capillary forms (Robbins, 1963). Thrombosis of the cavernous spaces and any stage of organization of the thrombi may be present. Some degree of fibrosis is usual and calcification can occur as was seen in our patient on radiological examination.

Considering the pathological anatomy of a haemangioma, it is possible to surmise that red-cell fragmentation could occur in such a situation as a result of coagulation in vascular channels especially when the tumour is very extensive.

References

- ADELSON, E., HEITZMAN, E.J. & FENNESSEY, J.P. (1954) Thrombohemolytic thrombocytopenic purpura. *Archives of Internal Medicine*, **94**, 42.
- BRAIN, M.C., DACIE, J.V. & HOURIHANE, D.O'B. (1962) Micro-angiopathic haemolytic anaemia: the possible role of vascular lesions in pathogenesis. *British Journal of Haematology*, **8**, 358.
- BRAIN, M.C., ESTERLY, J.R. & BECK, E.A. (1967) Intra-vascular haemolysis with experimentally produced vascular thrombi. *British Journal of Haematology*, **13**, 868.
- BRAIN, M.C. & HOURIHANE, D.O'B. (1967) Micro-angiopathic haemolytic anaemia: the occurrence of haemolysis in experimentally produced vascular disease. *British Journal of Haematology*, **13**, 135.
- BULL, B.S., RUBENBERG, M.L., DACIE, J.V. & BRAIN, M.C. (1968) Microangiopathic haemolytic anaemia: mechanism of red-cell fragmentation *in vitro* studies. *British Journal of Haematology*, **14**, 643.
- LOCK, S.P. & DORMANDY, K.M. (1961) Red-cell fragmentation syndrome: a condition of multiple aetiology. *Lancet*, **i**, 1020.
- ROBBINS, S.L. (1963) *Textbook of Pathology with Clinical Application*, 2nd edn, p. 468. W. B. Saunders, Philadelphia and London.
- RUBENBERG, M.L., REGOEIZI, E., BULL, B.S., DACIE, J.V. & BRAIN, M.C. (1968) Micro-angiopathic haemolytic anaemia: the experimental production of haemolysis and red-cell fragmentation by defibrination *in vivo*. *British Journal of Haematology*, **14**, 627.
- WOOTTON, I.D.P. (1964) *Microanalysis in Medical Biochemistry*, 4th edn, p. 144. J. & A. Churchill, London.
- WRIGHT, G.P. & SYMMERS, W.St.C. (1967) *Systemic Pathology*, 1st edn, Vol. 1, p. 664. American Elsevier Publishing Company, Inc., New York.

Postgraduate Medical Journal (February 1972) **48**, 113–115.

Autonomic dysfunction in syringomyelia

M. J. AMINOFF
B.Sc., M.B., M.R.C.P.

C. S. WILCOX
M.A., B.M., M.R.C.P.

Department of Neurology, The Middlesex Hospital, London, W.1

SINCE the clinical syndrome of postural hypotension was first described by Bradbury & Eggleston in 1925, numerous reports have appeared in the literature recording the association of this syndrome with disease of the nervous or endocrine systems. In this report, we describe a patient who developed autonomic dysfunction, with postural hypotension, in association with syringomyelia.

Case report

Mrs A.T., a 56-year-old housewife with syringomyelia was admitted to the Middlesex Hospital (No. G61625) under the care of Professor R. W. Gilliatt in April 1970, complaining of increasing weakness of the left leg.

The family history was non-contributory. The patient first noticed slight swelling and flexion deformity of the fingers of both hands at the age of 14 years and 2 years later found she could not feel heat normally with her hands. These symptoms gradually progressed and she noticed bruises and blisters on her hands, especially the left, from painless injuries and burns. At 25 years of age she developed weakness of the left arm and hand. She led an active life

as a housewife until she was 50 years old, when her legs became weak. At this time she also developed urinary frequency, with nocturia, urgency, occasional urinary incontinence and less frequently faecal incontinence, without loss of bladder or rectal sensation. In the last 4 years she experienced several episodes of unsteadiness which were suggestive of postural hypotension as they occurred immediately after standing. She did not remember having sweated in hot weather for many years.

Examination revealed a dorsal kyphosis and scoliosis. There was no intellectual deficit and the cranial nerves were intact. In the arms there was wasting of the intrinsic muscles of both hands and of the left shoulder muscles, a painless arthropathy of the metacarpophalangeal joints of the right hand and clawing of the fourth and fifth digits of the left hand. Several scars from past injuries were present over both arms and hands. She had generalized weakness of the left arm with distal muscle weakness in the right arm, and loss of all tendon reflexes in the upper limbs. The abdominal muscles were weak and she was unable to sit up without support; the abdominal reflexes were absent. There was a spastic paraparesis

with hyper-reflexia and bilateral extensor plantar responses. On sensory testing, pin-prick and temperature appreciation were impaired between the fourth cervical and eighth thoracic segments on the right, and between the second cervical and eighth thoracic segments on the left, with preservation of light touch, vibration and joint position sense. In the legs there was some impairment of joint position sense at the metatarso-phalangeal joints and of vibration at the ankles. She had a lax anal sphincter.

The following *investigations* were within normal limits: full blood count and sedimentation rate; blood urea, electrolytes and sugar; and serum calcium, phosphate and alkaline phosphatase. Serological tests for syphilis and for rheumatoid disease were negative. An electrocardiogram was normal. Three MSUs were sterile on culture.

A chest X-ray was normal. X-rays of the cervical and thoracic spine showed a gross kyphoscoliosis convex to the right with some anterior wedging of the mid-dorsal vertebral bodies. Tomography of the base of the skull showed no platybasia. X-rays of the right hand revealed partial subluxation of the second and third metacarpo-phalangeal joints and of the proximal interphalangeal joints of the ring and little fingers, with new bone formation at the head of the third metacarpal; in the left hand osteoarthritic changes were present in the terminal interphalangeal joint of the middle finger.

In view of her symptoms of orthostatic hypotension, her inability to sweat and her bladder and bowel disturbances, the patient was investigated for evidence of autonomic dysfunction. With the patient supine, her blood pressure varied between 130/85 and 95/50 mmHg; on standing there was a postural drop that varied between 0 and 35 mmHg in the systolic level, and 0–25 mmHg in the diastolic. For example, on one occasion her blood pressure fell from 115/70 mmHg supine to 80/40 mmHg on standing and her pulse rate increased from 78/min to 88/min; this was accompanied by transient dizziness but the patient did not become pale. With continuous blood-pressure recording via an indwelling radial artery needle the patient performed a Valsalva manoeuvre as described by Sharpey-Schafer (1953). During the forced expiration there was a continual fall in blood pressure without tachycardia, so that the reflex latent period (Gross, 1970) could not be measured; at the end of the manoeuvre there was no 'overshoot' of the blood pressure or bradycardia. This type of record is seen in patients with severely impaired circulatory reflexes. The afferent limb of this reflex arc in the ninth and tenth cranial nerves cannot be tested directly (Bannister, Ardill & Fentem, 1967) but chemoreceptor fibres in these nerves were shown to be intact by the rapid intravenous injection of lobeline 2 mg; an

involuntary cough was produced after a 14-sec latent period by stimulation of the chemoreceptors (Berliner, 1940). The vasomotor response of the patient to a deep inspiration was recorded in the index fingers of both hands using a plethysmograph; a marked inspiratory vasoconstriction occurred as a spinal reflex (Gilliatt, Guttmann & Whitteridge, 1948). The patient was warmed with a radiant heat cradle (800 W) over the trunk until her sublingual temperature had risen by 2.5° F, and quinizarin powder (Guttmann, 1940) used to detect sweating. Sweating occurred in the groins, axillae, palms and soles, and over the eyebrows, but there was a complete absence of thermo-regulatory sweating over the trunk, neck and limbs. However, the intradermal injection of acetylcholine (0.1 ml of 1% solution) in both thighs and forearms produced local piloerection and sweating through an axonal reflex in functionally intact postganglionic sympathetic fibres (Coon & Rothman, 1941; Janowitz & Grossman, 1950; Barany & Cooper, 1956). An intravenous pyelogram was normal. A micturating cinecystogram showed that the bladder held 400 ml; the rise of pressure in the cystometrogram was steep and there was spontaneous leakage around the catheter, leaving approximately 150 ml. The bladder emptied at a gross pressure of 70 cm water by straining. No spontaneous detrusor contraction was seen but the outflow tract appeared normal and there was no residual or reflux. The appearances were those of a 'systolic' bladder (Bates, 1971).

Discussion

The only previous report that we have found in the literature of an association between orthostatic hypotension and syringomyelia is by Ellis & Haynes, 1936, and their patient clearly had co-existent syringobulbia. Although they reached no definite conclusions about the site of the lesion responsible for the vasomotor disturbance, these authors suggested that it might be in the hypothalamus.

This patient with syringomyelia and symptoms of autonomic dysfunction was found to have postural hypotension, impairment of circulatory reflexes and thermo-regulatory sweating, and a 'systolic' bladder. The functional integrity of autonomic afferent fibres and of sympathetic ganglia and postganglionic fibres has been demonstrated by the normal evoked responses to intravenous lobeline and intradermal acetylcholine respectively. This implies that the autonomic nervous system is affected at an intermediate point. An abnormality at foramen magnum level, such as that which is frequently associated with syringomyelia (Gardner, 1965), may have interrupted descending autonomic fibres above the syrinx in the upper cervical cord. Alternatively, clinical assessment indicates that the patient has

a syrinx extending from the second and fourth cervical to the eighth thoracic segments and it may be presumed that this is involving the descending autonomic pathways. The medio-lateral columns or autonomic outflow paths in the spinal cord may also be affected, in which case the absence of Horner's syndrome and the preservation of the inspiratory vaso-constrictor reflex implies that this lesion is incomplete.

Acknowledgments

We wish to thank Professor R. W. Gilliatt for permission to publish this report, and for his advice and encouragement in its preparation. We are grateful also to the Department of Radiology at the Middlesex Hospital for performing the micturating cine-cystogram on this patient.

References

- BANNISTER, R., ARDILL, L. & FENTEM, P. (1967) Defective autonomic control of blood vessels in idiopathic orthostatic hypotension. *Brain*, **90**, 725.
- BARANY, F.R. & COOPER, E.H. (1956) Pilomotor and sudomotor innervations in diabetes. *Clinical Science*, **15**, 533.
- BATES, C.P. (1971) Hunterian Lecture. *Annals of the Royal College of Surgeons of England* (in press).
- BERLINER, K. (1940) Use of alpha lobeline for measurement of velocity of blood flow. *Archives of Internal Medicine*, **65**, 896.
- BRADBURY, S. & EGGLESTON, C. (1925) Postural hypotension: A report of three cases. *American Heart Journal*, **1**, 73.
- COON, J.M. & ROTHMAN, S. (1941) Sweat response to drugs with nicotine-like action. *Journal of Pharmacology and Experimental Therapeutics*, **73**, 1.
- ELLIS, L.B. & HAYNES, F.W. (1936) Postural hypotension with particular reference to its occurrence in disease of the central nervous system. *Archives of Internal Medicine*, **58**, 773.
- GARDNER, W.J. (1965) Hydrodynamic mechanism of syringomyelia; its relationship to myelocoele. *Journal of Neurology, Neurosurgery and Psychiatry*, **28**, 247.
- GILLIATT, R.W., GUTTMANN, L. & WHITTERIDGE, D. (1948) Inspiratory vaso-constriction in patients after spinal injuries. *Journal of Physiology*, **107**, 67.
- GROSS, M. (1970) The effects of posture on subjects with cerebro-vascular disease. *Quarterly Journal of Medicine*, **39**, 485.
- GUTTMANN, L. (1940) Topographic studies of disturbances of sweat secretion after complete lesions of peripheral nerves. *Journal of Neurology and Psychiatry*, **3**, 197.
- JANOWITZ, H.D. & GROSSMAN, M.I. (1950) The response of the sweat glands to some locally acting agents in human subjects. *Journal of Investigative Dermatology*, **14**, 453.
- SHARPEY-SCHAEFER, E.P. (1953) Effects of coughing on intrathoracic pressure, arterial pressure and peripheral blood flow. *Journal of Physiology*, **122**, 351.

Postgraduate Medical Journal (February 1972) **48**, 115-117.

Addison's disease, vitiligo and multiple autoantibodies

C. J. BURNS-COX
M.B., M.R.C.P.

J. E. G. PEARSON
D.M., F.R.C.P.

Bristol General Hospital, Bristol

'In some cases the discoloration occurs in patches, or perhaps certain parts are so much darker than others as to impart to the surface a mottled or somewhat chequered appearance; and in one instance there were in the midst of this dark mottling, certain insular portions of the integument presenting a blanched or morbidly white appearance, either in consequence of these portions having remained altogether unaffected by the disease . . . or, as I believe, from an actual defect of colouring matter in parts.' Thomas Addison's description of a patient with vitiligo in his first series of patients with disease of the adrenal gland (1855).

Recently evidence has accumulated that vitiligo often accompanies autoimmune responses. The patient described below had vitiligo, alopecia areata, Addison's disease of the adrenals and thyrotoxicosis—all diseases associated with the presence of autoantibodies.

Case report

In 1959 a married English housewife aged 29 was seen because of increasing weight loss, weakness and craving for salt for 1 year with amenorrhoea for 3 months. Previously, she had noticed bald patches on her scalp which had completely disappeared without treatment.

She had not been seriously ill before and had married 5 years previously but not become pregnant. Her father had died of rheumatic heart disease and pulmonary tuberculosis, her mother had been thyrotoxic but was well as were two siblings. She appeared thin and wasted and her blood pressure was 95/70 mmHg. There was brown pigmentation of the buccal mucosa and exposed areas of her skin but there were white patches over the knuckles. There was no poliosis or premature greying of the hair.

Haemoglobin 11.3 g/100 ml, ESR 48 mm/hr, plasma Na⁺ 133, K⁺ 6.1 and Cl⁻ 100.5 mEq/l.