MYASTHENIA GRAVIS IN TWO BANTU CHILDREN

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Myasthenia gravis is rare in the Bantu. One case has been reported in an adult.[1] The present cases are believed to be the first reported in Bantu children.

CASE I

On 5 January 1955 a 5-year-old female Bantu child was admitted to the Baragwanath Non-European Hospital, for investigation of bilateral external ocular paresis. The mother said the child had been well until 1 month before admission, when she noticed that the child's eyelids were drooping and that the eyes were not moving freely in all directions. On direct questioning she said the eyes appeared bigger in the early morning; also that the child could play tirelessly all day. She had not noticed any dysphagia or slurring of speech. There had been no preceding trauma or illness. There was no similarly affected person in the family.

Examination. The child was well nourished and showed marked bilateral ptosis and some sagging of the lower jaw. All the external muscles of the eyes showed weakness, but the external recti were affected to the most. No exophthalmos was present. The pupils were equal and reacted to light and accommodation. The fundi were normal. There was no evidence of hyperthyroidism. Muscle fatigability could not be demonstrated clinically, owing to lack of cooperation from the small patient. As far as it was possible to assess, no diplopia was present. The tendon jerks were normal and no muscle wasting could be detected. The rest of the physical examination was negative. A test dose of 0·5 mg. of Prostigmine was given by intramuscular injection and within 15 minutes ptosis was abolished and the range of eye movements was considerably improved (Fig. 1).

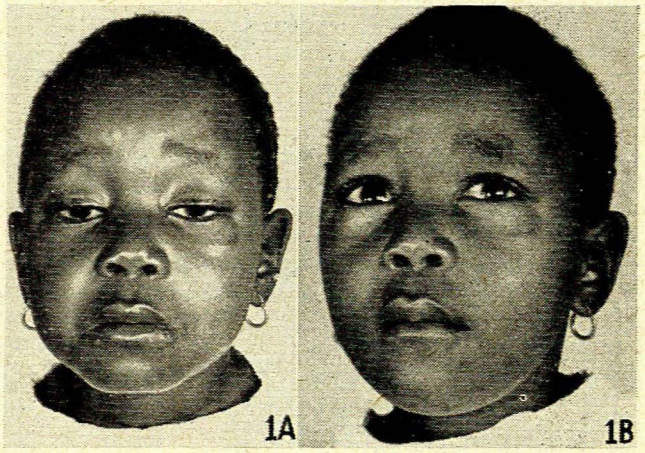

Fig. 1. (*a*) Case 1, before Prostigmine. (*b*) Case 1, 30 minutes after Prostigmine.

The urine was normal, the blood count normal, and the blood Wassermann negative. The serum potassium was 19 mg. per 100 c.c.

On X-ray of the chest the heart and lungs appeared normal, and no mediastinal mass was seen.

Treatment. Mestinon, 80 mg. by the mouth augmented by

1/8th gr. of ephedrine was given 6-hourly. The improvement which was noted within 30 minutes lasted for 3-4 hours. The child was maintained on this dosage until discharge and no side or untoward effects were noticed. At no time during the stay in hospital, was weakness noted in any other part of the body. The patient was discharged on 12 February 1955, with marked improvement of the external ocular paresis on the above dosage.

CASE 2

A 3-year-old Bantu female child was admitted on 11 October 1955 with a history of drooping of the eyelids, particularly on the left side, for 3 weeks before admission. The child was physically active and no disturbance of articulation or mastication had been noted. The parent volunteered that the weakness was maximal towards the end of the day. Before this illness, the child had

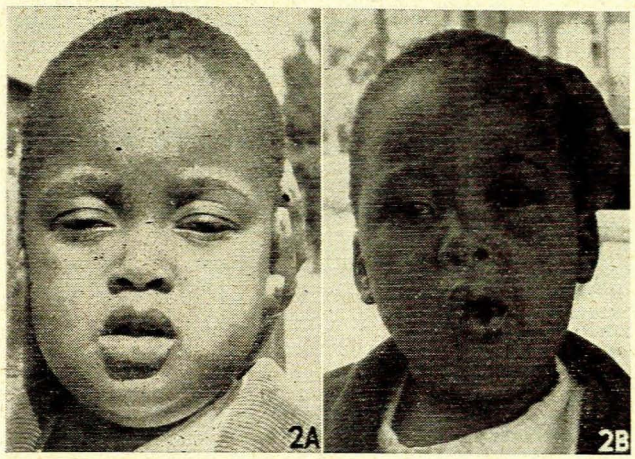

Fig. 2. (*a*) Case 2, before Prostigmine. (*b*) Case 2, 30 minutes after Prostigmine.

enjoyed good health. There was no other member of the family with a similar complaint.

Examination confirmed the ptosis, which was more marked on the left. The eye movements were limited in all directions and there was marked sagging of the lower jaw. The rest of the physical examination was normal. A test dose of 0·5 mg. of Prostigmine, together with 1/150th gr. of atropine, was administered by intramuscular injection, with a dramatic result. Ptosis of the right eye was completely abolished and that of the left eye considerably improved. The child was able to move her eyes freely in all directions and the jaw no longer drooped. Similar investigations were carried out as in case 1 and yielded no abnormal results. Muscle reactions were tested. The contractions of the orbicularis oris muscle became sluggish after 90 contractions produced by rapid faradic stimulation, which suggested a mild myasthenic reaction.

Treatment. The patient was well controlled on 15 mg. of oral Prostigmine plus 1/8th gr. of ephedrine 6-hourly. In view of the poor prognosis, it was decided to submit the child for thymectomy. The operation was performed on 7 December 1955 and the post-operative course was uneventful. On the day following operation, the Prostigmine was reduced to 7½ mg. 6-hourly, but this dose proved inadequate and, 1 week later, the original dose was reverted to. The child has required this dosage ever since. Histological section of the thymus showed this organ to be within normal limits.

DISCUSSION

In typical cases the characteristic symptoms are those of fatigue in voluntary muscles, increased by exertion. The muscles affected vary from case to case and, in a particular patient, from time to time.

The extrinsic muscles of the eyes are almost constantly affected in varying degree and may remain exclusively involved for many years. The mask-like or drowsy facies, is mainly due to the drooping eyelids and relaxation of the facial musculature. As the condition progresses, there may be generalized weakness with marked restriction of activity. In children the symptomatology is similar to that of adults. The loss of strength is not so great as in the muscular dystrophies. The patients are never so weak as to 'climb up themselves'. Remissions may occur, varying from several weeks to years. Death may result from the inhalation of food, from respiratory failure, or as the result of intercurrent infection.

Kibrick,[2] reviewing myasthenia gravis in childhood, classified cases into 2 groups:

Type 1—Transitory myasthenia of the newborn. In this group, infants born to mothers suffering from myasthenia gravis may present shortly after birth with hypotonia, inability to suck and difficulty in coping with secretions. Administration of Prostigmine may be life-saving in these cases. These infants recover completely after neonatal survival.

Type 2—True congenital myasthenia gravis. In this group, the signs of the disease may be present in infancy, but the cases differ from those in type 1, in that the onset is not as severe or as generalized. The mothers of infants in this group do not themselves suffer from the disease.

Differential Diagnosis

1. Post-diphtheritic paralysis was considered. There was no history of preceding illness. Paresis of the external ocular muscles, without any of the more common manifestations such as palatal palsy and weakness of the extremities and muscles of the neck, rendered this an unlikely diagnosis.

2. The after-effects of poliomyelitis was included in the differential diagnosis, but here again it was felt that the localization of weakness to the external ocular muscles excluded this possibility.

3. Familial periodic paralysis was considered. In this condition weakness commonly affects the proximal limb muscles and tends to occur after periods of rest. Tendon reflexes may disappear during episodes of paralysis. There is usually a familial incidence. The blood potassium is low. None of these features were present in these cases.

4. In meningo-vascular syphilis paresis is more widespread. Pupillary abnormalities, stigmata of congenital syphilis, mental deterioration, and a positive blood Wassermann, may be expected.

The typical history of increased weakness following exertion, the absence of wasting in affected muscles, with no evidence of sensory impairment, together with a dramatic response to Prostigmine, point conclusively to the diagnosis of myasthenia gravis.

I wish to thank Dr. E. Kahn and Dr. S. Wayburne, and Dr. J. D. Allen, Superintendent, Baragwanath Hospital, for permission to report these cases.

REFERENCES

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2. Kibrick, S. (1954): Pediatrics, **14**, 365.