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A Case for Cerebral Thromboangiitis Obliterans

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SUMMARY The existence of cerebral thromboangiitis obliterans (CTAO) has been controversial. The clinical, laboratory and angiographic features of a young woman with recurrent thrombophlebitis, digital gangrene and a bilateral anterior opercular syndrome (Foix-Chavany-Marie) are reported. The cerebral angiogram demonstrated significant narrowing of fronto-opercular branches of both middle cerebral arteries. Histology of small digital muscular arteries revealed segmental adventitial fibrosis, narrowing or occlusion of lumen and mild lymphocytic infiltrates; occasional veins showed phlebitis. An etiologic relationship between cerebral occlusive disease and peripheral thromboangiitis obliterans (TAO) is suggested.

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IN 1879, Von Winiwarter¹ published an account of angiopathy in a 57-year-old man with foot gangrene; he named the angiopathy endarteritis obliterans. Buerger,² in 1908, designated the condition thromboangiitis obliterans (TAO). Spatz,³ and Spatz and Lindenberg,⁴ in 1939, published comprehensive accounts of the cerebral form of TAO (CTAO) distinguishing 2 types according to distribution of the lesions. There is a controversy in recent literature⁵⁻⁸ on whether TAO and CTAO are distinct clinicopathologic entities.

A young woman affected by recurrent thrombophlebitis, digital gangrene and a Foix-Chavany-Marie (biopercular) syndrome⁹ is reported as representative of the association between cerebral and peripheral TAO. These observations buttress the present trend in angiology to consider TAO and CTAO as distinctive but rare entities.

Patient History

MOC, a 33-year-old black woman, was admitted to the hospital on August 31, 1980, because of sudden inability to talk and swallow. She had been in good health until 1966 when she had a thrombophlebitis in her left leg. One year later she developed a similar episode. In 1968 she had a right deep vein thrombosis. In 1973, at the age of 26, she suddenly had left

hemiparesis. Radionuclide brain scan at the time showed increased uptake over the right parietal region consistent with an area of infarction. Bilateral carotid angiography revealed mild segmental narrowing of several of the left operculofrontal branches and an area of "luxury perfusion" over the right parietal frontal region. Routine blood tests and CSF examination were normal. Electrocardiogram and echocardiogram were normal. Electroencephalogram showed mild slowing over the left hemisphere. During the ensuing weeks she gradually recovered from her hemiparesis. During the next year she had numerous episodes of pain in her fingers and toes aggravated by cold weather and/or water. A selective left brachial angiogram revealed multiple occlusions and no opacification of the interdigital arteries (fig. 1A). A femoral angiogram was unremarkable down to the level of the ankle but failed to visualize any vessel beyond that area. Due to recurrent episodes of painful digital ischemia, she required a right transmetatarsal amputation of the left ring, right index finger, left fourth toe, and left big toe. Histologic examination of the amputated digits revealed segmental involvement of small muscular arteries several of which showed narrowing or occlusion of lumens (fig. 1B) mainly due to intimal hyperplasia and sparse mononuclear infiltration. No evidence of atheroma or calcification was seen in these vessels. None of these arteries showed fresh thromboses or vasculitis with fibrinoid necrosis. Occasional veins showed phlebitis. Focally involved vessels were surrounded by concentric layers of increased connective tissue.

In November, 1979 she had a bilateral lumbar sym-

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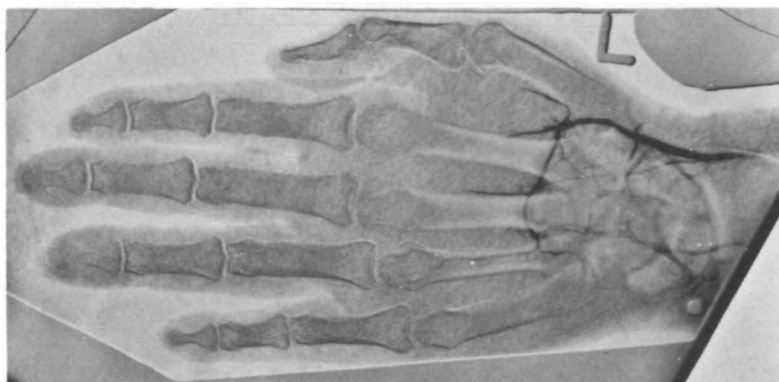


FIGURE 1A: *Left brachial arteriogram, with magnification views of the left hand demonstrates incomplete proximal palmar arch, extreme narrowing of the interosseus branches and no opacification of the interdigital arteries.*

pathectomy and inferior vena cava clipping because of recurrent thrombophlebitis.

Her past history revealed no evidence of lipid abnormalities, diabetes mellitus or migraine. She had smoked 10 cigarettes daily for many years. There was no history of intake of oral contraceptives, amphetamines or ergot derivatives.

On admission, she was unable to communicate orally, swallow or chew. Her mouth was half opened, and she had minimal voluntary control of her lips, tongue and jaw. There was bilateral facial paresis more severe on the the right side. The soft palate was immobile. No pathological reflexes other than bilateral hyperreflexia were found. Her neurovascular examination was normal. An awake electroencephalogram was reported as normal. The patient also had normal electrocardiogram, echocardiogram, and chest roentgenogram. Extensive blood and CSF tests were uninformative. Doppler flow velocity studies demonstrated decreased flow signals through most of the digital arteries and right posterior tibial artery. Radionuclide brain scan and CT scan showed an area of infarction in the left fronto-opercular area. On CT scan there also was a poorly marginated low density area in the right parieto-frontal region. Aor-

tocranial arteriography showed exaggerated tapering of the proximal segments of both middle cerebral artery branches, predominantly at the level of the frontal opercula (fig. 1C-D). The aortic arch and brachiocephalic vessels were unremarkable. Two months later she improved with increased strength in her facial and masticatory muscles, but swallowing difficulties remained essentially unchanged.

Discussion

The patient had recurrent thrombophlebitis before onset of neurologic dysfunction and digital gangrene. She also had numerous paroxysmal episodes of digital ischemia consistent with Raynaud's syndrome. She did not have evidence of cardiopulmonary, renal, hematologic, gastroenterologic, or collagen vascular disorders. Hyperlipoproteinemia with premature atherosclerosis, as well as intracranial infection and drug abuse, were excluded. There was no evidence of cardiac and extracardiac sources of emboli such as endocarditis, valvular heart disease, atrial myxoma, or paradoxical embolism arising from leg veins. Histologic examination of vessels from her amputated fingers and toes showed neither vasculitis, such as periarteritis nodosa, nor atherosclerotic plaques.

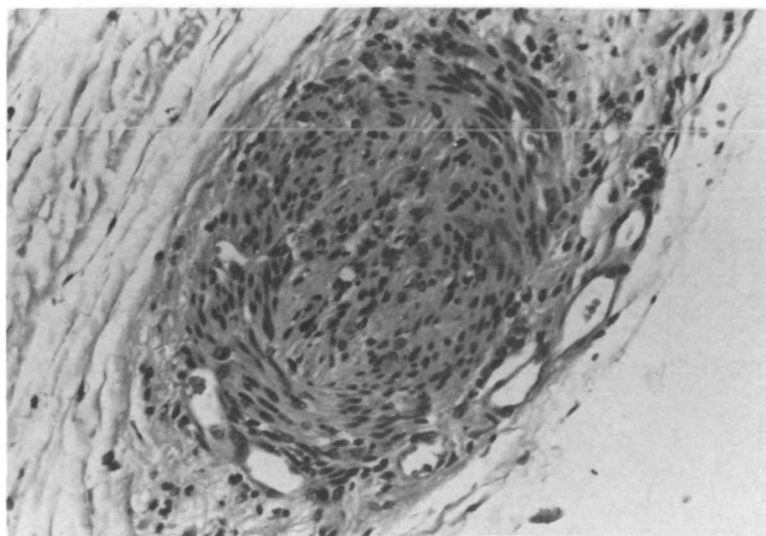


FIGURE 1B: *A slightly oblique section of a small muscular artery showing occlusion of lumen. Occasional lymphocytes are present in hyperplastic intima. H&E; Mag \times 280.*

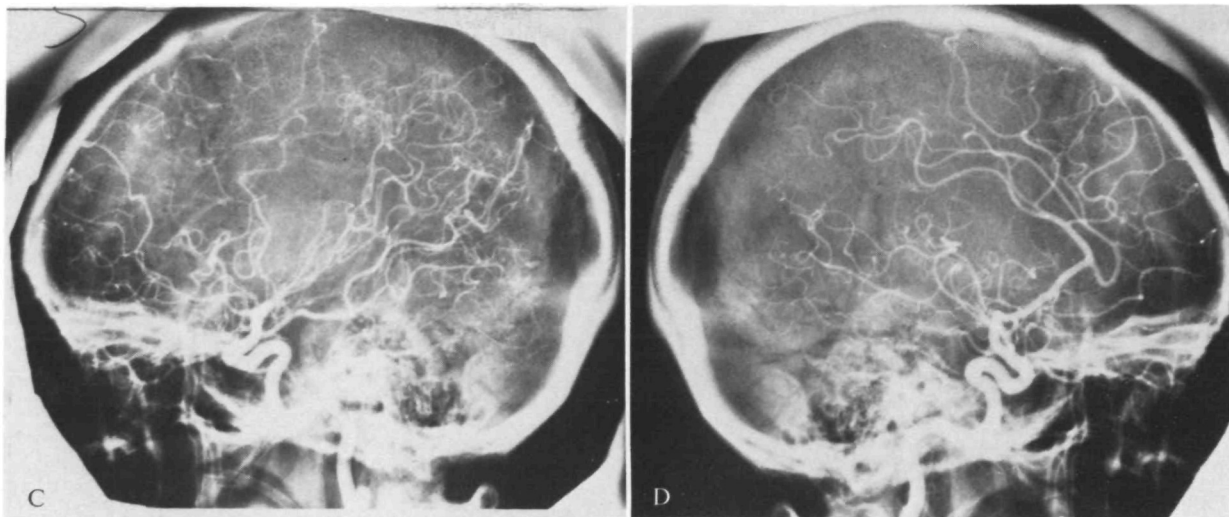


FIGURE 1C-D: Left and right carotid arteriogram demonstrating exaggerated tapering of middle cerebral artery branches in their proximal courses.

Although the microscopic appearance of the vessels did not suggest a distinct diagnosis, we consider this less important than the exclusion of vasculitis and atheromatous embolism or stenosis. Although it appears that in TAO the visceral arteries are somewhat more susceptible to atherosclerotic occlusions,¹⁰ the angiographic changes and the clinical course of our patient suggested TAO more than atherosclerotic disease. In light of her clinical course, angiographic appearances, and microscopic digital vascular changes, we believe that this patient's features were compatible with a diagnosis of TAO complicated by CTAO. The history of exposure to tobacco supports this diagnosis.¹¹

As this patient illustrates, the antemortem diagnosis of CTAO may depend heavily on the exclusion of other diseases that lead to digital gangrene and cerebral ischemic events. The incidence of CTAO in patients with TAO ranges between 0.5% and 18%.^{12, 13} Zülch¹⁴ found only 2 cases of CTAO among thousands of autopsies. In spite of the rarity of CTAO interest remains, as shown by publications emphasizing the selective aspects of CTAO^{5-6, 15-17} and TAO.¹⁸⁻²⁰ McLoughlin et al.²⁷ reported a greater prevalence of HLA-A9 and HLA-B5 antigens in patients with TAO when compared to patients with arteriosclerosis obliterans and normal subjects. Recent immunological studies²⁸ implicate a hypersensitivity reaction directed against arterial antigens as a cause of TAO. However, the basic question of whether TAO and CTAO are distinct clinico-pathological entities can not be answered until the pathogenesis of these diseases is clarified and specific pathological changes agreed upon. Strong clinical evidence and supportive pathologic data, as in the present patient, should continue to form the basis for considering the antemortem diagnosis of TAO and CTAO.

Acknowledgment

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Iatrogenic Carotid Cavernous Sinus Syndrome

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SUMMARY A hemodialysis shunt site, subclavian artery to internal jugular vein, resulted in a "pseudo" cavernous sinus syndrome. Recognition of this rare iatrogenic complication may assist in selecting other shunt sites and prevent potential visual loss and multiple surgical procedures.

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THE FULLY DEVELOPED clinical picture of a carotid cavernous sinus fistula (CCF) is not a diagnostic dilemma. These vascular malformations commonly are either traumatic or spontaneous.¹ This report is of a iatrogenic instance with clinical signs and symptoms of a carotid cavernous sinus syndrome following a subclavian artery to internal jugular vein shunt.

History

A 62-year-old right-handed man had received 6 years of hemodialysis treatment for membranous glomerulonephritis. Multiple episodes of thrombophlebitis eventually consumed all common sites for peripheral hemodialysis fistulas. Renal transplantation was unsuccessful. Since all peripheral shunt sites had failed, a left subclavian artery to left internal jugular vein shunt was selected. Within three weeks after the anastomosis, he experienced holocephalic headaches and mild generalized weakness, worse on his right side. A slow but progressive reddening of his left eye was noted. He denied diplopia, subjective bruit, pain and decreased visual acuity.

Neurological examination documented a mild but distinct pronation drift of the right arm without tendon reflex asymmetry. Ophthalmologic examination revealed a marked dilation and arterIALIZATION of the left conjunctival vessels and 2 mm of left proptosis

(fig. 1). An ocular bruit was not heard and both globes were neither tender nor pulsating (utilizing a Schiøtz tonometer). Visual acuity was correctable to 20/25 on the right and 20/20 on the left. Pupils were 4 mm bilaterally and equally responsive to light and accommodation. Applanation tonometry readings were 12 mm Hg on the right and 10 mm Hg on the left. Vergence, version and duction extraocular movements were normal. Fundus examination showed mild dilation of the left retinal veins.

Abnormal laboratory studies included: BUN 64 mg/dl, creatinine 8.5 mg/dl, total protein 5.4 gm/dl, albumin 2.9 gm/dl, prothrombin time 25.7 seconds (control 11.8) and partial thromboplastin time 67.3 seconds (normal less than 40 seconds). Urinalysis revealed a specific gravity of 1.017, 2+ glucose, 3+ protein, 2-6 wbc/hpf, and 1-3 casts/lpf.

An EEG showed slowing and decreased amplitude over the left frontal area. The CT scan suggested a left frontal chronic subdural hematoma. Within a week after the subdural was evacuated the right sided weakness and the EEG improved markedly.

A left carotid arteriogram, after removal of the subdural hematoma, showed slow arterial filling of the left cerebral hemisphere with all venous drainage from that side via the right transverse sinus and right internal jugular vein. A left subclavian arteriogram documented left cavernous sinus arterIALIZATION from retrograde flow in the left internal jugular vein, transverse sinus and petrosal sinus. Blood flow was from the shunt into the left carotid cavernous sinus (fig. 2). Since venous drainage from the left cerebral hemisphere occurred through the right internal jugular vein, the left internal jugular vein was ligated above the fistula. Two months later the patient was symptom

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