Short Communication

Angiographic Findings in a Young Man With Recurrent Stroke and Positive Fluorescent Treponemal Antibody (FTA)

MICHAEL S. ALDRICH, M.D., JAMES M. BURKE, M.D., AND SURENDRA M. GULATI, M.D.

SUMMARY A young man with a history of two strokes was found to have serologic evidence of a prior syphilitic infection, abnormal spinal fluid, and angiographic evidence of diffuse intracranial and extracranial angiopathy which we believe was due to tertiary syphilis, although his use of oral amphetamines cannot be ruled out as a contributing factor. This is the first reported case in which angiography has demonstrated both intracranial and extracranial disease arterial disease due to neurosyphilis. Since syphilis can produce diffuse cerebral arterial changes, this diagnosis should be considered in young patients with stroke even when non-treponemal serologic tests are negative.

NEUROSYMPHILIS (NS) remains an important and treatable cause of neurologic disease, even though its more familiar forms have become less common since the introduction of antibiotics. Several studies provide evidence that the relative frequency of vascular and other unusual forms of NS may be increasing, but angiographic findings in vascular NS have rarely been reported and these findings have been limited to intracranial vessels. In this paper, we will discuss the case of a young man with two strokes presumably resulting from NS and whose angiogram demonstrates diffuse intracranial and extracranial occlusive disease.

Case Report

A 25 year old right handed white male was admitted to the Ann Arbor Veterans' Administration Hospital in April 1982 complaining of speech difficulty and right-sided weakness. He had been well until October 1978 when he had the first of several transient episodes of left arm and leg weakness. When he was first admitted, in June 1979, physical examination revealed bilateral carotid bruits, mild left lower facial weakness, mild left hemiparesis with hyperreflexia, and a left Babinski sign. Complete blood count, sedimentation rate, triglycerides, lipoprotein electrophoresis, and electrocardiogram were normal. Spinal fluid glucose was 60 and protein was 42, but CSF VDRL was not done. A cerebral arteriogram demonstrated occlusion of the right internal carotid artery two centimeters above the common carotid bifurcation and severe stenosis of the left vertebral artery with narrowing of the left subclavian artery.

Following angiography he underwent right superficial temporal artery to the right middle cerebral artery bypass surgery. Biopsy of the right superficial temporal artery showed mild intimal thickening without evidence of granulomas.

He had no further symptoms until January 1981 when he experienced several complex partial seizures with secondary generalization which were treated with phenytoin. In June 1981, while hospitalized with mononucleosis, he complained of progressive decreased fine motor function of his right hand and on neurologic exam rapid alternating movements of his right hand were impaired. A right Babinski sign was present but reflexes were symmetric. Spinal fluid glucose was 57, protein was 60, there was one white blood cell and CSF VDRL was negative.

He had no additional neurologic complaints until April 1982 when, after a night of heavy alcohol and marijuana use, he awoke with right arm and leg weakness and speech difficulty. By the time of admission, three weeks later, his symptoms had gradually improved.

He revealed a history of venereal infection acquired in the Philippines several years earlier, and treated with an oral antibiotic. He used marijuana and alcohol frequently, as well as amphetamines and mescaline on weekends, but denied intravenous use of drugs. He had smoked two or three packs of cigarettes per day for about eight years. He took phenytoin irregularly but no other prescribed medications. There was a family history of myocardial infarction and hypertension.

The general physical examination was normal except for a loud left carotid bruit and bilateral femoral bruises, and there were no signs of intravenous drug abuse. Neurologic examination demonstrated a mild right lower facial weakness and a right hemiparesis with right sided hyperreflexia. A left Babinski sign was present. Speech was slow with a spastic dysarthria. Sensory examination was normal.

Chest radiography was normal. An electrocardiogram showed first degree atrioventricular block but two-dimensional echocardiography was normal. The
following blood studies were normal or negative: complete blood and platelet count, electrolytes, liver enzymes, blood urea nitrogen, prothrombin and partial thromboplatin times, antinuclear antibody, rheumatoid factor, hepatitis B surface antigen, serum protein and lipoprotein electrophoresis, C3, C4, CH50, cryoglobulins, cold agglutinins, antithrombin III, Raji cell assay, serum viscosity, leptospira antibodies, febrile agglutinins, and urinary homocysteine. Westergren sedimentation rate was two. Serum fluorescent treponemal antibody was positive but VDRL was negative. Spinal fluid glucose was 60, protein was 52, IgG was 6 mg/dl, and VDRL was negative. CSF electrophoresis showed a minimonoclonal band in the IgG portion which was also present in serum. Head CT scan showed evidence of loss of brain substance in the right parietal and left frontal lobes (fig. 1).

Aortic arch and four vessel cerebral angiography demonstrated a number of abnormalities. There was a patent surgical anastomosis between the right superficial temporal and middle cerebral arteries (fig. 2). The right internal carotid artery was occluded from its origin to the supraclinoid portion (fig. 2, 3). Some terminal branches of the right posterior cerebral artery appeared occluded, with collateral flow from right middle cerebral artery branches. The proximal portions of the right middle cerebral artery and the right anterior cerebral artery were severely narrowed.

There was irregular narrowing of the left internal carotid artery at the level of the carotid canal and at the junction of the cavernous and precavernous segments (fig. 4). Most of the left middle cerebral artery branches were occluded, with several large branches in the region of the basal ganglia serving as collaterals. Both anterior cerebral arteries filled from the left internal carotid artery. The left subclavian artery was severely stenotic proximal and distal to the left vertebral artery (fig. 5). The left vertebral artery appeared nearly occluded at its origin, and was probably entirely occluded in the high cervical region; the left posterior
cerebellar artery was never seen with certainty. The right vertebral artery was small and possibly occluded distal to the origin of the right posterior cerebellar artery. The basilar artery appeared occluded between its origin and the superior cerebellar arteries.

A diagnosis of probable neurosyphilis was made and the patient received a 14 day course of high dose intravenous penicillin. Four months later, his rehabilitation continued without progression of his neurologic disease.

Discussion

Vascular NS was a relatively uncommon condition in the preantibiotic era, and accounted for just 5% of 2019 patients with symptomatic neurosyphilis in Kierland's series. However, the clinical presentations appear to have changed since the introduction of antibiotic treatment and vascular and other atypical presentations now account for from 13% to 50% of cases of neurosyphilis. Heubner’s endarteritis is the most common pathologic finding in vascular neurosyphilis. Adventitial lymphocytic infiltrates and fibrous intimal proliferation can lead to occlusion of several types of arteries including medium caliber intracranial arteries, meningeal branches, and large arteries at the base of the brain.

Only a few of the reported cases of vascular neurosyphilis have included angiographic findings, perhaps because angiography was rarely used in the era when NS was widespread. Of the nine studies we have reviewed which did report angiography, only one reported more than one case. Aupy et al described six patients with NS; in four of them angiography demonstrated occlusions of the left or right middle cerebral artery (MCA) or posterior cerebral artery.

Each of the following five reports concerns a single case. Vatz et al reported a 42 year old man with NS who presented with a left hemisphere stroke. In this patient, angiography demonstrated narrowing of the supraclinoid portion of both internal carotid arteries, with stenosis or occlusion of the right anterior cerebral artery (ACA), and branches of the left MCA and left ACA. Liebeskind et al reported a 65 year old woman with NS; angiography demonstrated diffuse irregularity and “beading” of the ACA and MCA bilaterally. Mondrup et al reported a 17 year old patient with meningovascular NS whose angiogram demonstrated multiple stenoses and occlusions involving the internal carotid arteries, ACA’s, and MCA’s. Reignier et al described the case of a 21 year old woman with paresthesias and right leg weakness; angiography demonstrated segmental changes of anterior cerebral vessels. Suchenwirth reported a 28 year old man with NS who died several days after the sudden onset of right hemiplegia. Angiography showed occlusion of the left internal carotid artery and narrowing of the right internal carotid and right ACA.

Three case reports have described patients with NS and local angiographic abnormalities. Brain gummas were associated with focal hypervascularity in one patient, and irregular narrowing of both ACA’s in the area of the gumma in the other. Angiography in the third patient disclosed a localized blush in the left parietal area; biopsy of this region revealed chronic perivasculitis.

Our patient not only demonstrated angiographic findings similar to those of most of the patients mentioned above, but also showed evidence of extracranial disease. Although a number of extracranial angiographic changes have been attributed to tertiary syphilis (including dilation and calcification of the thoracic aorta, occlusion of the coronary ostia, aneurysm of the sinus of Valsalva, retinal periarteritis, and popliteal artery aneurysms), we have found no reported cases in which angiography has demonstrated both intracranial and extracranial arterial disease due to neurosyphilis.

In the differential diagnosis of this case, only a few of the wide spectrum of vasculitides involving cerebral vessels are diagnostic considerations in this case. Our
FIGURE 5. The left subclavian artery is markedly narrowed proximal and distal to the vertebral artery. The left vertebral artery appears nearly occluded at its origin and is probably entirely occluded in the cervical region.

patient's laboratory studies make inflammatory diseases such as periarteritis nodosa, systemic lupus erythematosus, Takayasu's arteritis and giant cell arteritis unlikely. The CSF findings rule out bacterial or tuberculous meningitis. Cerebral angiographic changes associated with drug abuse include irregular narrowing with "beading" of large and intermediate size vessels, and complete or partial occlusion of vessels less than one millimeter in diameter. 22 Although our patient did not have a history of parenteral drug abuse, he had used drugs orally, and this use cannot be ruled out as a contributing factor in his angiopathy. This case shows some features of moyamoya disease but extracranial findings are not typical. Accelerated atherosclerosis secondary to unknown factors cannot be completely ruled out although the pattern of arterial involvement is unusual because of the occlusion of multiple small vessels without involvement of arterial bifurcations. Syphilitic arteritis, which can produce diffuse arterial changes, appears the most likely diagnosis and is strongly supported by the serologic evidence of a prior syphilitic infection. The negative serum and CSF VDRL with mild CSF abnormalities are not uncommon findings in neurosyphilis. 1, 23

References
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