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SPONTANEOUS THROMBOSIS OF INTERNAL CAROTID ARTERY

A natural history of giant carotid cavernous aneurysms

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Abstract – **Objective:** To describe five cases of giant carotid cavernous aneurysms which evolved with spontaneous thrombosis of internal carotid artery (STICA), with emphasis at epidemiology, clinical presentation, natural history, related factors and neurological outcome. **Method:** There were 711 consecutives patients with 802 aneurysms with and without surgical treatment during a period of 19 years. We selected 35 patients with 40 carotid cavernous aneurysms (5%) of which 20 (50%) were giant aneurysms. Among those cases, 5 patients evolved with STICA (25%). Symptoms and findings at presentation were recorded and compared with those at outcome. **Results:** Clinical presentation was commonly related to atherosclerotic factors such as elevated blood pressure (80%), diabetes mellitus (40%) and dislipidemy (40%). All patients presented with hemicranial headache, ophthalmparesy and retro bulbar pain, and after STICA all presented improvement of symptoms. After STICA, 4 patients had regression of deficit, 2 partial and 2 complete. Four patients had sensorial trigeminal neuropathy in V1 and V2 territories, also showing improvement of symptoms after STICA. **Conclusion:** STICA is a common outcome in giant carotid cavernous aneurysms, and is related with significant improvement of symptoms; however, it may be catastrophic for those patients without efficient collateral circulation.

KEY WORDS: carotid cavernous aneurysms, carotid thrombosis, atherosclerosis.

Trombose espontânea da artéria carótida interna: a história natural dos aneurismas gigantes intracavernosos

Resumo – **Objetivo:** Relatar cinco casos de aneurismas gigantes intracavernosos que evoluíram com trombose espontânea da artéria carótida interna (TEACI), estudando-se: prevalência, apresentação clínica, história natural, fatores associados e prognóstico neurológico. **Método:** Análise de 711 pacientes consecutivos com diagnóstico de 802 aneurismas cerebrais submetidos a tratamento clínico ou cirúrgico num período de 19 anos. Foram identificados 40 aneurismas intracavernosos, sendo que 20 desses eram gigantes. Dentre esses, 5 pacientes com aneurismas gigantes intracavernosos que evoluíram com TEACI (25%). Os sintomas e sinais neurológicos da apresentação foram registrados e comparados ao término do acompanhamento. **Resultados:** Todos pacientes apresentavam cefaléia hemicraniana, apresentavam oftalmoplegia e dor retro-orbitária. Freqüentemente estavam associados a fatores ateroscleróticos como HAS (80%), diabetes melito (40%) e dislipidemia (40%) e após a TEACI evoluíram com melhora desses sintomas. Após a TEACI, 4 pacientes tiveram regressão do déficit, sendo que em 2 a regressão do déficit foi total. Quatro pacientes apresentavam hipostesia no território de V1 e V2. Todos apresentaram melhora desse sintoma. **Conclusão:** A TEACI é uma evolução comum em aneurismas intracavernosos gigantes, e está associada à melhora importante dos sintomas. No entanto pode ser catastrófica naqueles pacientes sem circulação colateral eficiente.

PALAVRAS-CHAVE: aneurisma intracavernoso, trombose carotídea, aterosclerose.

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Carotid cavernous aneurysms (CCA) represents approximately 3 to 5% of all intracranial aneurysms[1] and 15% of those originated in the internal carotid artery (ICA)[2]. CCAs can arise from any segment of cavernous carotid artery (Fig 1), but most commonly are originated in the horizontal segment, being projected forwardly and laterally, with the superior orbitary fissure and below the anterior clinoid process[3]. CCA morbidity and mortality indices are low[4-7], however, pain and neuro-ophthalmologic deficits due to neurovascular compression are frequent, what highlights the possibility of surgical treatment[7,8]. The vast majority of intracranial intradural aneurysms can be micro-surgically treated, commonly through aneurysmatic isolation without vascular occlusion, while CCA, when operated, frequently are through occlusion of ipsilateral ICA, with cerebral ischemia and amaurosis risks[9-11].

Spontaneous thrombosis of ICA is a complication in patients with CCA, most commonly associated with giant aneurysms[11-13], due to vascular compression. Most clinical investigators have found an increasing risk of thrombosis with an increasing degree of stenosis[14-19] and preceded by rupture of atherosclerotic plaques[20,21], but observations concerning plaque ulcerations[12-16] and intraplaque hemorrhages[22,23] have been more uncertain and controversial. The occlusion of ICA can be a dangerous complication to patients without a patent collateral circulation[24-27], manifested as an ischemic scenario, with a devastating cerebrovascular accident, or results in spontaneous therapeutic with a patent collateral circulation.

We report 5 cases of giant CCA developing spontaneous thrombosis of ICA.

METHOD

After approval from the Institutional Review Commission, the Discipline of Neurosurgery of Santa Casa Medical School of São Paulo studied patients with the diagnostic of cerebral aneurysms in the period between January 1989 and April 2007.

These patients were analyzed regarding genre, age, site and number of aneurysms, being selected for a second phase of the study those with CCA between (C3) lacerus segment and (C5) clinoid segment of ICA, selecting after that patients that evolved with spontaneous thrombosis of ICA. There were excluded from the study patients that presented aneurysms with partial or total intradural or subarachnoid colon, displasic aneurysms (beyond segment C4 of ICA) and traumatic or infectious aneurysms. All the selected patients were submitted to a full neurological exam and underwent radiological study with contrasted cranial computed tomography scan (CT), complete cerebral angiography (CAG) with and without subtraction and magnetic resonance imaging scan (MRI) with slices from the paraselar region after 90's.

The data from patients' files were completed afterwards during medical appointments. The following items were verified: age of diagnostic, age during treatment, genre, ethnic, morbid antecedents, site and size of aneurysm, presence of other aneurysms, neurological and visual signs and symptoms, therapeutic options and complications after treatment.

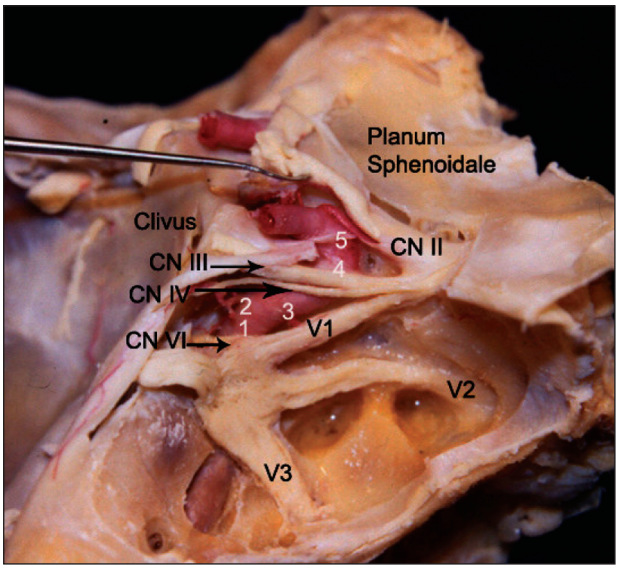

Fig 1. Cavernous sinus dissection, demonstrating cranial nerves: II-III-IV-V1-V2-V3-VI; [1] The posterior vertical segment; [2] The posterior bend; [3] The horizontal segment; [4] The anterior bend; [5] The anterior vertical segment (By Castro JAF, MD).

As to measure, according to Linskey et al.[27], pain symptoms and neurological deficits, the pain was graduated in severe, moderate, weak or absent, while neurological deficits were classified as severe, in the presence of cavernous sinus syndrome including trigeminal neuropathies; moderated, if there were complete involvement of III, IV and VI cranial nerves; weak, if there were deficits in one or two cranial nerves; and absent.

Each patient was classified taking into account his initial and final presentation during overcome: 0, absence of symptoms; 1, weak pain or neurological deficit; 2, moderate pain or neurological deficit; 3, severe pain or neurological deficit.

RESULTS

In our service during the period between January 1989 and April 2007 there were 711 consecutives patients with 802 aneurysms with and without surgical treatment during a period of 19 years. Symptoms and findings at presentation were recorded and compared with those at outcome (Table 1). We selected 35 patients with 40 carotid cavernous aneurysms (5%) of which 20 (50%) were giant aneurysms. Among those cases, 5 patients evolved with STICA (25%).

Average age at diagnosis was 62 years and 80% of patients were females. Clinical presentation was commonly related to atherosclerotic factors such as elevated blood pressure (80%), diabetes mellitus (40%) and dislipidemy (40%). All patients presented with hemicranial headache and retro bulbar pain. After STICA all presented improvement of symptoms. All patients had ophthalmparesy and

Table 1. Localization and treatment of cerebral aneurysms.

	N aneurysms	Observation	Clip	Ligature / Balloon	Embolization	By-pass	Trapping
Right end of ICA	12		12				
Left end of ICA	8		8				
Right ophthalmic	17	3	12	1	1		
Left ophthalmic	7	2	4		1		
Right CMA	125	18	106				
Left CMA	100	15	84	1			
Right ACA	8		8				
Left ACA	11	9	2				
Right PCoA	147	23	120	2	2		
Left PCoA	82	9	73				
Right intracavernous	20 (2.5%)	10		7	1	1	1
Left intracavernous	20 (2.5%)	11		4	4		1
Right Ant. choroidal	3	1	2				
ACoA	138	9	129				
Basilar	20	13	7				
Hypophyseal	2		2				
Right PCA	3		3				
Left PCA	6	2	2		2		
Others in right ICA	19	6	13				
Others in left ICA	23	8	13	1			1
Right PICA	14	1	13				
Left PICA	5	1	4				
Other	12	2	10				
Total	802	143	627	16	11	1	3

Ant: anterior; ICA: internal cerebral artery; CMA: cerebral medium artery; ACA: anterior cerebral artery; PCoA: posterior communicanting artery; PICA: posterior inferior cerebellar artery.

after STICA, 4 patients had regression of deficit, 2 partial and 2 complete. Four patients had sensorial trigeminal neuropathy in V1 and V2 territories, also showing improvement of symptoms after STICA.

Cases

Case 1 – Female 47-year-old patient with systemic arterial hypertension history presented with right acute pulsatile hemicranial headache and severe right retrobulbar pain followed by alterations on IIIrd, IVth, VIth, V1 and V2 cranial nerves (CN). Patient underwent cranial CT which showed a giant right ICA aneurysm. The patient was treated conservatively, and after six months of follow-up, presented with spontaneous resolution of headaches and retrobulbar pain. Afterwards underwent cerebral angiography with ICA occlusion. After an 11-year follow-up, patients evolved without pain, showing CN deficits and a MRI showing no vascular alterations at carotid cavernous.

Table 2. Intracavernous aneurysms' aspects.

Size	Total (n=40)	Observation (n=21)	Surgery (n=19)
Giant (>2.5 cm)	20	10	10
Large (1–2.5 cm)	13	8	5
Side			
Unilateral	30	13	17
Right	15	7	8
Left	15	6	9
Bilateral	10	8	2

Case 2 – Female 44-year-old patient with systemic arterial hypertension history presented with severe left hemicranial headache, retrobulbar pain, palpebral ptosis and alterations on IIIrd, IVth, VIth, V1 and V2 cranial nerves, with a diagnosis of a giant left ICA aneurysm. The patient underwent conservative treatment. After 3 months of follow-up, patient evolved with complete resolution of

headache and palpebral ptosis, maintaining the left VIth nerve deficit. Patient underwent cerebral angiography with left ICA occlusion. The posterior communicanting artery supplies the middle cerebral artery. During 6 years of follow-up the patient has not presented pain, and shows partial improvement of neurological deficits.

Case 3 – Female 65-year-old patient with systemic arterial hypertension, diabetes mellitus and dyslipidemic disease presented with acute right and severe hemicranial headache, right retrobulbar pain, and diplopy. Patient was diagnosed by MRI with a giant right CCA. With conservative treatment, the patient evolved with intensification of deficits and after 3 years started presenting palpebral ptosis and alterations on right IIIrd, IVth, VIth, V1 cranial nerves. Cerebral angiography showed giant intracavernous aneurysm and cavernous ICA occlusion of 70%. After 5 years of initial presentation patient there was spontaneous resolution of pain symptoms. By this time, cerebral angiography showed ICA thrombosis. After 7 years of follow up patient maintains CN deficits without pain.

Case 4 – Previously healthy 19-year-old male patient presented with left severe hemicranial headache with left retrobulbar pain and alterations on left IIIrd CN. CT showed aneurysmatic dilatation of left ICA. After 10 days of clinical observations patient presented ptosis's resolutions and underwent cerebral angiography, witch showed left ICA occlusion and carotid system irrigations by contralateral carotid and vertebral systems. After few months there were neurological deficits resolutions without other clinical manifestations.

Case 5 – Female 84-year-old patient with systemic arterial hypertension history presented with severe left hemicranial headache, left retrobulbar pain, and alterations on IIIrd, IVth, VIth, V1 and V2 cranial nerves. CT showed aneurysmatic dilatation of left ICA with local bone erosion (Fig 2A). MRI showed partial thrombosis of a giant aneurysm on left cavernous carotid artery (Fig 2B, 2C and 2D). Cerebral angiography showed left ICA obstruction (Fig 2E and 2F). After 3 months of clinical follow up patient evolved with resolution of pain symptoms and partial improvement of CN deficits, maintaining alterations in left IIIrd and VIth CN and semi-ptosis.

DISCUSSION

Using Pubmed data base there were identified only three studies with large series of CCAs, including one with natural history of 79 aneurysms with a follow up of 10 years[11] and other two studies which emphasized patients' neuro-ophthalmological prognostic: one with 40 CCAs for

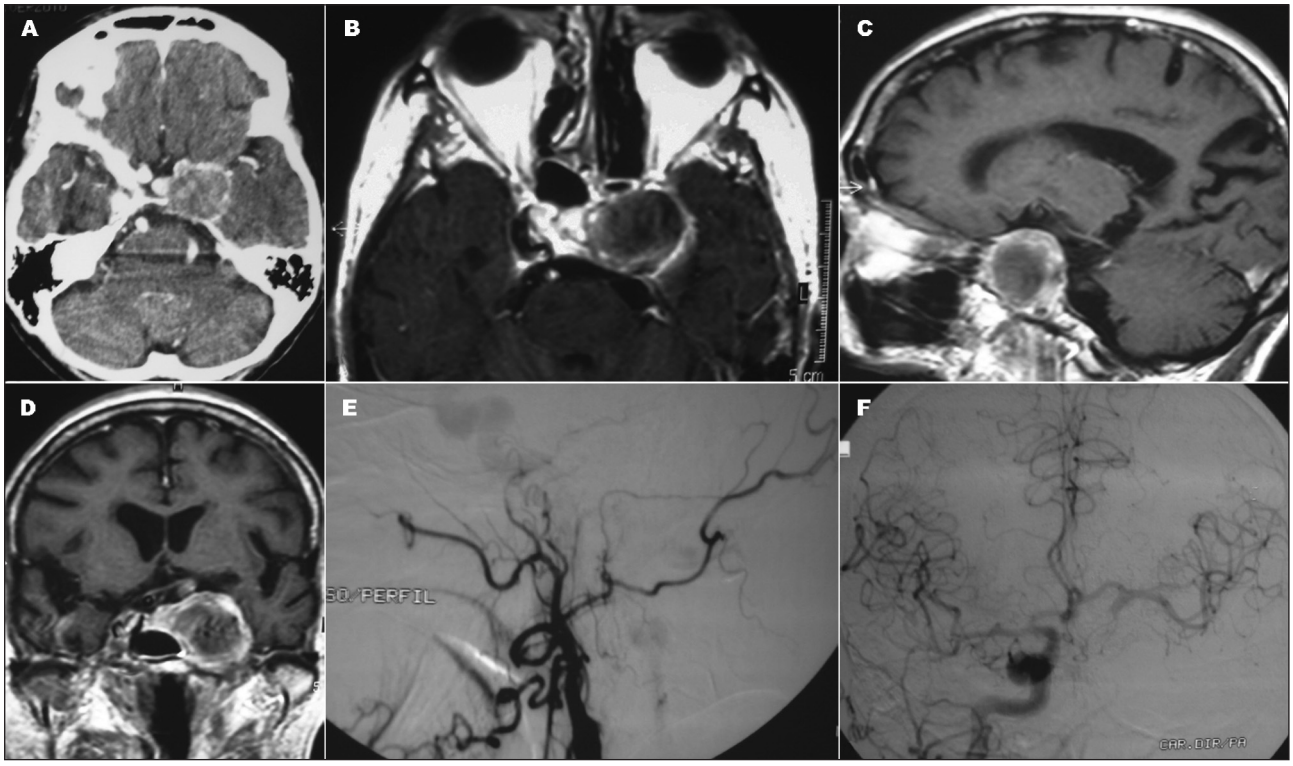

Fig 2. [A] CT scan showing giant aneurysm of left ICA, partially occluded by thrombosis and with bone erosion in a female 84-year-old patient with headache, retro bulbar pain, full III cranial nerve, left IV, VI, V1 and V2. [B, C and D] MRI scan showing important dilatation of a partially occluded aneurysm with thromboses. E and F: Angiography showing left ICA spontaneous thrombosis with a patent collateral circulation.

30 years[13] and other, a multi-centric study with 206 aneurysms with a follow up of 16 years[28]. There is no report of spontaneous thrombosis of ICA in patients with CCA.

As expected, common pathogenetics factors for the precipitation of thrombosis, as hypertension, diabetes and dyslipidemia could be demonstrated in 80% of our cases, without measurement of the degree of atherosclerosis.

The presence of bilateral cavernous aneurysms and intracranial aneurysms in other sites, not seen in this report, suggest that a degenerative process secondary to a genetic factor of fragility in the vascular wall should be present[2,3,8,9,10,13].

Rarely do CCAs suffer rupture with and subarachnoid hemorrhage by the time of diagnostic, due to the fact that cavernous sinus are composed by dural slices, which lay over the body of sphenoid and are, infrequently, projected towards the subarachnoid space[2,8]. We show one case of bone erosion (20%), without rupture or epistaxis.

CCAs frequently determine symptoms as consequence of neurovascular compression, being the most prevalent diplopia, as seen in all cases. This symptom follows VIth cranial nerve lesions due to its position within cavernous sinus, as shown in Fig 1[2,7]. The association with other nerves (IIIrd, IVth, V1 and V2), located in the lateral wall of cavernous sinus characterizes the complete cavernous sinus syndrome[6,8,13,28].

Considering our patients been submitted to conservative treatment, it could be verified a relapsing and fluctuating course of pain symptoms and a progressive impairment of cranial nerves' function, determining a poor neurological outcome till the spontaneous occlusion of ICA[27], when the pain scenario, determined by headache, hemifacial pain and retrobulbar pain, was improved or extinguished in all cases.

Patients submitted to interventionist treatment, i.e., endovascular treatment with coils, stent and ICA occlusion with balloon, as well as simple ICA ligature, ICA ligature associated with external carotid to media cerebral artery by-pass and ICA trapping, could be compared with our cases for having as final result, aneurysmatic or carotid exclusion from circulation.

The lack of a pre-occlusion assessment of collateral circulation should be avoided. Brain infarcts did not occur, but the high complications rates in the literature can be devastating[10,26].

In conclusion, CCA developing spontaneous thrombosis of ICA is a common outcome in giant carotid cavernous aneurysms and determines significant improvement of initial deficits. However, it can be catastrophic for those patients without efficient collateral circulation. Complete cerebral angiographic study, associated with internal carotid occlusion test with balloon is important, since it can indicate by-pass surgery for these patients, avoiding potentially dangerous outcome[29].

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