A case of Sandifer's syndrome with hand tremor

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A 1.5-month-old boy with Sandifer's syndrome is described. After an uneventful delivery, he presented torticollis, seizure-like dystonic neck movements usually associated with feeding, episodic vomiting, inspiratory stridor and hand tremor in the first month of life. Barium esophagogram demonstrated gastroesophageal reflux, for which medical therapy was started. Children with torticollis and dystonic movements should be evaluated for Sandifer's syndrome. Early diagnosis and treatment of gastroesophageal reflux may prevent complications.

Key words: Sandifer's syndrome, hand tremor, dystonic movement.

Sandifer's syndrome, first described by Kinsbourne[1] in 1964, is a rare clinical entity which is characterized by dystonic body movements, paroxysmal torticollis and gastroesophageal reflux (GER) with or without hiatal hernia[2-4]. The mechanism of dystonic movements in Sandifer's syndrome has not been clearly understood. Treatment of GER improves symptom complex completely[5]. Identification of this syndrome is very important in order to avoid unnecessary investigations and achieve good therapeutic results. Here, we present a 1.5-month-old boy with Sandifer's syndrome who displayed recurrent vomiting, torticollis, paroxysmal dystonic movements and hand tremor.

Case Report

A 1.5-month-old boy, the first child of first-degree consanguineous parents, was referred for evaluation of torticollis. His birth weight was 3,500 g. He had history of mild hypoxia during delivery after an uneventful pregnancy. He had suffered for one month from recurrent vomiting and associated intermittent movements like bending backward, tilting the head to lateral and back side and shaking in his hands. Parents reported these movements occurring immediately after feeding. He was conscious during attacks. On physical examination, his length and weight were 58 cm (50p) and 4 kg (3p), respectively. His head circumference was 38 cm (50p). Inspiratory stridor was prominent. Dystonic movements, characterized by backward curving of trunk in opustotonus position, tilt of head and coarse tremor in hands, were observed during examination, lasting 30 seconds. They were recorded by video (Fig. 1). Cervical X-rays were normal. Direct laryngoscopy revealed normal anatomical structure of larynx, consistent with laryngomalacia. Electroencephalography record during an attack of dystonic movement was interpreted as normal. Cranial computerized

Fig. 1. (a)

Fig. 1 (a-d). Sequential images of a dystonic movement.

Fig. 1. (b)

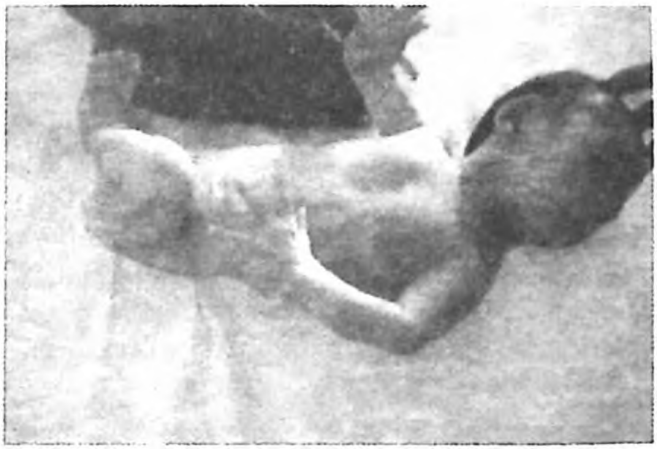

Fig. 1. (c)

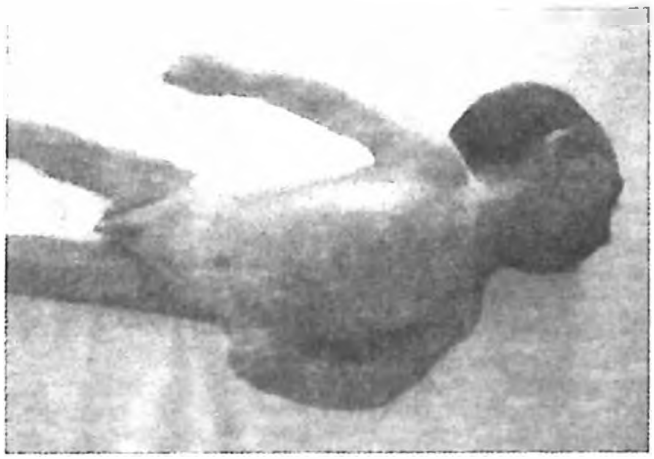

Fig. 1. (d)

tomography (CT) was normal. Biochemical investigation, including glucose, calcium and magnesium, was normal. Plasma and urinary amino acids were normal. Thyroid hormones were normal. Barium esophagogram showed significant GER (Fig. 2). We started at first with cisapride, but had no opportunity to follow-up. We later learned that he had died as a result of an aspiration episode.

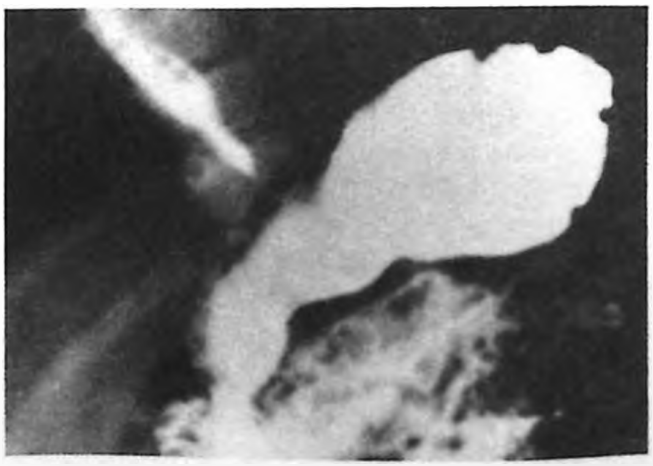

Fig. 2. Barium esophagogram revealing gastroesophageal reflux.

Discussion

Sandifer's syndrome is more common than generally recognized[4]. It is difficult to estimate the real frequency of the syndrome, as misdiagnosis may occur due to variable clinical presentation and features overlapping with other diseases[4,6]. Both normal and mentally retarded infants manifest Sandifer's syndrome. Neurological manifestations of Sandifer's syndrome in children with brain damage or metabolic disease may be falsely attributed to their basic disorder.

Gastroesophageal reflux could play a role in the pathogenesis of Sandifer's syndrome. However, most children with GER do not develop abnormal posture, and the connection between reflux and dystonia has been a matter for speculation[7,8]. Some authors postulated that children with this syndrome have an increased esophageal sensitivity to refluxed gastric contents so that even minimal exposure to acid may initiate the symptom complex[4]. It has been suggested that the movements in some way

bring about symptomatic relief[1,5]. Manometric study of a patient with Sandifer's syndrome showed low amplitude and slow propagation of esophageal peristalsis. And, dystonic posturing was shown to produce an increase in the velocity and amplitude of the peristaltic waves in the esophagus, relieving the symptoms by promoting the clearance of acid from the lower esophagus[7]. Subsequently, esophageal dysmotility was again found as the most frequent alteration in manometric studies of eight children with Sandifer's syndrome[9]. Furthermore, delayed gastric emptying was shown in a patient with Sandifer's syndrome by ultrasonography[10]. These evidence imply motility defect which may be primary or secondary to esophagitis.

Sometimes, unexplained irritability and paroxysmal movements like jerks and shaking may accompany this symptom complex[4]. Our patient also had coarse tremors in his hands. We could find no underlying cause for the tremors. It may have been an early manifestation of this association.

Medical therapy of GER generally relieves symptoms[3]. If medical therapy fails, then surgery should be performed[11,12].

We conclude that children with torticollis, dystonic body movements, unexplained jerks and hand tremor should be evaluated for Sandifer's syndrome.

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