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MEDICAL MEMORANDA

Pulseless Disease Presenting with Isolated Abducens Nerve Palsy and Recurrent Cutaneous Angiitis

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The clinical features of pulseless disease are the result of inadequate blood supply to certain organs such as the brain, eyes, and upper limbs due to narrowing or occlusion of the main branches of the aorta. The outstanding presenting symptoms are visual disturbances, ranging from photopsia and blurring to blindness, and syncopal attacks, often with convulsions. We report here a case of pulseless disease which presented with isolated abducens nerve palsy. We believe this is the first report of an isolated cranial nerve palsy associated with this disease. Cutaneous lesions similar to those described by Takezawa *et al.* (1966) as tuberculide-like eruptions and by Grimaldi (1970) as cutaneous microangiitis were also present.

Case Report

A 20-year-old man was admitted to hospital because of headache and double vision. He had been well until two years before, when he started having intermittent throbbing headache, which usually began in the right frontal region but soon radiated to the occiput and on some occasions also to the left side. The headache was worse on waking in the morning and sometimes it awakened him during the night. There was no associated nausea, vomiting, or fever. The attacks had been almost daily but with occasional interludes of up to three months. Three months before admission he had had pain in and around the right orbit and a month later had developed double vision. Five years previously he had had intermittent skin eruptions, which he described as "black dots," over the upper parts of both arms and over the back, and for the past two years these had persisted.

On admission the patient's blood pressure was 120/70 mm Hg in the right arm and 100/90 in the left. It was unobtainable in either leg. There was hyperpigmentation of the dorsal aspect of both thighs and papulonecrotic eruptions measuring 3-5 mm in diameter over the chest wall, the back, and all the extremities. The white cell count was 14,100/mm³ with 67% neutrophils, 23% lymphocytes, and 10% monocytes. Serial erythrocyte sedimentation rates over a two-week period were 35, 37, 96, and 79 mm in the first hour (Westergren). Serum protein electrophoresis showed the albumin to be 2.6 g/100 ml and globulin 4.16 g/100 ml, the globulin

fractions being alpha₁ 2.4%, alpha₂ 10.8%, beta 18.7%, and gamma 19.5%. All other findings, including examination of the cerebrospinal fluid, were within the normal range. A purified protein derivative tuberculin skin test was negative at 1:1,000 but positive at 1:100.

A radioactive brain scan was normal. Electroencephalography showed background activity of moderate amplitude with waves of alpha frequency seen posteriorly. There was some higher-amplitude, irregular, slow activity from the right hemisphere, more prominently from the posterior temporal region. X-ray examination of the chest, skull, and cervical spine and an intravenous pyelogram showed nothing abnormal. A retrograde transfemoral aortogram (see fig.) showed complete obstruction of the left common carotid artery at its origin with good collateral circulation via the thyrocervical trunk and intercostal arteries. The left subclavian artery

Retrograde transfemoral aortogram showing complete obstruction of left common carotid artery at its origin and no evidence of left subclavian artery.

was not seen and there was no evidence of subclavian steal syndrome. An abdominal aortogram showed partial obstruction and doubling of the right renal artery. The right internal carotid artery did not fill on right brachial angiography. The right vertebral artery and the posterior communicating artery were enlarged, feeding blood to both cerebral hemispheres through collateral circulation.

Biopsy of tissue from a papulonecrotic lesion on the back showed focal spongiosis of the epidermis leading to intraepidermal vesicle formation. Patchy cellular infiltration was present in the vicinity of blood vessels and adnexal structures of the dermis. The cellular infiltrate consisted of lymphocytes with a few neutrophils in some areas. Giant cells were also present. There was narrowing of the capillary lumen from oedema of the endothelial cells. The findings were consistent with papulonecrotic tuberculide and pityriasis lichenoides et varioliformis acuta of Mucha and Habermann, both of which represented two forms of vasculitis.

The patient's throbbing headache persisted in hospital, but his stay was otherwise uneventful and he was discharged after 25 days.

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The right abducens nerve palsy gradually improved. Six weeks after discharge lateral movement of the right eye was apparently full but the diplopia persisted and he complained of blurring in the left eye. Prednisolone was then given, but a week later vision in the right eye became blurred. The left carotid and the left radial pulses were absent and the right carotid pulse was weak. A short, soft systolic bruit was audible over the right carotid artery. Visual impairment was believed to be due to retinal ischaemia. Three months after discharge the patient still had headache and intermittent blurring of vision. The left auriculotemporal arterial pulsation was absent. Nicotinic acid added to the treatment gave no benefit. Four months after discharge he developed another crop of papulonecrotic eruptions over the back, buttocks, arms, legs, and face. The headache was intense and there was polyarthralgia involving the wrists, ankles, and knees. These symptoms persisted for over a month despite prednisolone in doses of up to 60 mg/day. Prednisolone was stopped and cyclophosphamide by mouth 150 mg/day gradually reducing to 100 mg/day was given instead. The headache became less frequent and less severe.

Aortography nine months later showed the state of the extra-cranial cerebral vessels to be unchanged.

Comment

The unilateral abducens palsy in this case might be attributed to a reduced blood supply to the nucleus of the nerve

in the pons. The lack of any other evidence of brain-stem lesion, however, was against that explanation. Isolated oculomotor palsy associated with various causes of arteriopathy such as diabetes mellitus and syphilis is well known, but, unlike those disorders, the lesions in Takayasu's (1908) disease are confined to the large arteries.

The onset and course of the abducens palsy in our patient were similar to those of a palsy due to infarction of the nerve trunk resulting from occlusion of the vasa nervorum, and the view that this could have been the nature of the lesion in our case is supported by the presence of widespread cutaneous microangiitis, manifested as papulonecrotic, tuberculide-like eruptions. Alternatively, the nerve may have been directly affected by a granulomatous process as in cranial neuritis, or polyneuritis cranialis—a condition that seems to be highly prevalent in some countries in the Far East, including Thailand (Steele and Vasuvat, 1970).

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Kidney Transplantation in Fabry's Disease

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Renal transplantation is of particular interest in congenital metabolic disorders. In this group, graft viability may be affected by the underlying disease, or the normal transplant may have a beneficial effect on the metabolic defect. Our observations in a patient with Fabry's disease who developed renal failure and received a renal allograft in 1967 provide further information about the value of renal transplantation in this inborn error of metabolism.

Case Report

Proteinuria detected on routine urine analysis prompted the examination of a 33-year-old man of Italian origin. Pertinent findings included slight burning discomfort of his hands and feet, purple-black telangiectasia, hypohidrosis and hyperkeratosis of the skin of the lower

abdomen, scrotum, and buttocks. These lesions had been present since childhood and increased in size and number since that time; biopsy confirmed the angiokeratosis. Slit lamp examination of the eyes showed opacities of cornea and lens. A 24-hour urine specimen contained 1.5 g of protein and the urinary sediment showed birefringent foam cells. Creatinine clearance was 60 ml/min.

Four years later the patient presented with a blood pressure of 220/110 mm Hg, pitting oedema, and a creatinine clearance of 15 ml/min. Shortly thereafter, in August 1967, haemodialysis was begun. Nephrectomy was performed two months later for intractable hypertension. Histologically extensive hyalinization of nearly all glomeruli and pronounced interstitial fibrosis were found. Fat stains showed lipid deposits in glomerular epithelial cells, mesangial cells, interstitial macrophages, and tubules, and in the walls of intrarenal blood vessels (figs. 1 and 2).

Enzyme measurements showed a total lack of ceramide trihexosidase in the acetone-dried powder of this patient's kidney, whereas simultaneously measured normal kidneys contained 4.6 nmol ceramide trihexosidase/mg protein/hr (Dubach *et al.*, 1969).

FIG. 1—Kidney section showing hyalinization of glomeruli, atrophic tubuli with protein casts and desquamated tubular foam cells, lipid deposits in arteriolar and glomerular epithelial cells, tubules, interstitial macrophages; adaptive intimal fibrosis of vascular walls. (P.A.S. X 60.)

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