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AN ACUTE VASCULITIS RESEMBLING POLYARTERITIS NODOSA*

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Peripheral vascular diseases are structural and functional abnormalities of the peripheral blood vessels that produce blood flow irregularities. The signs and symptoms relate either to ischemia or inflammation of the involved vessels, or to both. A previously healthy 4.5-year-old girl was referred to our hospital with bruises on fingers and toes. She had no history of environmental causes, including drug intake. Symptoms of abdominal pain, fever, and a swollen face preceded the symptoms of her extremities. On physical examination, her blood pressure, which could be obtained only on the thighs, was high. There were no pulses on the upper extremities or on either the a. dorsalis pedis or the a. tibialis anterior dextra. There was massive necrosis on fingers which led to dry gangrene. A rise in acute phase reactants accompanied the physical findings, and a segmental obstruction was found proximally to a. brachialas, and distally to a. radialis dextra and a. dorsalis pedis dextra on digital subtraction angiography (DSA). High-dose methylprednisolone, cyclophosphamide, salicylates and dipyridamole were given; as these medications did not relieve the symptoms, a thoracal sympathatectomy was performed. The peripheral circulation improved, but a demarcation zone developed on the fingertips, leading to amputation.

This patient is presented because she had an acute vascular spasm, of unknown etiology, resembling polyarteritis nodosa. *Key words: inflammatory vasculitides, polyarteritis nodosa, etiology, therapy.*

Inflammatory vasculitides are syndromes in which the various patterns depend on the size and location of the affected vessels. Vasculitis may be a primary disease or may be secondary to connective tissue disorder, infection, or other processes[1]. Peripheral vascular diseases are structural and functional abnormalities of the peripheral blood vessels that produce blood flow irregularities. The signs and symptoms relate to ischemia or inflammation of the involved vessels, or to both (changes in local temperature and skin color, as well as trophic changes that may be prone to ulceration, diminished arterial pulsations and local gangrene)[2]. When the injury is severe and prolonged enough that the cells cannot survive, various ultrastructural and histologic changes can be recognized. Tissue necrosis is obvious both histologically and clinically. There are two broad types of tissue necrosis, coagulative and liquefactive. Coagulative

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necrosis is the most common and is usually the result of arterial obstruction or inherent vascular disease that leads to sudden cessation of blood flow or ischemic infarction[3]. An attempt to classify vasculitides serves to emphasize the complexity of the subject and the heterogeneity of the clinical spectrum of these diseases[4]. In polyarteritis nodosa, (PAK), medium-sized arteries are the sites of inflammatilon. This necrotizing vasculitis affects all age groups, but is rare in childhood; males are affected more frequently than females. The cause is unknown, but the disease has been reported to follow specific drug exposure[1]. In this paper, a patient having an acute vascular spasm of unknown etiology resembling polyarteritis nodosa is presented.

Case Report

A previously healthy 4.5-year-old girl was referred to the pediatric rheumatology unit with bruises on fingers and toes. She had no history of environmental causes, including drug intake. Symptoms of abdominal pain, fever and a swollen face preceded the symptoms of her extremities. On physical examination, her hands were cold and edematous and the fingers were cyanotic. She had massive necrosis on fingertips with an infected wound on the left index finger, which led to dry gangrene (Fig. 1). Her feet were also cold and mildly cyanotic (Fig. 2).

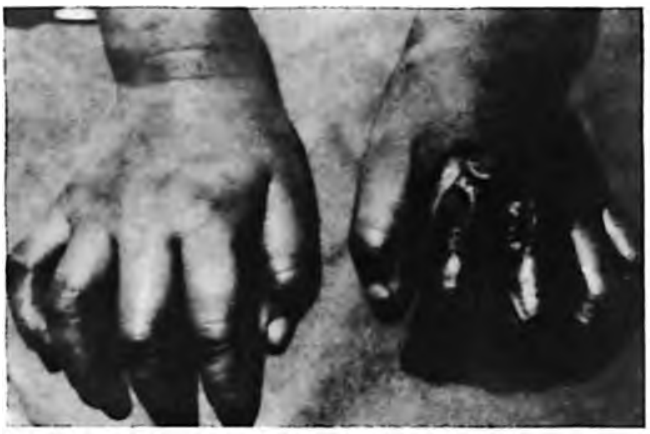

Fig. 1: There is massive necrosis on the fingertips and an infected wound on the left index finger.

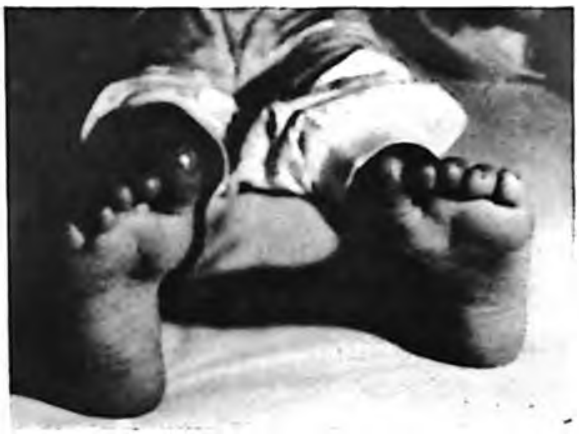

Fig. 2: There is mild cyanosis on the toes.

No pulses were detected on the upper extremities and the blood pressure, available only on the thighs, was 170/120 mmHg. Laboratory test results showed: leukocytosis (12.6×10^9 cells/L), C-reactive protein: 300 mg/L, erythrocyte sedimentation rate: 80 mm/hr, Hb: 8.5×10^9/L, Htc: 27%, and erythrocyte count: 3.2×10^9/L; the erythrocytes were hypochromic and microcytic. Renal function tests, including urine analysis, were normal. Prothrombin and partial thromboplastin time were within normal ranges. No hypocomplementemia was detected. Levels of antithrombin III (98%), protein C (2.20 mg/L), protein S

(13.8 mg/L), and anticardiolipin antibodies (ACA IgM: 1.4 MPL U/ml, ACA IgG: < 1.0 GPL U/ml) were within normal limits. Hepatitis B antigen was negative. On digital subtraction angiography, segmental obstruction was found proximally to a. brachiales and distally to a. radialis and a. ulnaris sinistra (Figs. 3-7). The

Fig. 3: Right brachial artery is obliterated in the proximal region and is refilled with the collateral arteries at the middle.

Fig. 4: Right brachial artery in the forearm is obliterated again just before the bifurcation and no filling is seen at the distal portion.

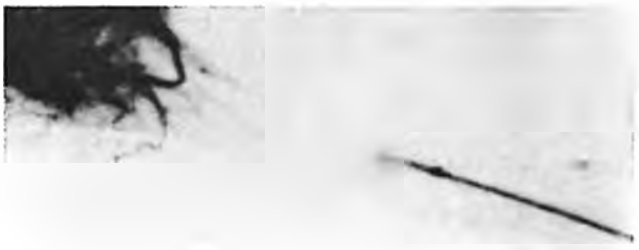

Fig. 5: Left brachial artery is obliterated in the proximal region, but it is refilled with larger collaterals.

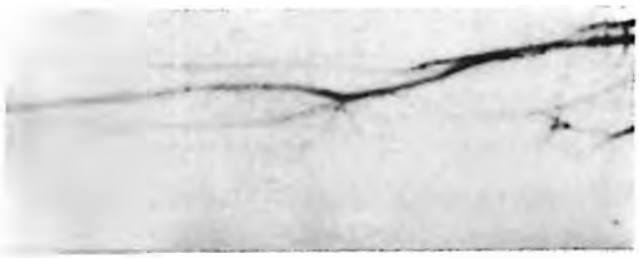

Fig. 6: Left brachial and radial artery can be seen in their entirety while ulnar and radial arteries cannot be filled at the level of the wrist.

patient was treated with high-dose methyl prednisolone (30 mg/kg/day) and cyclophosphamide (2 mg/kg/day) for immune suppression; acetyl salicylic acid (150 mg/day) and dipyridamole (5 mg/kg/day) as thrombolytic agents; nifedipine (2 mg/kg/day) for hypertension; piperacillin and amikacin for the infected lesion. Although she felt better and no signs of new skin necrosis on extremities were identified, the continued lack of pulses on the upper extremities necessitated a sympathatectomy, and the gangrenous fingers were amputated at the level of proximal interphalangeal joints (Fig. 8). Marked improvement was noted at the surgery sites, as well as in the general condition of the patient, within a week. She is being followed regularly since discharge.

Fig. 7: Left radial artery is refilled with the collaterals after being obliterated in the wrist line and forms the medial curve. The vascularizations of the third and fourth fingers are better than the others. The curve cannot be formed on the ulnar side.

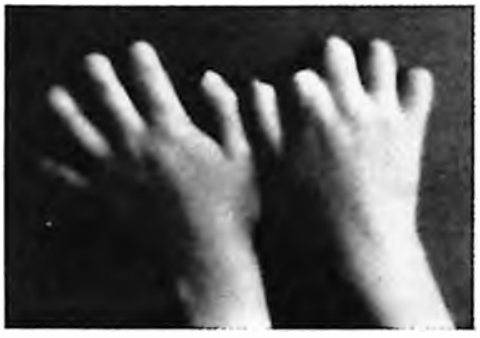

Fig. 8: The patient's hands after amputation. The thumbs are largely spared.

Discussion

The clinical features that suggest a vasculitic syndrome include weight loss, fever of unknown origin, abdominal pain, dermal necrosis, leukocytosis, and elevation of C-reactive protein and erythrocyte sedimentation rate[5,6]. This patient's symptoms of abdominal pain, fever, swollen face, hypertension, elevation of acute phase reactants, and gangrene of the fingers, strongly suggested an inflammatory vasculitis. Marked absence of pulses in a young girl should always bring to mind the most common giant cell arteritis of childhood, Takayasu's arteritis. The diagnosis of this disease is often delayed extensively because of the insidious onset, as well as the nonspecific nature of early manifestations.

The digital subtraction angiography of the entire aorta and its major branches showed no pathological changes, and helped to rule out this entity. The polyarteritis nodosa (PAN), though rare in a young pediatric age group, exhibits no single pattern of clinical presentation. However, abdominal pain, peripheral or central nervous system disease, arthritis and skin lesions occur in most children with PAN. Cutaneous involvement includes purpura, edema, peripheral gangrene, and nodular vasculitis. Although it can be suspected from a typical clinical presentation, diagnosis can be confirmed only by pathologic demonstration of characteristic lesions, or by radiologic documentation of aneurysms[7-9]. This patient had no typical lesions for a diagnostic biopsy. Digital subtraction angiography was done to determine the presence of abnormalities of the vascular system. There were obliterations in brachial arteries, but no aneurysm was detected, which is typical for polyarteritis nodosa. Ergotism is an acute or chronic poisoning caused by the ingestion of grain, or its products, infected by the ergot fungus, Clavicep purpurea. Patients may experience colic, coldness and cyanosis of the extremities, Raynaud's phenomenon, absence of peripheral pulsations, and gangrene of the digits[2]. As it is known that toxins are one of the factors triggering vasculitis, the symptoms and findings suggesting peripheral vasoconstriction in this patient could, presumably, be attributed to an ingestion of food or plant containing ergot or its derivatives, as the child's symptoms developed following three months spent in a village, where she might have been in close contact with the substances mentioned above.

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