Syphilitic arteritis involving the origin of the cervical internal carotid artery

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ABSTRACT

في هذا التقرير نعرض حالة مرض الزهري أصابت الأوعية الدموية و أغشية المخ لامرأة شابة بالغة تعاني من شلل نصفي أيسر وذلك بسبب قرب انسداد الشريان السباتي و حدوث جلطة غير حادة عبر الشريان المخي الأوسط على أساس النتائج الإيجابية للدم والسائل الشوكي لمرض الزهري، و دوبلر الشريان السباتي، وتصوير الأوعية بالرنين المغناطيسي، فضلا عن التحسن بعد علاج البنسلين. في هذا التقرير نعرض حالة نادرة الحدوث من حيث إصابة أصل الشريان السباتي بالزهري ونتائج التصوير التي وصفت وكذلك المراجع المماثلة.

We report a case of meningovascular syphilis in a young adult woman presenting with left hemiparesis due to near occlusion of proximal cervical internal carotid with subacute middle cerebral artery territory infarction. Diagnosis was made on the basis of positive serum, and spinal fluid serology for syphilis, carotid Doppler, and magnetic resonance angiography, as well as improvement after intravenous penicillin therapy. In this case report, the imaging findings were described and related literature was reviewed.

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Syphilis is caused by the spirochete *Treponema pallidum*, and remains an important and frequently encountered sexually transmitted disease. If untreated, it progresses, through primary, secondary, and tertiary stages. In around 30% of untreated patients late disease of the heart, CNS, or other organs develop. The incidence of syphilis decreased significantly after the penicillin era in 1940. Unfortunately, the prevalence again rose dramatically during the recent acquired immunodeficiency syndrome (AIDS) era worldwide.[1] The infectious course of syphilis has 3 stages. The primary stage is characterized by the presence of a chancre, a round painless sore or ulceration at the inoculation site, and regional adenopathy. If untreated, within 2-6 weeks, hematologic dissemination and the secondary form of syphilis occurs. Clinically, this manifests as a rash on the palms and soles, and may develop characteristic oral mucous patches or other lesions in moist areas of the body called condyloma lata. Tertiary syphilis, occurs after resolution of the secondary stage, usually 5 years or more after the primary infection, and involves many organ systems, including the CNS, cardiovascular system, bones, joints, skin, and mucous membranes.[2] Early neurosyphilis occurs within weeks to months to the first few years of infection, and coexists with primary or secondary syphilis. The clinical syndromes include asymptomatic or symptomatic meningitis, cranial nerve palsies, and meningovascular syphilis. In contrast, late or tertiary neurosyphilis occurs years to decades after the initial infection, and includes parenchymatous, and meningovascular syphilis. Late parenchymatous neurosyphilis syndromes are rare nowadays and include general paresis, a rapidly progressive dementing illness due to syphilitic encephalitis, tabes dorsalis affecting the dorsal roots and posterior column of the spinal cord presenting as sensory ataxia with incontinence, pain, and optic atrophy.[3] Meningovascular neurosyphilis, may appear any time from months to a decade after the primary infection and presents as stroke syndrome with focal neurologic deficits, or subacute illness with global CNS dysfunction.[4] Our objective in presenting this case was to highlight the importance of considering syphilis as a remerging cause of carotid arteritis resulting in stroke in the non-atherosclerotic cohort of patients.

Case Report. A 44-year-old Indian women, working in Kuwait, married with 2 children, presented to the emergency room of Al-Sabah Hospital, Kuwait with weakness of the left upper and lower limbs, and dropped left angle of the mouth of one day duration. There was

no fever, fits, or disturbed level of consciousness. She is a non-smoker nor drug abuser. Her history was remarkable for hypertension, and diabetes controlled by medical treatment, with no history of cardiac or peripheral vascular disease. On examination she had blood pressure of 118/76, regular pulse 92 beats/min, no fever, conscious, with oxygen saturation 97%. Clinical examination of chest, heart, and abdomen were unremarkable, and no carotid bruit detected. The CNS examination showed left upper and lower limb weakness, with power 2/5, decreased muscle tone, decreased sensation on the left side was for all modalities, left upper motor neuron facial nerve palsy. It was difficult to elicit deep tendon reflexes, and there were no signs of meningeal irritation. Routine blood tests were unremarkable with low density lipoprotein-cholesterol 2.1 mmol/L, high density lipoprotein-cholesterol 1.32 mmol/L, total cholesterol 3.5 mmol/L, triglycerides 1.62 mmol/L, fasting blood glucose 5.2 mmol/L, creatinine 65µmol/L (normal up to 82 µmol/L), and normal electrolytes. Urgent CT showed right temporal-parietal recent infarct (**Figure 1**), and left occlusive cerebrovascular accident was diagnosed. Carotid Doppler showed diffuse mural thickening with severe concentric narrowing at the carotid bifurcation, with around 90% diameter stenosis (**Figure 2**), peak systolic/diastolic velocity 490/170 cm/sec, with no flow detectable in the internal carotid artery (ICA), more distal, indicative of near total occlusion. An MRI of the brain, including MRI angiography (MRA) of neck arteries and circle of Willis, confirmed the recent right cerebral infarct, and showed occlusion of the origin and proximal segment of right cervical ICA (**Figure 3**). Other cervical arteries and more distal intracranial circle of Willis were unremarkable. Transesophageal echo did not show any intracardiac thrombi. Vasculitis, including syphilitic was considered, erythrocyte sedimentation rate

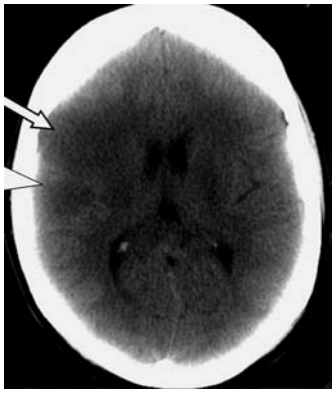

Figure 1 - Axial non contrast CT brain on day of admission, showing right hemispheric recent infarct, with cortical, and sub cortical ill defined hypodensity and brain swelling, loss of grey white matter interface and effacement of right insula.

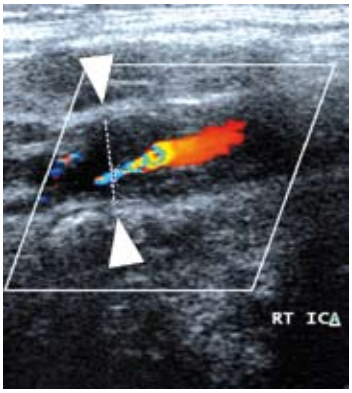

Figure 2 - Color Doppler, showing diffuse circumferential narrowing of the proximal right internal carotid artery, with around 90% diameter stenosis, color aliasing, and absent flow signal more distally.

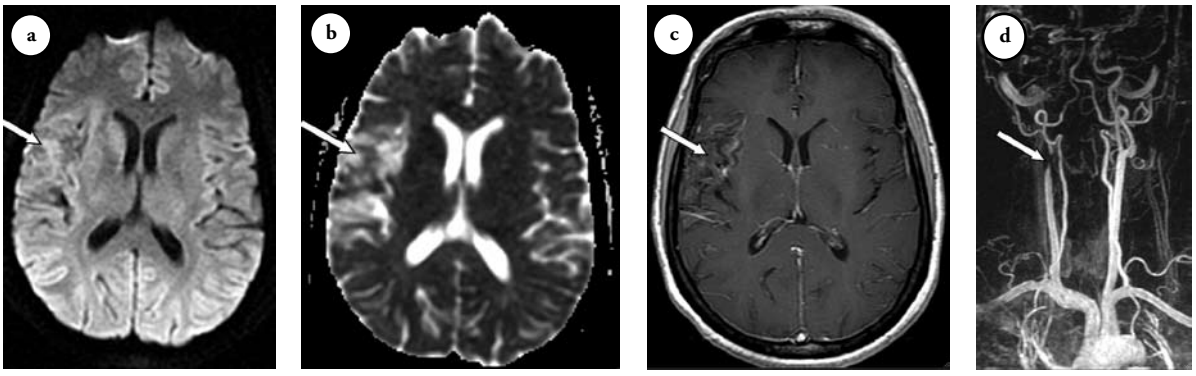

Figure 3 - Brain MRI on day 3 after admission showing **a)** axial diffusion weighted at b=1000 sec/mm^2, **b)** corresponding apparent diffusion coefficient (ADC) map, **c)** corresponding axial T1 post intravenous gadolinium showing bright signal of the right temporoparietal region on b=1000 and ADC map, and gyral enhancement post intravenous gadolinium confirming right hemispheric subacute infarct, and **d)** magnetic resonance angiography of neck vessels showing absent signal of right internal carotid artery signifying occlusion, or near total occlusion of the proximal right internal carotid artery.

was raised 102mm/1st hr. Serology for syphilis including Venereal Disease Research Laboratory (VDRL), and Treponema pallidum hemagglutination test (TPHA) were strongly positive, VDRL and TPHA for CSF were also positive. The CSF also showed white blood cell (WBC) count of 23 cells/μL, 90% of WBCs were lymphocytes with red blood cells 2 cells/μL, and protein level was 650mg/L. Serology tests for vasculitis were negative. Accordingly, the diagnosis of neurosyphilis with Huebner arteritis was made. Testing for HIV to exclude possible association with neurosyphilis was negative. She was treated by benzyl penicillin 4 million units/4 hourly for 3 weeks (European guidelines),[5] and physiotherapy, with improvement in her clinical condition regarding power of left upper limb, which improved to 4/5, and power of left lower limb improved to 3/5. Lumbar puncture was repeated after 6 months and showed decrease of WBCs count to 3 cells/μL and drop of protein to 420mg/L. Carotid endarterectomy for our patient, following the general guidelines for treating symptomatic atherosclerotic carotid artery stenosis, was considered.[6] However, as she showed continuous clinical improvement, and her condition remained stable up to 6 months follow up, the procedure was not undertaken.

Discussion. Meningovascular syphilis causes both an arteritis and meningitis; clinically, it may manifest as an acute or subacute stroke syndrome or, more commonly, as a subacute illness, with a prodrome of weeks to months. Symptoms include headache, vertigo, seizures, transient hemiplegia, insomnia, and psychological disturbances.[2] Syphilitic arteritis is of 2 forms; Huebner arteritis, which is the more common form, affects the medium and large arteries, and is characterized by fibroblastic proliferation of the intima, thinning of the media, and adventitial fibrous and inflammatory changes. The second is the Nissl-Alzheimer form of arteritis, which affects the small arteries and arterioles and is manifested as endothelial and adventitial thickening. Both types of arteritis produce narrowing and focal dilatation of the vessel lumen, correlating with the segmental "beading" seen angiographically. Vascular occlusion results in ischemia and subsequent infarction.[3] A wide range of imaging findings can be seen in meningovascular syphilis, and many cases have no imaging abnormalities.[7] The CT and MR findings of the arteritis form of meningovascular syphilis consist of infarcts affecting both gray and white matter in single or multiple arterial distributions, meningitis can be detected as meningeal thickening with enhancement. The demonstration of multicentric process leads to the consideration of vasculitis, and includes an autoimmune vasculitis such as lupus erythematous or polyarteritis nodosa, infectious basilar meningitis, and amphetamine induced vasculitis.[8] An MRA of cervical, and cranial arteries, may demonstrate concentric narrowing of large-and medium caliber arteries, and irregularities of smaller vessels. Moyamoya appearance is rarely reported, infarct enhancement, and overlying cortical vascular blush have been described.[9]

In our present case, the diagnosis of meningovascular syphilis was made in a 44-year-old woman presenting with subacute stroke syndrome, with left hemiparesis, on the basis of positive serum and spinal fluid serology, suggestive carotid Doppler, and MRA findings, and response to antibiotic therapy. The MRA showed smooth concentric narrowing of the carotid bifurcation, and near total occlusion of the origin and proximal cervical right ICA. These Doppler, and MRA findings are different from those associated with atherosclerotic vascular disease and correlate well with the usual pathologic features of meningovascular syphilis.[3,7,10] Holland et al[11] reported 3 cases of meningovascular syphilis in which the angiographic findings showing multiple vessel involvement with segmental constriction and occlusion, which included supraclinoid ICA, proximal M1, and A1 segments of middle cerebral artery (MCA), and anterior cerebral artery (ACA), basilar artery, posterior cerebral, and superior cerebellar arteries. Brightbill et al,[7] in a series of 35 patients of documented neurosyphilis, reported 8 cases of meningovascular syphilis (23%), presenting with cerebral infarctions involving the cortex and subcortical white matter of different vascular territories, as well as pontine, thalamic, and ganglio basilar infarcts, 2 of the 4 cases who underwent cerebral angiography demonstrated high grade concentric narrowing of intracranial supraclinoid ICA, and basilar artery. Tien et al,[9] in a report of 6 patients with neurosyphilis in HIV carriers, 5 presented with acute or subacute stroke, 4 of these involved bilateral basal ganglia corresponding to the vascular territories of the lenticulostriate arteries, and one case involved the left MCA. Asdaghi et al[12] reported 2 cases of meningovascular syphilis presenting with infarcts of bilateral caudate, and anterior limbs of internal capsule signifying bilateral recurrent artery of Heubner territory, and another case with bilateral narrowing of the M1 segment of the MCA presenting with lacunar infarcts of right midbrain crus, left thalamus, and left pons. Umashankar et al[13] reported a case of acute infarction in the medial posterior inferior cerebellar artery territories bilaterally. Our case reports near occlusion of the origin of cervical ICA due to meningovascular syphilis presenting with subacute MCA territory infarction.

In conclusion, neurosyphilis is difficult to diagnose because most patients with the disease are asymptomatic. However, the clinical picture of cerebral infarction, particularly in a young adult patient, should generate investigation into possible causes of vasculitis and the diagnosis of meningovascular syphilis, particularly in those who are HIV-positive should be considered. Meningovascular syphilis is typically shown as single or

multifocal infarcts with evidence of segmental narrowing or occlusion of small or large intracranial or extra cranial carotid or vertebrobasilar circulation, and is confirmed by positive serology and CSF for syphilis. The diagnosis of meningovascular syphilis prompts important antibiotic therapy, which improves patient outcome.

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