MYOPATHIA LARYNGIS

By D. S. SARDANA (London)

INVOLVEMENT of pharyngeal musculature in myopathies is reported by various workers but weakness of laryngeal abductors due to myopathy has not been found to be mentioned even after a fairly exhaustive search of the literature.

Dysphagia is mentioned in association with dystrophia myotonica by Thomson and Negus. Ludman also records dysphagia in dystrophia myotonica. Walton found dysphagia in 50 per cent. cases of ocular myopathy. Leach states that generalized muscular diseases may present as pharyngeal dysphagia, palatal weakness, hoarseness and pooling of saliva in pyriform fossae. Vocal cords however, move normally. Usually these diseases are familial. He does not mention weakness of abductors of larynx. Over half a dozen cases of pseudomuscular hypertrophy were examined by the author only to find that they had no abnormality in the larynx but could manage only small boluses of food and ate very slowly.

An exhaustive report on laryngeal paralysis in children and adults by Cavanagh (1955) and later by Hollinger does not mention myopathy as a direct cause of laryngeal paralysis. In 1936, Jacod, as quoted by Cavanagh, reported a child with bilateral abductor paralysis in which no cause could be found but the mother suffered from progressive muscular atrophy.

This report deals with two sisters suffering from inspiratory dyspnœa with stridor in whom no cause could be found for the poor abduction of the vocal cords. Since both of them have some sort of generalized myopathy it seems plausible to conclude that the laryngeal abductors are also affected similarly.

The term "Myopathia Laryngis" may or may not be scientifically correct but has been used as it describes the condition fairly adequately. The entity that is described here should not be confused with myasthenia laryngis (Jackson) which means weakness of thyroarytenoideus muscle, a laryngeal adductor, as a result of vocal abuse.

Case I: Baby Gajan, 4½-year-old female, was admitted on November 18th, 1960, to Safdarjung Hospital, New Delhi. She had acute inspiratory dyspnœa and stridor for 2 days. Immediately tracheotomy was performed. Stridor had been present for 18 months and was worse at night. A younger sister was reported to have similar symptoms in a milder form. She had normal birth at full term. No history of anoxia, jaundice or convulsions during perinatal period was elicited. Cry was weak up to 5 months' age. She starting holding her head at

https://doi.org/10.1017/S0022215100064409 Published online by Cambridge University Press

8 months, sitting at 12 months, standing at $2\frac{1}{2}$ years and walking at 3 years. She had started speaking monosyllables at 1 year's age. Hearing and eyesight were normal. Examination showed a wasted child without any lymphadenopathy. Though the chest was deformed yet heart, lungs and abdomen were normal. Though stated to be $4\frac{1}{2}$ years old yet she looked only 2 years old.

Nervous System: No sensory or motor loss was detected. Reflexes were normal. Cranial nerves were normal. Marked hypotonia and wasting of the lower part of the body (Fig. 1, 2) and waddling gait was demonstrable. Lumbar lordosis was present. To stand up she climbed herself up from the sitting posture.

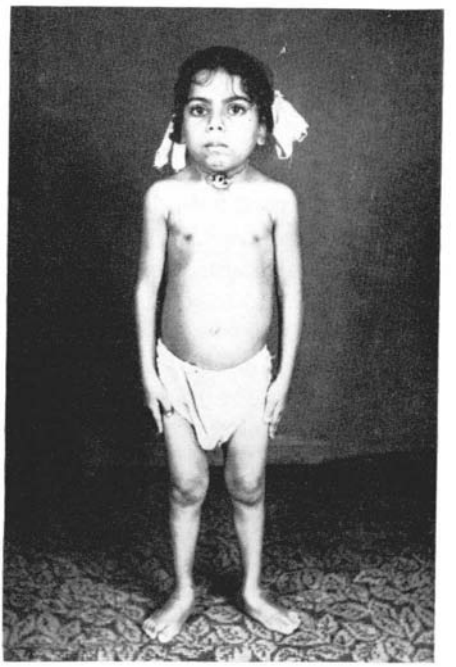


FIG. 1.
Case No. I wearing a tracheotomy tube for four years. The legs are very thin.

Investigations: X-ray of chest was normal. Blood calcium 9 mgm. per cent. ESR 32 mgm. in first hour (Wintrobe method). Mantoux test was negative. Barium swallow. No abnormality seen. Endoscopic examination of larynx, bronchial tree and œsophagus done twice did not reveal any cause for laryngeal obstruction.

On November 19th, 1960, patient was decannulated as the diagnosis was considered to be laryngismus stridulus in spite of facts apparently against this diagnosis.

On March 29th, 1961, readmitted with dyspnœa and stridor although a small tracheotomy wound remained patent due to poor healing. Bronchoscopy did not reveal any cause again. On parent's insistence tracheotomy wound was closed.

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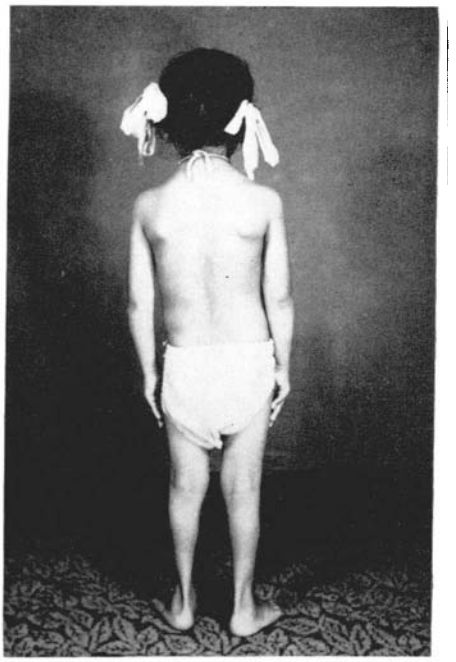


FIG. 2.
Case No. I: scapulae are prominent.

However, the patient became acutely dyspnœic within the next 24 hours and tracheotomy tube had to be reinserted.

On April 11th, 1961, direct laryngoscopy was performed without anæsthesia to study movements of the vocal cords. They lay near the midline and did not move out. This physical sign had been missed at previous examinations due to ignorance about the occurrence of this type of laryngeal paralysis in children.

On April 18th, 1961, direct laryngoscopy repeated under open ether anæsthesia. Cords adduct but do not abduct at all.

Patient was afebrile throughout and in spite of large doses of Vitamin D and calcium showed no improvement. The chest was at first wrongly considered to be ricketty. It is realized now that the deformity is due to laryngeal obstruction.

On November 15th, 1961, no improvement noticed. There was no return of abduction movement of vocal cords. The child could not be decannulated. Patient and mother had put on weight, perhaps due to restful sleep at night. Dr. A. K. Basu, pædiatrician and Dr. P. Sikand examined the child again and were of the opinion: "This is a case of myopathy. Motor milestones alone are delayed, which can be easily accounted for by myopathy. Intelligence and sensory development seem to be normal. We do not know whether laryngeal paralysis is a part of generalized muscular hypotonia." Muscle biopsy was suggested but she did not agree to submit to it.

On July 29th, 1964, the vocal cords movements did not show any sign of recovery. Muscle biopsy taken from peroneal compartment of left leg lacks

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features of striated muscle and resembles fibro-fatty tissue consistent with diagnosis of myopathy (Figs. 5 and 6). She cannot tolerate partial blocking of the tube for more than 10-15 minutes. Thus there has been no return of function in 4 years.

Case II: Krishna, younger sister of Case I, aged 3 years and 3 months, was seen on January 16th, 1962, after great persuasion as prior to that the parents would brush away our request by saying "cure baby Gajjain first and then we will show you Krishna".

She was delivered normally at full term. Second stage was a bit prolonged. She did not cry well after birth and was quite limp at birth. She did not suck well for 48 hours after birth.

She started holding her head up at 6-7 months. She sat at 1 year. She started standing with support at 2 years. She does not walk as yet. Speech appeared at 7 months, only as monosyllables, although hearing was quite normal.

Examination: Revealed generalized hypotonia. There was some drooping of right upper eyelid. Slight internal squint of right eye was noticed. Nervous system: cranial nerves were normal; motor power was found diminished in all muscles (Figs. 3, 4). There was no loss of sensation. All reflexes were normal. Heart, lungs and abdomen were normal. Dr. A. K. Basu's impression was "it seems to be a case of myopathy but anoxia neonatorum may be an ætiological factor—electroencephalogram may be helpful".

Skiagram chest and wrists were reported normal. Soon after the pædiatric examination the child was examined by me. She had stridor, but little dyspnœa, of 2 years duration which was worse during sleep, crying and eating. She coughed and vomited quite frequently. She had had treatment for talipes in Irwin Hospital, New Delhi by being immobilized in plaster of Paris and now wears special shoes. Another elder sister (10 years old) and a brother 6 years old are quite healthy. The first two were delivered in the home while the last two (the diseased ones) were delivered after prolonged labour in hospital. No surgical interference was done. Both were small sized infants at birth.

Direct laryngoscopy under open ether revealed midline position of the vocal cords with poor abduction. Re-examined on July 29th, 1964, there is neither any improvement nor deterioration in the laryngeal and general condition.

Follow Up

Both children have been followed up to July 29th, 1964. Both show muscular hypotonia. Their difficulties in standing and walking persist. Case I still cannot be decannulated due to midline position of vocal cords. Case II still has stridor but the dyspnœa is so mild that she does not require any tracheotomy.

Comments

It has been impossible to find any local cause for weakness of abductors. Perhaps the associated myopathy is the cause. It has not been possible to carry out electromyography, electroencephalography, or serum aldolase, or serum creatine kinase estimations which help in confirming the diagnosis

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FIG. 3.
Lower limbs of the younger sister are very thin, practically devoid of muscles.

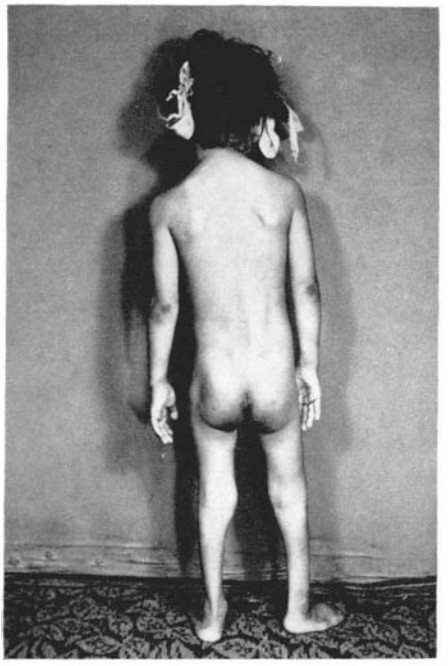


FIG. 4.
Case No. II: scapulae are prominent.

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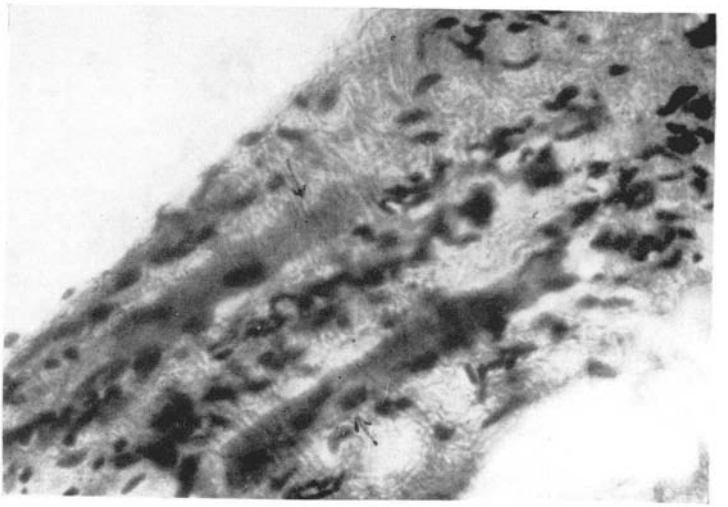


FIG. 5.
Microphotography of the peroneal muscles from Case No. 1 showing two muscle fibres with early loss of striations and fraying. There is proliferation of nuclei and replacement by loose connective tissue in-between. (H. and E. × 270.)

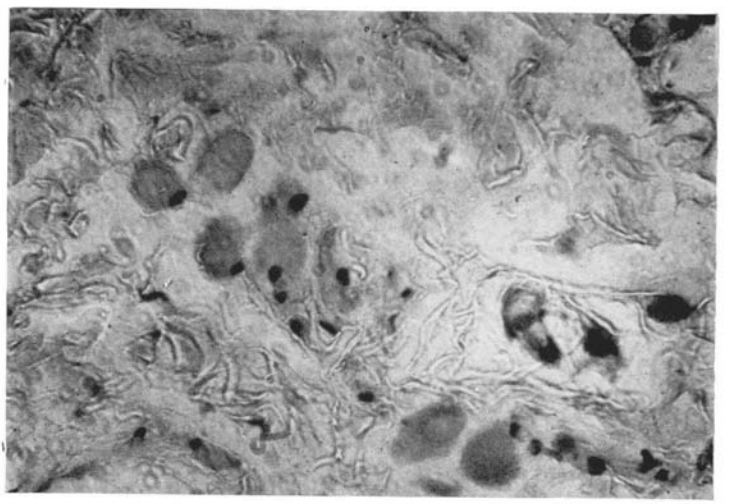


FIG. 6.
Microphotograph of the peroneal muscle from a different site from Case No. 1 showing almost complete replacement by fibrofatty tissue. A few atrophied transversely cut muscle fibres can be seen. (H. and E. × 270.)

of the myopathies. The first two investigations are of equivocal value in some types. Muscle biopsy findings are similar to those found in myopathies. As the tissue has been taken from peroneal muscles and not laryngeal muscles it is not absolutely confirmatory. Recently Walton has suggested

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a new classification of the muscular dystrophies and emphasized the difficulties faced in such a task. Our case could be classified under the heading: congenital muscular dystrophy. The younger child also presents a squint and thus presents some features of ocular myopathy. What part anoxia at birth has played in the second infant is again hard to decide. About 30 per cent. cases of laryngeal paralysis evade ætiological diagnosis in spite of extensive investigations. Could it be that some of them suffer from a localized form of myopathy?

Treatment also presents problems. Operations designed to abduct the vocal cords in the elder sister will only lead to worsening of the voice. Also in view of her young age tracheotomy tube with expiratory valve is preferable at present.

For the younger sister we have to await some form of medical treatment. At present there is no known satisfactory treatment.

Summary

Case reports of two sisters suffering from laryngeal obstruction due to generalized myopathy affecting the laryngeal abductors are presented.

Acknowledgments

My grateful thanks are due to Dr. A. K. Basu, D.C.H., and Dr. P. Sikand, M.D., M.R.C.P., for their valuable help in the diagnosis of these cases. Special thanks are due to Dr. K. C. Kasliwal for help in follow up. Dr. Das Gupta and Dr. K. C. Samuel, M.D., are to be thanked for the histological opinion. Col. R. D. Ayyar, Supt. Sufdarjung Hospital, New Delhi, is to be thanked for permitting me to publish these two cases.

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D. S. Sardana, M.S., F.R.C.S., D.L.O. (London),
Reader and Head of the E.N.T. Department,
G.S.V.M. Medical College,
Kanpur.
E.N.T. Dept.,
G.S.V.M. Medical College,
Kanpur,
India.

https://doi.org/10.1017/S0022215100064409 Published online by Cambridge University Press