

A CASE OF ADIE'S SYNDROME*

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In April, 1943, I saw a male patient, aged 21 years, who complained of a minor ailment, a slight weakness of the abdominal wall following appendicectomy. At the same time it was observed that his pupils were unequal, the right being about two thirds the size of the left. When he was questioned he replied that he knew of this inequality of the pupils and that he had first noticed it about one year previously when he was looking into a mirror, at a small stye. He had since, paid little or no further attention to the pupillary inequality.

In July, 1941, he noticed that his vision at times was momentarily blurred when he looked from a near to a distant object and vice versa. The blurring of vision did not last longer than two seconds. Occasionally he also experienced diplopia, also momentarily. Because of this he consulted an ophthalmologist who diagnosed a compound hypermetropic astigmatism and lenses were accordingly prescribed, obtained and worn with a beneficial result. The wearing of the lenses did not restore the equality of the pupils and the probability that the abnormality was not due to an error of ocular refraction was considered and investigated.

His father and mother are both well and have a good medical history.

Apart from the pupillary abnormality the eyes were normal in every other respect, no irregularity nor eccentricity, synechiae nor pathology such as atrophy of the iris. The lenses, fundi, optic discs and ocular tension were normal. The right pupil was two thirds the size of the left. When the pupils were tested for convergence accommodation there was no apparent pupillary response of the right eye only. There was apparent abolition of the light reflex and of the consensual photomotor reflex from the same eye. Exposure to darkness for five minutes and subsequent exposure to light did not elicit the light reflex, nor did forced closure of the eyelids (Piltz—Westphal). Pinching of the cervical skin failed to stimulate dilatation of the pupils. There was no hippus nor a change in the size of the pupils when the eyes were abducted (Tournay's) but corneal stimulation produced a just detectable contraction.

A mydriatic instillation of $\frac{1}{2}$ per cent. atropine sulphate solution resulted in fairly rapid dilatation of both pupils but when both were maximally dilated the relative sizes of the two pupils

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remained the same, the right two thirds the size of the left. As the effect of the atropine wore off the pupils retained their relative sizes.

The instillation of $\frac{1}{2}$ per cent. eserine into the eyes resulted in equal maximal contraction of the pupils.

A test suggested by Kyrieleis Werner was made. Strychnine sulphate, gr. 1/60 was subcutaneously injected and the pupillary reactions tested two minutes after the injection, then after thirty minute, sixty minute, 12 hour and 36 hour intervals. At the end of the thirty minute interval pupillary accommodation to convergence was just detectable but delayed over about 10 seconds; but there was no light reflex and at the end of the sixty minute interval this reflex was doubtful. Twelve hours after the strychnine injection his convergence accommodation was present but minimal and so was his light reflex but not the consensual photomotor reflex. Thirty-six hours later there were delayed convergence accommodation, consensual photomotor reflex and light reflex, each taking about 5 seconds for completion but the range of dilatation and contraction was small and there was no extreme contraction, the resting size of the pupil being a third the diameter of the cornea. Ten days later convergence accommodation was apparent only after 20 seconds. The light and consensual photomotor reflexes were again not apparent. The size of the pupil did not appear to vary from day to day nor was it found to be larger in the early morning than it was later in the day. Fifteen days later when again examined there was apparent but slight delayed reaction to accommodation and to light which took 5 seconds in both instances for completion of the reflex; but there was no consensual reflex apparent. The pupil with the eye at rest tended consistently to maintain a diameter of 3 mm. throughout the day and from day to day.

Examination of the nervous system disclosed the following abnormalities. The right knee jerk was absent although a weak contraction of the quadriceps muscle could be detected, only with "re-inforcement." Both ankle jerks were absent. These deep reflexes remained constant. Superficial reflexes were normal. There were no pains or cramps in the limbs nor a history of such symptoms. Both triceps jerks and the left knee jerk were normal. The Kahn reaction of the blood serum gave a negative result. In all respects the patient was a fit and healthy individual leading a perfectly normal life and there was evidence that in 1940 all his tendon reflexes were normal.

Conclusion

This case of Adie's syndrome did not present all the features of the typical syndrome, persistently. At times the delayed "tonic"

pupillary reactions were evident and at other times were not apparent or doubtful. The consistent smallness of the right pupil compared with the left and its strong tendency to maintain a constant size, at first seems to indicate a parasympathetic tonus of the sphincter pupillae; but a fact, that complete dilatation of the pupil did not follow atropine mydriasis suggests the contrary, a hypotonic state of the dilatator pupillae, so that it is reasonable to assume

- (1) that the relative smallness of the pupil was due also, to a relative hypertonicity of the sphincter pupillae over the dilatator or
- (2) because the actual size of the pupil remained moderate that there was an almost atonic nervous balance.

However, it appears that the one-time anatomic and physiologic distinctions between the sympathetic and para-sympathetic systems do not carry quite the same weight and that there is a strong tendency to name the two parts of the autonomic nervous system, acetylcholine secretory (cholinergic) and adrenaline-like substance secretory (adrenergic).

Recent evidence has also been forthcoming that the autonomic and somatic nervous systems are closely knit and as Boelke has shown, the sympathetic nerve fibres are closely associated with the fibres of voluntary muscle, and Hunter and Royle believed that sympathetic nerve fibres had much to do with muscle tone.

These endeavours, to illustrate the functional import of the sympathetic with the somatic system, together with the correlation of the functional activities of the sympathetic and parasympathetic systems, bring even stronger indications of the interdependence of the autonomic and somatic nervous systems.

It is, therefore, submitted, that the delayed "tonic" pupillary reflexes of Adie are in reality "atonic" and due to a generalised nervous asthenia, the degree of which might vary from time to time with a corresponding effect upon the pupils which are much more sensitive to such variations than are the absent or sluggish tendon reflexes. Kyrieleis Werner's procedure appears to support this.

The benign and non-syphilitic nature of the syndrome in a young and healthy individual, who is capable of leading a normal life, is confirmed and its detection appears to be a matter of chance. Its recognition, however, from the point of view of insurance companies should become established among insurance company doctors, otherwise the likelihood of a prospective policy being refused because of the absent reflexes with the usual implication, is great.

Summary

1. A case of Adie's syndrome is described.
2. Clinical tests which have at one time or another been advocated were carried out.
3. A hypothesis to explain the syndrome is submitted.
4. Its importance from the insurance companies' point of view is stressed.

REFERENCES

- Medical Annual*, 1940.
Lancet, Vol. II, p. 224, 1941.
Gray's Anatomy, 25th edition, p. 1082.

ANNOTATION

"Service" Burns

The burns inflicted on airmen, naval gunners from the back-flash in a turret, and tank crews affect the exposed parts of the body, that is the face and hands, and are characteristically encircling burns. Both the palmar and dorsal surfaces of the hands are commonly involved and so are the ears and scalp. The ophthalmologist is concerned in burns of the face where a third degree burn of the eyelids often ends in cicatricial ectropion with such complications as exposure keratitis and corneal ulceration. Those who survive by escaping within 5 seconds of the onset of the fire are not burnt in parts of the body protected by even one layer of clothing. The majority of accidental fires occur from a air-crash either shortly after the take-off or on landing. At such times it seems to be the habit of pilots to have their gloves off and their goggles pushed up on to the forehead. Gloves are put on when they are in the air and their course is set but goggles are sometimes discarded in combat. Naval gun crews are provided with an asbestos helmet and vizor which covers the scalp, face and neck leaving an aperture for the eyes, and gauntlets. It is probable that the asbestos is unnecessary and that any clothing material of reasonable thickness would give protection. Here, also, such protection is not favoured by the men and its importance unappreciated.

Much controversy has raged during the last twenty years about the treatment of burns and this subject is still under discussion. It is quite certain that the coagulants such as tannic acid should be condemned in the treatment of "service" burns of the face and hands. In the case of the face, tannic acid preparations applied