CASE REPORTS

COMPLETE HEART BLOCK IN MARFAN'S SYNDROME

BY

Y. S. BAWA, P. D. GUPTA, AND B. G. GOEL

From the Department of Clinical Medicine, Rajindra Hospital, Patiala, India

Marfan, in 1896, described a patient with unusually long extremities, skeletal deformities, and generalized loss of subcutaneous tissue. Since then patients showing these abnormalities and having arachnodactyly and certain other bony deformities such as high arched palate, and kyphoscoliosis have been grouped under the description of Marfan's syndrome. Marfan himself was unaware of any associated ocular or cardiovascular abnormalities and he even failed to correlate the presence of congenital cataract in one brother and strabismus in the sister of the original patient. Salle (1912) and later on Piper and Irvine-Jones (1926) stressed the association of cardiovascular abnormalities with arachnodactyly. Conduction defects have rarely been described. A case of Marfan's syndrome who took ill with Stokes-Adams attacks due to complete heart block is now presented.

Case Report

A man aged 30 years, working as a labourer for the last 15 years, was admitted on July 4, 1961, with attacks of unconsciousness. He had the first attack in June 1960, when, while sitting, he felt a tingling sensation all over the body, which was followed by unconsciousness lasting for about half a minute. A year later he had the second attack which was accompanied by convulsions. The frequency of attacks gradually increased and on the day of admission, they were recurring in quick succession. He had been complaining of breathlessness during heavy exertion for about one year preceding this illness. He was unmarried and had no brother or sister. His parents died during his childhood.

On examination, he was thinly built with long face and extremities. His weight was 120 lb. (54.4 kg.), height 5 ft. 6½ in. (166 cm.), arm span 5 ft. 10 in. (177 cm.), upper segment (vertex to pubis) 31.5 in. (80 cm.) and lower segment (pubis to heels) 35.25 in. (89 cm.). Skin was of normal texture with little subcutaneous fat. The pupils were normal and there was congenital cataract in the right eye. The palate was high arched. The fingers were long and thin, and hyperextension was present at metacarpo-phalangeal and interphalangeal joints. The feet were also long and there was congenital hallux-valgus deformity of the left foot. The spine showed scoliosis in the dorsal region with convexity to the right side and the sternum was depressed in the lower part. The blood pressure was 140/80 mm. Hg and the pulse 30 a minute with occasional extrasystoles. Venous pulsation was visible in the neck and there were occasional cannon waves. The apex beat was in the fifth interspace in the anterior axillary line. On auscultation the first sound varied in intensity, and there was a soft systolic murmur. Examination of the respiratory, alimentary, and nervous systems revealed nothing abnormal. Routine examination of blood, urine, and stools gave normal results. The fasting blood sugar was 106 mg./100 ml.; blood urea, 20 mg./100 ml.; serum calcium, 9.8 mg./100 ml., sodium, 320 mg./100 ml.; potassium, 17.8 mg./100 ml.; serum total protein, 6.5 g./100 ml.; albumin, 4.4 g./100 ml.; globulin, 2.1 g./100 ml.; albumin:globulin ratio 2:1; and V.D.R.L. and Kahn's test negative. A radiograph of the heart showed an aortic configuration with slight increase in transverse diameter and normal lung fields. A radiograph of the dorsal spine showed scoliosis with convexity to the right side. Radiographs of the pelvis and skull were normal. Metacarpal index calculated by the method of Sinclair, Kitchin, and Turner (1960) was 8.2. A cardiograph showed complete atrio-ventricular heart block, the atrial rate being 74 a minute and the ventricular rate 32 a minute. Right bundle-branch block was also present (Fig. 1).

FIG. 1.—Cardiograph shows right bundle-branch block and atrio-ventricular heart block.

The patient was given ephedrine orally, adrenaline injections, sodium phenobarbitone, and a course of heavy doses of prednisolone. He remained under observation for eight months. The attacks of unconsciousness disappeared after two months, the cardiac rhythm became regular, and the heart rate varied from 28 to 42 a minute. Electrocardiogram taken on March 20, 1962, showed no alteration.

Comments

The case reported here showed features characteristic of Marfan's syndrome. He was underweight with little subcutaneous tissue, and had a long face, with high arched palate, the extremities were long and thin with spider fingers; in the feet there was hallux-valgus deformity; the spine showed scoliosis. His arm span exceeded the height by 3.25 in. (8 cm.); the lower segment was longer than the upper segment by 3.75 in. (9.5 cm.). These two measurements, though not diagnostic, were corroborative.

In addition to these typical features the patient had a complete heart block which was obviously the cause of Stokes-Adams attacks. Conduction defects are rare in cases of Marfan's syndrome. Baer, Taussig, and Oppenheimer (1943) reported a case with prolongation of P-R interval, and another was found from the case records of Massachusetts General Hospital (1950): in these two there was no evidence of dropped beats. Marvel and Genovese (1951) reported bundle-branch block in one patient, and a similar patient was reported by Strayhorn and Wells (1948). Sinclair *et al.* (1960) reported a case of partial heart block with a ventricular rate of 30 a minute, who developed complete heart block six years later and the following year had reverted to partial heart block. We have found no record of any patient showing as severe a conduction defect as in the present instance.

For one year preceding the present illness the patient gave a history of breathlessness on exertion. It is believed that the cause of this was an associated myocarditis. Myocarditis with various

stages of heart failure in Marfan's syndrome has been reported by Whitfield, Arnott, and Stafford (1951), Tung and Liebow (1952), and Sinclair *et al.* (1960). It seems probable that in the present case the illness set in with myocarditis; consequent fibrosis later involved the conducting system, and resulted in complete heart block. However the pathological basis of conduction defect in association with Marfan's syndrome must remain a matter of conjecture at the present time.

Congenital cataract, which was present in this patient, again is an uncommon finding in cases of Marfan's syndrome. The commonest ocular anomaly reported is ectopia lentis; less frequent are tremulous iris, myopia, retinal detachment, blue sclera, iritis, cloudiness of cornea, glaucoma, small pupils and poor mydriasis due to hypoplasia of the dilator muscles.

Summary

A case of Marfan's syndrome presenting with Stokes-Adams syndrome is reported. The patient had complete heart block and right bundle-branch block. He also had a congenital cataract in the right eye. These cardiac and ocular lesions are both rare in Marfan's syndrome.

References

- Baer, R. W., Taussig, H. B., and Oppenheimer, E. H. (1943). *Bull. Johns Hopk. Hosp.*, **72**, 309.
 Marvel R. J., and Genovese, P. D. (1951). *Amer. Heart J.*, **42**, 814.
 Massachusetts General Hospital, Case Records (1950). *New Engl. J. Med.*, **243**, 346.
 Piper, R. K., and Irvine-Jones, E. (1926). *Amer. J. Dis. Child.*, **31**, 832.
 Sinclair, R. J. G., Kitchin, A. H., and Turner, R. W. D. (1960). *Quart. J. Med.*, **29**, 19.
 Salle, V. (1912). *Jb. Kinderheilk.*, **75**, 540.
 Strayhorn, D., and Wells, E. B. (1948). *Trans Amer. clin. climat. Ass.*, **59**, 205.
 Tung, H. L., and Liebow, A. A. (1952). *Lab. Invest.*, **1**, 382.
 Whitfield, A. G. W., Arnott, W. M., and Stafford, J. L. (1951). *Lancet*, **1**, 1387.