PULSELESS DISEASE*

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TAKAYASHU (1908) reported a case of complicated cataract with unusual features, and Onishi and Kagoshima reported two similar cases at the same meeting with the added observation of absent radial pulses in one of them. It was not, however, until Shimizu (1948) reviewed the two articles that the disease became generally recognized by Japanese ophthalmologists as a rare but definite entity. Since then about 100 cases have been reported, most of them from Japan, a very few from America, and half a dozen from India (Misra, Prakash, and Agrawal, 1959; Damany and Shah, 1960; Pahwa, Pandey, and Gupta, 1959; Shikhare, 1921).

About 60 per cent. of the cases reported from Japan have occurred in persons aged 15 to 40 years; females greatly predominate and the condition is sometimes called "female arteritis". A non-specific arteritis produces gradual occlusion of the innominate, the left common carotid, and the left subclavian arteries where they leave the aorta.

20 per cent. of such cases are due to syphilitic changes in the arteries; this occlusion of the main branches of the aortic arch is commoner in males.

The condition has also been called segmental thrombo-obliterative arterial disease, aortic arch syndrome, Martorelli's syndrome (by Damany and Shah, 1960), Takayashu's syndrome, and ophthalmo-angiopathica hypotonica due to low central retinal artery pressure.

Signs, Symptoms, and Prognosis

Clinical.—The presenting symptoms are protean, depending upon the nature and extent of the obliterative process and the adequacy of the collateral circulation. The following are the classical findings:

- (1) Absence of pulse in the upper extremities, commonly in the radial and carotid arteries.
- (2) Arterio-venous anastomosis surrounding the optic nerve.
- (3) Hypersensitive carotid sinus, frequently resulting in attacks of syncope and vertigo.

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- (4) Development of cataracts—usually of the nuclear type.
- (5) Absence of detectable blood pressure in the upper extremities.

Ocular.—The most frequent early ocular symptom is that of recurrent transient blurring of vision, especially upon turning the face upwards, or when quickly assuming the upright position. This blurring of vision may be associated with the sensation of flashes of light. As the occlusion of the vessels progresses the visual symptoms occur more frequently and last longer.

Dilatation of the conjunctival and episcleral blood vessels, pigmentary deposits in Descemet's membrane, Krukenberg spindles, mydriasis, iris atrophy, unequal pupils, vascularization of the iris (occasional), and nuclear cataracts have been recorded. In the fundus, obliteration of the peripheral arterioles of the retina, very low blood pressure in the central retinal artery (which may be the only early sign), and occasional occlusion of the central retinal artery with primary optic atrophy may be seen.

Large retinal haemorrhages, with cotton-wool exudates (greyish-white flecks located around the macula) may also be present, and some cases show retinal detachment and retinitis proliferans.

General

CIRCULATION.—Absence of palpable arterial pulsations and detectable blood pressure in the upper extremities, and a hypersensitive carotid reflex.

C.N.S.—The neurological disturbances resulting from cerebral ischaemia vary from relatively mild symptoms of headache, vertigo, and faintness to more severe convulsions, aphasia, and actual paralysis. Hemiplegia is occasionally encountered.

E.N.T.—Perforated nasal septum, ulceration of the palate, atrophy of the facial muscles and bones, and enlarged thyroid.

SKIN AND APPENDAGES.—Very delicate skin, fine poorly-pigmented hair, and loss of hair.

TRUNK AND UPPER EXTREMITIES.—Arterio-venous aneurysms on tips of fingers, and pulsating subcutaneous arteries over the back, chest, and upper abdomen.

LOWER EXTREMITIES.—Hypertension in legs.

Laboratory Findings.—Raised erythrocyte sedimentation rate and leucocytosis, positive tuberculin test, increased prothrombin time, and high blood cholesterol.

Pathology.—Thrombosis of subclavian and carotid arteries. Arteritis affecting the adventitia and media with disorganization of internal elastic membrane.

Shimizu and Sano (1951), who felt that the lesion was suggestive of tuberculosis, noted the presence of Langhans' giant cells. No organisms were found by smear, guinea-pig inoculation, or culture.

Differential Diagnosis.—The condition must be distinguished from temporal arteritis, Buerger's disease, and periarteritis nodosa.

Prognosis.—This is poor, both for vision and longevity. Cataract extraction does not help as the retina is ischaemic.

Treatment.—None is known to be effective.

Case Report

A Hindu female aged 40 years was admitted to hospital with the complaints of fainting attacks and dizziness whenever she tried to rise from a lying position for the past 1½ years, and complete loss of vision in the right eye and marked diminution of vision in the left eye for 15 days. During the attacks, which lasted for only a couple of minutes, the hands became numb and felt colder than the feet, and there was marked but transient blurring of vision. A few months before admission she had pain in the right temple accompanied by severe pain in the right ear with discharge. This pain gradually spread to the right eyeball and then to the whole of the right side of the face.

There was also a nasal discharge which persisted for some time. The pain later spread to the left side of face and left eyeball, but gradually disappeared without treatment.

There had been a gradual diminution of vision in both eyes even between the attacks of syncope.

Her past and family history showed nothing noteworthy. She had had four children; the first was stillborn after prolonged labour, and the other three were alive and healthy.

General Examination.—She looked anaemic with a pale face. She was comfortable in a lying position, but a syncopal attack occurred as she was getting up.

Pulse.—Very feeble brachial pulsation in both arms and in the neck. Radial pulses not palpable. Heart rate 92 per minute. Blood pressure: left arm 76/70, right arm 70/70, right leg 170/86, left leg 164/88 (mm. Hg).

Heart size normal. No intercostal pulsations visible. Markedly visible pulsation on right side of base of neck, but no pulse could be felt.

Ocular Examination

Right Eye.—Pupil dilated and fixed. Aqueous flare + + +. Fine pigmentary deposit at back of cornea. Iris atrophic and pattern lost. Lens showed fluffy cataract. Tension 12 mm. Hg (Schiotz).

Left Eye.—Pupil dilated and fixed. Aqueous flare +. A few pigmentary deposits at back of cornea. Lens showed fluffy cataractous changes along the Y-suture. Tension normal. Visual acuity 4/60.

The left optic disc was hyperaemic, margins slightly blurred, arteries markedly attenuated. Veins congested and full with uneven calibre and fragmented circulation. New blood vessels on and around the disc between the upper blood vessels. Very slight pressure on the upper lid made the fundus pale and the optic disc white. No haemorrhages or exudates seen. Foveal reflex absent and macular area showed pigmentary disturbances. No abnormality at periphery.

E.N.T.—Right eardrum perforated. Nasal septum almost absent, with very large perforation.

Laboratory Investigations

Mantoux Test: 1:10,000 — — —
1: 1,000 + — —

Blood: Red blood count 3.4 million/c.mm. Haemoglobin 9.15 g. per cent. White blood

count total 7,500/c.mm., (polymorphs 57 per cent., lymphocytes 42 per cent., eosinophils 1 per cent.) Bleeding time: 3 min. Coagulation time: 5 min. Prothrombin time: 17 sec. Wassermann reaction and Kahn test negative. Erythrocyte sedimentation rate 110 mm./1st hr.

Urine: Negative.

Fasting Blood Sugar: 100 mg. per cent.

X-rays: Chest and skull normal.

Operation.—Cataract extraction was done in the right eye but no visual improvement followed. The fundus showed similar changes to those in the left eye.

Result.—Complete blindness ensued.

Comment

The present case differed from those hitherto described in that there was no leucocytosis and an almost negative Mantoux test. However, the erythrocyte sedimentation rate was very high, and this is a constant finding in such cases.

Summary

The literature of pulseless disease (Takayashu's syndrome) is briefly summarized. A case of the syndrome is described in which the aetiology was non-syphilitic.

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