RETINAL ANGIOMA WITH CERVICAL CORD LESION

REPORT OF A CASE

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Y.P.H. aged 42 male Chinese was first seen in September 1955. He was a dispensing attendant for many years. When the illness began, he noticed some stiffness of his right arm coming on gradually without any systemic complaint. This stiffness made him clumsy in handling things, and he was apt to drop bottles when holding them in his right hand.

A month later, he burned his fingers accidentally with strong carbolic acid quite badly, and was surprised to feel no pain. Unaware of the anaesthetic properties of carbolic acid, he grew worried, and began to inflict pain on his right arm by pricking the skin. He found that the right arm was devoid of any painful sensation.

Ten days later, the right arm became so weak that he was alarmed. He used to be able to carry a weight of 40 lbs. with his right hand, but he found that he could only manage 20 lbs. then. Simultaneously, he began to notice numbness and tingling in the radial side of the right upper limb, and also some weakness in the right lower limb, but there were no abnormal sensations in the legs.

Bowels and micturition were normal and he had no history of previous illness nor was there any relevant family history of nervous diseases. 10 years ago his right eye was injured during a football game and had to be removed.

On examination at entry in September 1955, he was found to have normal mental functions. There was a deep ulcer on his right index finger which he said was painless. There was a total loss of pain and thermal sensation from C2 to T3 on the right side, but tactile sense was preserved. The right arm and right leg showed some weakness with a power of 4 on muscle power grading. Reflexes were absent in the right upper limb and increased in the right lower limb. Abdominal reflexes were absent on the right side and right plantar response was equivocal. The cranial nerves were normal and the left side of the body was normal apart from a slight weakness of grip.

Investigations showed a normal cerebrospinal fluid, Kahn test was negative for blood and

C.S.F. and X-ray of skull and cervical spine was normal.

A provisional diagnosis of an elongated lesion in the cervical region near the central canal was made? Syringomyelia.

Progress: He was kept under observation for 2 months when it was evident that he was perceptibly though very slightly getting weaker. In December 1955, he was given a course of deep X-ray to his neck — 4000 r. At the conclusion of the deep X-ray irradiation, he found that although the sensory disturbance remained unchanged, his motor power improved until his left hand was normal and his right hand was much stronger. A month later, he was able to return to work.

He continued to come up for inspection every few months or so and the general condition remained much the same until 1958 when he stopped attending as he felt he was able to work, and also he was not making further progress.

From the middle of 1960, he had trouble with his vision and in January 1961 was admitted to the Ophthalmic ward. There he was found to be essentially the same as far as his cervical lesion went, but, in addition, it was found that he had haemorrhages and exudates in the left retina, retinitis proliferans near the disc, and some peculiar vascular abnormalities. The inferior nasal and temporal vessels were dilated, especially the inferior nasal artery which on being traced peripherally seemed to split and lead up to a tumour like structure on the surface of the retina nasal to the disc. On careful examination it appeared to be an angiomatous growth covered by glial tissue and exudate. No papilloedema was seen. Two shallow detachments of the retina were seen, one near the angiomatous malformation, and the other near the macula (Figure 1). A diagnosis of Retinal Angiomatosis probably associated with Syringomyelia was made.

Discussion: Dissociated anaesthesia and the distribution of the motor weakness suggested a linear lesion of some dimension near the central canal. At this age, the probabilities are either a syringomyelic cyst or an eppendymoma, since

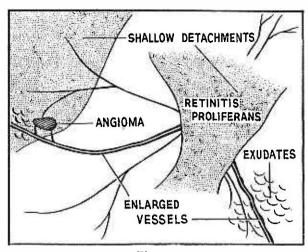


Fig. 1.

the skeleton was normal and there were no other detectable anomalies. Both these conditions would respond to deep X-ray, but the gross difference between the deficit in the right and left seems more in keeping with syringomyelia than eppendymoma, as the latter in the central canal would tend to exert pressure on all directions, and hence produce a relatively symmetrical deficit, whereas syringomyelic cyst is well known for its ramifications and extensions in one or more directions leading to quite confined lesions.

The absence of progress after deep X-ray over a period of six years also favours the diagnosis of syringomyelia which tends to be slow in progress, and may even remit. The finding of the eye in 1961 seems to suggest that angioma was the primary lesion and its presence through bleeding has led to retinitis proliferans and retinal detachment. A normal fundal examination in 1955 strongly points to the possibility of a recent acquisition of the eye complications. The retinopathy associated with angiomatosis retinae is usually seen between the 2nd to 4th decade and it is not unlikely that a single angioma situated far out in the periphery and small to start with might be missed if the associated retinopathy had not yet developed.

Angiomatosis of the retinae is associated with angiomata of the nervous system such as described by Lindau, and it would be tempting to suggest that the cervical lesion has been also an angioma which by its situation simulates a syringomyelic cyst. On the other hand, syringomyelia can be associated with many congenital defects, and vascular defect is a known association (Wilson) and the possibility of this being a case of retinal angioma associated with cervical syringomyelia cannot be entirely ruled out.

Since this report was written, he has remained static neurologically and his vision has improved till he can see enough for his daily requirements.

REFERENCE

Kinnier Wilson, S.A. (1954): Neurology. 2nd Ed., page 1187. London, Butterworth & Co., Ltd.