurysms. *Stroke*, 2000, **31** : 392-397.
13. LARSON J. J., TEW J. M., TAMSICK T. A. *et al.* Treatment of aneurysms of the internal carotid artery by intravascular balloon occlusion : long-term follow-up of 58 patients. *Neurosurgery*, 1995, **36** : 26-30.
14. NIRO M., SHIMOZURU T., NAKAMURA K. *et al.* Long-term follow-up study of patients with cavernous sinus aneurysm treated by proximal occlusion. *Neurol. Med. Chir. (Tokyo)*, 2000, **40** : 88-97.
15. ZENTENO M., FREITAS J. M. M., ABURTO-MURRIETA Y., KOPPE G., MACHADO E., LEE A. Balloon-expandable stenting with and without coiling for wide-neck and complex aneurysms. *Surg. Neurol.*, 2006, **66** : 603-610.
16. LINSKEY M. E., SEKHAR I. N. Which cavernous sinus aneurysms should be treated ? *J. Neurosurg.*, 1993, **78** : 1008-9.
17. KAI Y., HAMADA J. I., MORIOKA M., YANO S., MIZUNO T., KURODA J. I. *et al.* Treatment strategy for giant aneurysms in the cavernous portion of the internal carotid artery. *Surg. Neurol.*, 2007, **67** : 148-155.
18. DO SOUTO A. A., DOMINGUES F. S., ESPINOSA G., WAINBERG E., CHAGAS H., TRAGANTE R. *et al.* Complex paraclinoidal and giant cavernous aneurysms. *Arq. Neuropsiquiatr.*, 2006, **64** : 768-773.
19. FIELD M., JUNGREIS C. A., CHENGELIS N. *et al.* Symptomatic cavernous sinus aneurysms : management and outcome after carotid occlusion and selective cerebral revascularization. *AJNR Am. J. Neuroradiol.*, 2003, **24** (6) : 1200-7.
20. VAN ROOIJ W. J., SLUZIEWSKI M., SLOB M. J., RINKEL G. J. Predictive value of angiographic testing for tolerance to therapeutic occlusion of the carotid artery. *AJNR Am. J. Neuroradiol.*, 2005, **26** : 175-8.
21. CHIBBARO S., TACCONI L. Extracranial-intracranial bypass for the treatment of cavernous sinus aneurysms. *J. Clin. Neurosci.*, 2006, **13** : 1001-1005.
22. DERDEYN C. P., CROSS D. T., MORAN C. J. *et al.* Postprocedure ischemic events after treatment of intracranial aneurysms with Guglielmi detachable coils. *J. Neurosurg.*, 2002, **96** : 837-43.

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