Stroke in the young - angiitis aortic arch following varicella zoster infection

Dr. Hassan Ranganath, M.D. and Dr. Shahid Bartas, FRCPE,
Dr. Muhammad Mubarak, Facharzt

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Chickenpox in adults usually runs a severe and protracted course and can be occasionally associated with fatal complications. In children, however, it is often a trivial, febrile illness.

We report an adult who developed cerebral infarction and stroke following 2 weeks of chickenpox infection involving arch of aorta and its main tributaries. Early initiation of high dose prednisolone therapy resulted in neurological improvement and partial regression of arthritis.

To the best of our knowledge, this is the first case reported from the Middle East of angiitis of aortic arch in a patient with chickenpox, leading to stroke.

Case History. A thirty-year old Somalian was admitted with headache, confusion, fever and on-and-off vomiting of four days' duration. This progressed to drowsiness, inability to talk, painful swallowing and right-sided body weakness. Seven days prior to admission there was eruption of a generalized body rash. Past medical history was insignificant.

General physical examination revealed an ill-looking young man, who was confused, agitated, drowsy and dehydrated. There was extensive vesiculopapular rash in different stages of development, especially involving the chest and abdominal wall. Mouth was full of vesicles and ulcerations. Neurological examination showed conscious patient with global aphasia, dense hypotonic areflexic right hemiplegia.

Following a 10 day stay, in-hospital routine examination showed no radial, brachial and carotid pulsations on either side, with good pulsation at femoral, popliteal and dorsalis pedis arteries. Blood pressure in the lower limbs was 120/70.

Laboratory workup, which included complete blood count, urea, creatinine, electrolytes, blood sugar, liver function tests, electrocardiography, chest X-rays and arterial blood gases, were all normal. Blood ESR - 16, Blood culture, HIV serology, anti DNA double stranded antibodies, VDRL were all negative. ANCA not done.

CSF analysis was normal. Color doppler ultrasound could not detect any blood flow in both common carotids and arteries of upper limbs. Computerized axial tomographic (CAT) brain scan (Fig. 1) revealed cerebral infarction in the territory of left middle cerebral artery with brain oedema. The temporal artery biopsy was carried out but was inconclusive.

Aortic arch angiography showed complete occlusion of innominate, common carotids and subclavian arteries at site of their origin (Fig. 2). The patient was treated with acyclovir and steroids. His general condition improved. The chickenpox rash became dry and crusted.

Four weeks later, he started to improve and a repeat doppler examination showed a partial resumption of blood flow in carotids and brachial arteries. Eight weeks later, he was able to walk with the help of a zimmer. He was discharged after 8 weeks on prednisolone 10 mg/day, and maintained his progress on follow-up visits. Repeat angiography was not carried out in view of clinical recovery.
Discussion Central nervous system involvement in chickenpox has been well described and is due to cerebral vasculitis.

In our patient, clinical and angiographic features were highly suggestive of arteritis involving the arch of the aorta with its main tributaries. Arch of the aorta has been reported to be involved in various types of vasculitides and the most common being the Takayasu’s arteritis.

Medium and small size cerebral arteries have been involved in various systemic disorders which include polyarteritis nodosa and systemic lupus erythematosis, Becher’s disease, Wegner’s granulomatosis, temporal arteritis, etc. but required meningeal and brain biopsy which was beyond our scope here. Our patient had none of the clinical or laboratory features of these diseases. Granulomatous angiitis of the central nervous system has been reported in association with chickenpox and herpes zoster ophthalmicus.\textsuperscript{3,4,5} The varicella zoster virus has been implicated as the inciting agent. The viral nucleocapsids and antigens have been found in the walls of the affected arteries.\textsuperscript{6} Veins and arteries of various size including large cerebral arteries, may be involved.

Granulomatous angiitis is the likely diagnosis in our patient due to concomitant chickenpox infection. Unfortunately biopsy did not favor our
diagnosis. Aortic arch angiography confirmed the site of involvement and occlusion. The disease is usually progressive and fatal and response to various treatment modalities is unsatisfactory. Immunosuppressive therapy may be of help if initiated early in the course of the disease. Our patient received high dose prednisolone (60 mg/day) from the beginning and showed considerable clinical improvement along with the return of some blood flow in occluded vessels indicating partial regression of arteritis.

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References