The number of patients in the present series represents a high incidence relative to the total number of patients suffering from lumbar disk disease. The incidence is also high compared with other series. This may be due to the method of reference of patients with disk disease to the neurosurgical unit of the Maida Vale Hospital. All in this series had been examined by one or more consultants before transfer.

However, the cauda equina syndrome as produced by lumbar disk prolapse or extrusion is not so rare as is generally supposed. In the first nine months' tenure of my present appointment I have had three patients suffering from this syndrome referred to my care. Fig. 2 is a photograph of a " disk sequestrum " removed at operation from the last of these patients.

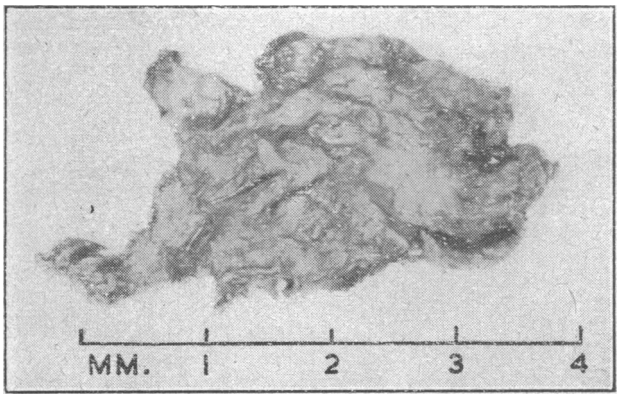

FIG. 2.—Single extruded fragment of disk comprising bulk of nucleus pulposus.

Mention is required of the association of manipulation and the cauda equina syndrome. Such a manœuvre precipitated the onset of the syndrome in one patient in this series. If there is any evidence to suggest a central disk prolapse not yet producing a cauda equina syndrome, manipulation should be avoided. The finding at operation in a number of these patients of a thinned annulus and posterior common ligament, and the fact that so many of the frank cases of the syndrome follow relatively trivial traumata—acting on an already diseased disk—indicate the danger of manipulation.

The profound disturbance of function which follows acute central prolapse or extrusion of a lumbar disk is clear from a study of the patients forming the basis of this paper. The slow return of this function, with delay in diagnosis and operation, is also apparent. These lesions do not of themselves endanger life. However, when no relief is afforded, the paralysis of bladder and sensory function leads to ascending urinary infection and decubitus ulcer formation, and so result in a shortening of the life-span. Uraemia or septic absorption from bed-sores brings to an early end a miserable and limited existence.

Summary

In order to improve the prognosis attention is drawn to the need for early diagnosis and operative treatment of patients with the cauda equina syndrome due to lumbar central disk prolapse or extrusion.

Thirteen patients have been chosen for study, being nearly one-third of those having central lumbar disk lesions and one-tenth of those with compressing disk lesions verified at operation and seen over a six-year period.

The 13 patients are classified into two groups. The main diagnostic features are described, the role of injury and the rapid development of the syndrome in most cases being specially stressed.

The manner in which the cauda equina may be involved is described.

Factors in prognosis are mentioned: pain; sphincter, sensory, and motor involvement; and duration of the syndrome. Their relative importance is discussed.

Five illustrative case-histories are given, and the main clinical features of the whole series are tabulated.

The discussion includes remarks concerning the diagnosis and frequency of the syndrome. The possible danger of manipulation is mentioned and allusion made to the plight of the unrelieved sufferer from this condition.

I thank Mr. Valentine Logue for granting me access to his personal notes of these patients and for allowing me to perform many of the operations, I am indebted to Dr. D. Sutton for permitting me to include Fig. 1. My thanks are due to the medical committee of the Maida Vale Hospital for allowing me to publish. I am grateful to Mrs. Davis for secretarial assistance.

REFERENCES

Jennett, W. B. (1956). *J. Neurol. Neurosurg. Psychiat.*, **19**, 109.
O'Connell, J. E. A. (1950). *Ann. roy. Coll. Surg. Engl.*, **6**, 403.

PULSELESS DISEASE, OR TAKAYASHU'S DISEASE

CASE REPORT

BY

J. M. PAHWA, M.S., D.O.

M. P. N. PANDEY, D.O.

Eye Hospital, Sitapur, India

AND

D. P. GUPTA, M.D., M.R.C.P.Ed.

Medical Officer in Charge, District Hospital, Sitapur, India

Cases of unilateral absence of pulse due to aortic aneurysm have been known since 1757, when William Hunter described the condition, but bilateral absence of the arterial pulses is exceedingly rare. In 1908 Takayashu reported a peculiar case with cataracta complicata and unusual ocular findings. At the meeting at which the case was described Onishi and Kagoshima reported two similar cases, with the addition that radial pulsation was absent in one of the patients. It was not, however, until the review by Shimizu and Sano (1953) that this pulseless-disease–aortic-arch syndrome, or reversed coarctation, became recognized by Japanese ophthalmologists as a rare but definite entity. Since that time 69 cases have been reported from Japan, and 34 from Europe and America, including several in recent years. The present paper records a further case, the first one from India.

Clinical Signs and Symptoms

Clinically, pulseless disease is characterized by three cardinal signs; the most common is absence of palpable

arterial pulsation in the upper extremities, in the carotid, and in the superficial temporal arteries. The other two signs are peripapillary arteriovenous anastomoses and microaneurysms, and a hypersensitive carotid sinus which frequently cause attacks of syncope and vertigo. Two additional signs often considered classical are the development of cataract and the absence of blood-pressure in the upper limbs. Syncopal attacks are frequently associated with convulsion, and sometimes are followed by transient hemiparesis.

Other reported ocular findings are obliteration or irregularity of arteries in the periphery of the retina, with clumping of blood and pallor of the fundus, mydriasis, iris atrophy, dilatation of conjunctival and episcleral vessels, pigmented deposits on the back of the cornea, retinal and vitreous haemorrhages, retinitis proliferans, and very low central artery blood-pressure. Dodo (1950) therefore applied the term " ophthalmo-angiopathica-hypotonica " to these ocular manifestations.

In addition to these ocular findings, the following have been noted: delicate skin ; fine poorly pigmented hair ; attacks of syncope ; intermittent claudication in the upper limbs, but rarely gangrene ; speech disturbance ; pulsation of subcutaneous arteries over the back, chest, and upper abdomen ; attacks of pain in the orbit, eyeball, and chest ; hemiplegia ; hypomenorrhoea and amenorrhoea. The most common early symptom is recurrent transient blurring of vision, especially when turning the face upwards or quickly assuming an upright position. These patients often hold their head, with the face directed downwards, as they find that vision is thereby often improved because of the increased blood supply to this area. A high blood-pressure in the legs has been reported in about half the cases. Ross and McKusick (1953) quote convincing experimental evidence and suggest that hypertension arises from progressive cerebral ischaemia. The disease slowly worsens, and after a period of blindness death usually ensues from cerebral infarction.

Aetiology

Pulseless disease has a predilection for young females, who outnumber males by 10 to 1. It is seen during or after puberty. An acute infection, such as scarlet fever or tonsillitis, may precede the onset of the disease.

There appear to be two definite forms of pulseless disease. One is associated with pathological findings compatible with syphilitic aortitis at post-mortem examination. The other is more common in females, and fits more closely Takayashu's description of the disease. In these cases the serological test is negative. Post-mortem examination has been reported in only six of the non-syphilitic cases. The pathological process was restricted to large arteries close to their origin, and consisted of thickening and hardening of the adventitia, disruption of elastic lamellae of media by collagenous connective tissue, and infiltration by round cells and plasma cells ; intima was thickened by atheromatous plaques, and the lumen occluded by organized thrombus. Biopsy of peripheral vessels revealed simple narrowing due to diminished blood flow, but there was no sign of arteritis. As an aid to diagnosis, the site of obstruction of the brachiocephalic trunks at their origin can be confirmed by aortography.

The cause of the condition is obscure. The sedimentation rate is high, and the presence of lupus erythematosus (L.E.) cells has not been reported. Probably the disorder is caused by non-specific arteritis at an unusual site. Shimizu and Sano (1953) felt that histologically the picture was suggestive of a tuberculous lesion, and the presence of Langhans giant cells was noted ; however, no organisms were found by smear, by guinea-pig inoculation, or by culture.

Other theories advanced as the cause of the second group of cases include Buerger's disease, allergy, panarteritis of unknown aetiology, and collagen disease.

Pathological Physiology

The fundamental lesion is an occlusion of the great vessels, usually the brachio-cephalic trunks or sub-clavian and carotid arteries close to their origin from the aorta, with consequent ischaemia of the brain, eyes, and upper limbs (Fig. 1). To compensate for the

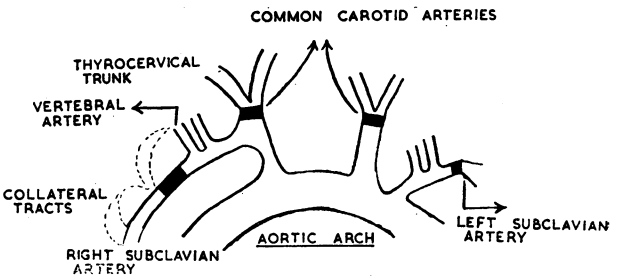

FIG. 1.—Diagram showing probable site of lesion in pulseless disease.

diminished blood supply there is a development in the collateral system. The collateral blood supply to the face is often sufficient to provide the necessary nutritive material. This is done by way of the subclavian artery to the thyroid gland, on to the superior thyroid artery, then to the external carotid and the vessels of the face. The enlarged thyroid in pulseless disease may result from this process.

The collateral blood supply to the brain via the vertebral arteries of the circle of Willis, then to the middle cerebral and anterior cerebral arteries, is often insufficient and may result in hemiparesis and loss of memory. Because of this, nutrition to the eyes is also affected and may result in structural atrophy, especially of the retina.

The collateral blood supply to the upper limbs via the pectoral branch of the thyro-cervical trunk on to the thoraco-dorsal artery of the humerus and thence to the deep brachial artery is usually sufficient for nutrition of this area. A continuous murmur may be audible over large collateral vessels, and in the neck over main arteries, owing to post-stenotic dilatation.

Whenever there is a constant and greatly decreased arterial pressure, such as is found in the retinal arterioles, the finer capillary channels are short-circuited and arteriovenous anastomoses occur, thus explaining the early arteriovenous anastomosis in the periphery of the retina and the large arteriovenous anastomosis seen around the optic nerve head in the late stages of the disease. The clubbing of the fingers and the arteriovenous anastomosis in the tips of the fingers sometimes seen can be explained in a similar manner. Because of the decreased blood supply to the area, a lowered arterial pressure in the retinal arteries can be easily demonstrated by pressure on the globe,

and may be an early finding of pulseless disease. Cataract formation can also be explained because of decreased nutrition, which also leads to gradual atrophy of the peripheral retinal elements.

Attacks of dizziness, convulsions, or unconsciousness seem to be associated with a hypersensitive carotid sinus and are often produced merely by the patient stretching his neck, or by pressure on the carotid sinus. The hypersensitivity of the carotid sinus is explained on the basis of an insufficient blood supply to this area over a long period. Pinkham (1955) and De Bes *et al.* (1955) reported the following findings in the eye:

Lens: Coagulation necrosis. Ciliary body: Atrophy of muscle and ciliary processes. Retina: Clumping of rods and cones; decrease in number of ganglion cells; formation of new vessels in nerve fibre layers. Central retinal artery: Marked proliferation of the intima. Optic nerve: Demyelinization.

Differential Diagnosis: Prognosis

Temporal arteritis, Buerger's disease, and panarteritis nodosa may be considered in differential diagnosis, but these can be easily differentiated and ruled out.

The prognosis is poor, though some patients can carry on, with limited activity, if their collateral circulation to the brain is sufficient. As regards vision, prognosis is unfavourable. In a few cases cataract extraction has been done, only to reveal an atrophic retina with poor perception of light. The course of the disease ranges from 1½ to 14 years. Death may result from chronic debility due to cerebral ischaemia, though it may be the result of myocardial disease.

Treatment

No known treatment is effective. Caccamise *et al.* (1952, 1954) have suggested that cortisone and corticotrophin may be of value if treatment is initiated before irreversible changes have occurred; therefore cortisone and anticoagulant therapy is worth a trial in early phases of the disease. Lessof (1958) reports considerable physiological and symptomatc improvement in a patient on prednisolone treatment. A thrombectomy performed through the external carotid artery, coupled with denervation of the carotid sinus and removal of the carotid body, has been tried by Shimizu and Sano (1953) with some relief of symptoms. In another case they tried grafting of a piece of vein into the area where the thrombosis had been resected. This relieved the attacks of syncope, but there still appeared to be little flow of blood to the brain through this segment. Ross and McKusick (1953) described the successful relief of distressing syncope by resection of the carotid sinus. Sympathectomy of the cervical sympathetic ganglion has also been tried, but with poor results.

Case Report

A Hindu female, aged 27, was admitted to hospital on May 6, 1958, with a history of attacks of fainting fits for a year and diminution of vision for 20 days.

Nearly one and a half years previously she had intermittent headache on one side, and the eye on this side became red, but since the fainting attacks started the headache has subsided. The fainting is precipitated on getting up from a lying-down position, on looking up again, and on attempting to unlock or open the *Kundi* of the doors of the house. At first she used to fall many times and hurt herself during the attack of fainting, but she has now learnt to sit down or lie down at the onset of the attacks. If she puts her hand under her head or pillow while resting, the fingers become numb and flexed.

Past History.—She had typhoid fever in 1947. There was no history of rheumatic pain. Last year she was admitted to the general hospital, and the absence of a pulse was thought to be due to weakness and debility.

General Examination

The patient was a young, slightly built woman, who looked more than her age. Married 10 years ago. Two children, both healthy. The eyes appeared to some extent enophthalmic. Heart slightly enlarged, but sounds normal. Radial, brachial, and temporal arteries not palpable, and without pulsation. Blood pressure could not be recorded in the upper limb, but in the lower limb (popliteal artery) it was 230/90. Bruits audible on right side of chest in front, ½ in. (1.3 cm.) above and external to the mid-clavicular line and on either side of the back in the ninth intercostal space 4 in. (10 cm.) from the midline.

Eye Examination

She had perilimbal injection with some prominence of conjunctival and episcleral vessels. The cornea had pigmented deposits over its posterior surface, more on the right side. Both pupils were dilated, and sluggish to light and convergence. The iris was atrophic and showed many vessels, mostly in the ciliary part (different from rubeosis of diabetes, etc.). Tensions were: right side, 27 mm. Hg; left, 25 mm. (Schiøtz). Gonioscopic examination revealed vessels in the angle, which was very narrow and had some pigment deposits.

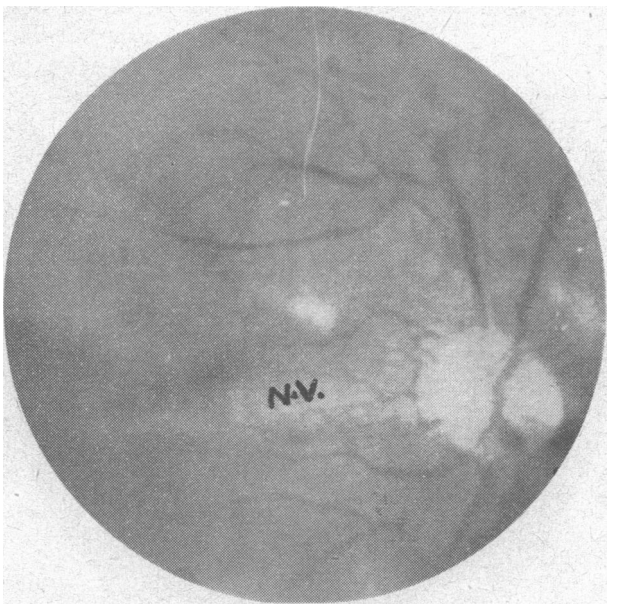

FIG 2.—Fundus photograph of right side showing new vessels (N.V.) near the disk, with somewhat dilated veins.

Fundus.—Right disk was covered with new blood vessels anastomosing between themselves (Fig. 2). A patch of proliferative retinopathy extended from the temporal side of the disk to the macula in the form of a broken girdle (Fig. 3). Veins were tortuous; arteries were contracted, with faint pulsation, and at places showed microaneurysms and arteriovenous anastomoses (Fig. 3). Vessels could not be traced up to the periphery, which was pale. The veins on the left side were tortuous and prominent. Many arteriovenous anastomoses were seen, with new vessels near the disk (Fig. 4). Some of the vessels on both sides showed

peri-sheathing and were obliterated. Pressure in the central arteries was observed by ophthalmodynamometer and found to be very low.

Laboratory Examination

Urine, total and differential W.B.C., and W.R. were normal; E.S.R. was 18 mm. in one hour; haemoglobin was 65%. *X*-ray films of the chest showed slight enlargement of the heart. Patient was given local cortisone drops, and prescribed "deltacortril" tablets with not much relief.

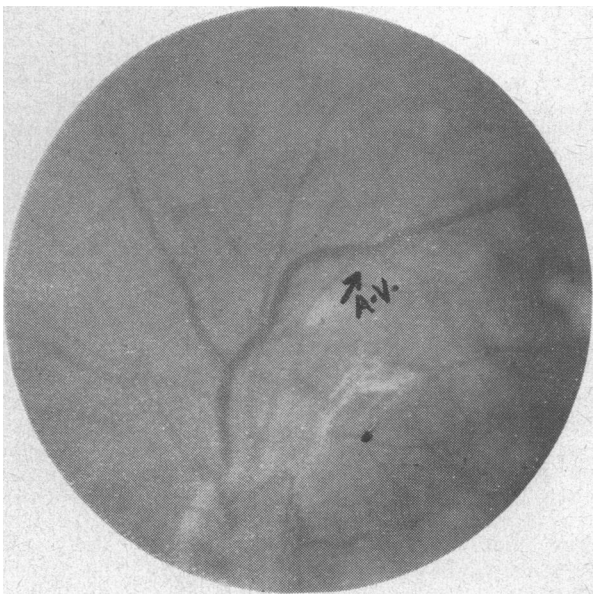

FIG. 3.—Fundus photograph of right side showing arteriovenous aneurysms and patch of proliferated retinopathy extending from temporal side of disk.

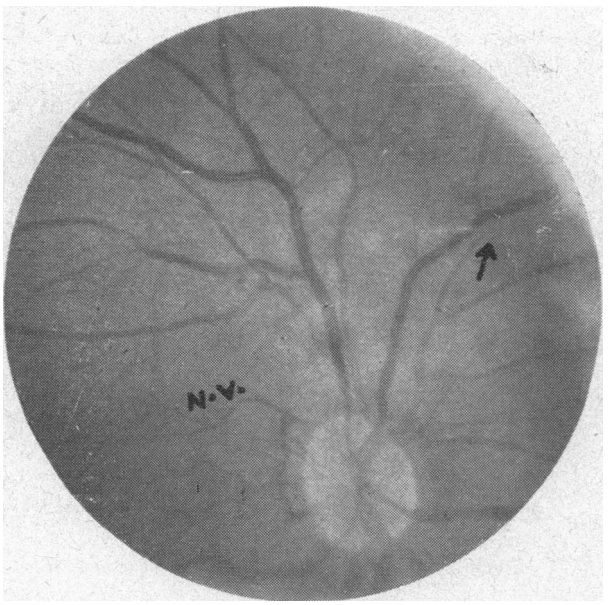

FIG. 4.—Fundus photograph of left side showing new vessels and arteriovenous aneurysms.

The patient came to this hospital again on July 20, 1958, with the history of attacks of headache on one or other side, lasting for a day or two. She also complained of total loss of vision in the right eye. On examination the right pupil was dilated and had mature cataract (duration 14 days). Vision in the left eye was 2/60, with early vacuolation in the lens cortex as seen by slit-lamp. The whole fundus, including the disk was pale. The vessels, particularly the arteries, were very attenuated; they became indiscernible as we traced them towards the periphery, and were completely invisible even before the equator. Loupes of new vessels were present near the disk. Perivascular sheathing, and even obliteration of the arteries, were now quite obvious.

The patient returned on November 26, 1958, with total loss of vision in both eyes, and cataract had developed in the second eye also. Pupils were dilated and fixed, with perception of light only. Cataract extraction (extracapsular, due to raised tension) was done, but under small peripheral iridectomy the iris bled because of the vascularization, and took a long time to absorb. The fundus was pale and there were many arteriovenous vessels near the disk. Cataract extraction did not improve her sight.

Summary

A typical case of pulseless disease in a young woman, the first from India, is described, with typical fundus changes, absent pulsation in the upper limbs, cataracts, attacks of syncope, and raised blood pressure in the lower limbs, with tendency to rise of ocular tension, probably due to new vessels and pigment deposits in the angle of the anterior chamber. The case passed on to complete blindness, though cataracts were removed successfully.

BIBLIOGRAPHY

Aggeler, P. M., Lucia, S. P., and Thompson, J. H. (1941). *Amer. Heart J.*, **22**, 825.
Applemans, M., Michiels, J., and Missotten, L. (1956). *Bull. Soc. belge. Ophtal.*, No. 110, p. 141. (Abstracted in *Amer. J. Ophthal.*, 1956, 41, 1117.)
Ask-Upmark, E. (1954). *Acta med. scand.*, **149**, 161.
Boccardelli, V. (1955). *Progr. Med. (Napoli)*, **11**, 687. (Abstracted in *J. Amer med. Soc.*, 1956, **160**, 1261.)
Caccamise, W. C., and Okuda, K. (1954). *Amer. J. Ophthal.*, **37**, 784.
—— and Whitman, J. F. (1952). *Amer. Heart J.*, **44**, 629.
Chang, Hsioh-Teh, Chang, An, and Ch'iu, Fu-Hsi (1955). *Chin. med. J.*, **73**, 163.
De Bes, L. T., Lucas, J. G. S., and Barcons, F. B. (1955). *Brit. Heart J.*, **17**, 484.
Dodo, T. (1950). *Jb. Kurashiki Zentralhosp.*, **21**, 2.
Hunter, W (1757). *Medical Observations and Inquiries*, 1, 323.
Lawson, L. J. (1954). *Amer. J. Ophthal.*, **37**, 788.
Lessof, M. (1958). *Guy's Hosp. Rep.*, **107**, 53.
Mengis, C. L., Dubelier, W., and Barry, K. G. (1958). *Amer. Heart J.*, **55**, 435.
Pinkham, R. A. (1955). *Acta XVII Concilium Ophthal., 1954*, **1**, 348.
Ross, R. S., and McKusick, V. A. (1953). *A.M.A. Arch. intern. Med.*, **92**, 701.
Segal, F., and Berezowski, A. (1958). *Amer. Heart J.*, **55**, 443.
Shimizu, K., and Sano, K. (1953). *J. Neuropath. clin. Neurol.*, 1, 37.
Skipper, E., and Flint, F. J. (1952). *Brit. med. J.*, **2**, 9.
Sökmen, C. (1958). *Amer. Heart J.*, **55**, 433.
Takayashu, M. (1908). *Acta soc. ophthal. japon.*, **12**, 554.

"Most men of science would disclaim any great concern with philosophy and especially with metaphysics. But what is metaphysics? In the last resort it is the search for a statement subsuming the ultimate nature of all reality. Science has more modest hopes, and modern physics led the way to a considerable renunciation in this respect which has had a profound influence on philosophy itself. Yet in science the total neglect of philosophical aspects may be unfortunate. It can lead, on the one hand, to statements which are sensational but unsound, and, on the other, to an unhealthy indifference to the relationship of science to existence in general. And in any event, even though science may seem to decline metaphysical statements, it not only throws out challenges to philosophy but is frequently itself confronted with problems inescapably philosophical in nature." (Anniversary address by Sir Cyril Hinshelwood, P.R.S., November 30.)