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TORTICOLLIS WITH HIATUS HERNIA IN CHILDREN*
Sandifer Syndrome

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SUMMARY: Şenocak M.E, Arda İ.S, Büyükpamukçu N. (Department of Pediatric Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey). Torticollis with hiatus hernia in children: Sandifer syndrome. Turk J Pediatr 35: 209-213, 1993.

Sandifer syndrome is an unusual combination of gastroesophageal reflux with or without hiatal hernia and torticollis. It can be considered to be part of an expanded syndrome of unusual presentation of gastroesophageal reflux. After successful surgical repair of the gastroesophageal reflux and the hiatus hernia, there is a gradual but permanent disappearance of the torticollis. An infant with this extremely rare syndrome is presented along with a review of the related literature. *Key words: Sandifer syndrome, torticollis, gastroesophageal reflux, hiatus hernia.*

The Sandifer syndrome is an uncommon disease characterized by an unusual combination of gastroesophageal reflux (GER) with or without hiatal hernia and torticollis. Torticollis disappears completely after surgical correction of the hiatal hernia and gastroesophageal reflux.

In this paper a 5 ½-year-old girl with a history of torticollis, failure-to-thrive, vomiting, and anemia is decribed with the aim of reminding the clinician that this rare entity should be included in the differential diagnosis when investigating patients presenting with these symptoms.

Case Report

A 5 ½-year-old girl was referred to our hospital because of a history of vomiting which began when she was four-months-old, and failure-to-thrive. The vomitus was mostly curdy, but had occasionally become blood-stained. She had been vomiting at least five times a day. Physical examination revealed a pale, irritable child with a weight of 8.1 kg (below the 3rd percentile). She had a left-sided torticollis, but the head could be brought easily to a normal position (Fig. 1). No abnormalities of the sternocleidomastoid muscle were found.

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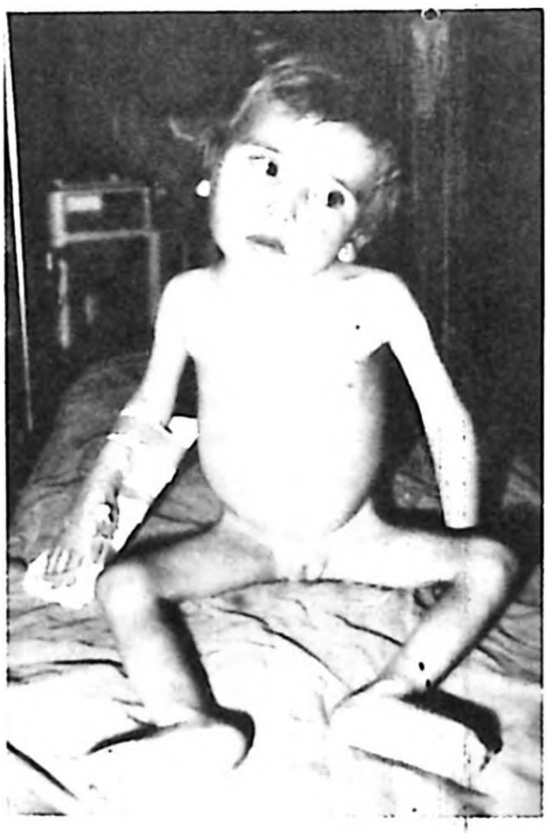

Fig. 1: Photograph showing left-sided torticollis.

The hemoglobin level was 7.3 g/dl, denoting an iron-deficiency anemia. Guaiac positive stools were noted. Roentgenograms of the cervical spine were normal and scoliosis was not present.

An upper gastrointestinal series revealed a small sliding hiatus hernia with evidence of reflux and esophagitis. The distal esophagus was moderately narrowed and irregular, suggestive of stricture (Fig. 2). Esophagoscopy at the time

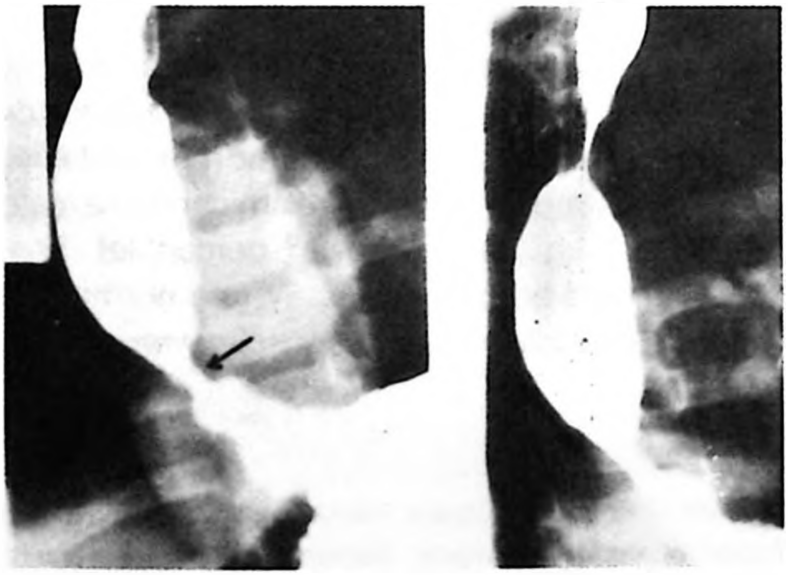

Fig. 2: Preoperative esophagogram demonstrating a small hiatus hernia with a moderately narrowed and irregular (arrow) distal esophagus.

of operation showed moderately severe esophagitis with bleeding in the lower one third of the esophagus.

At laparotomy, the entire thickness of the lower esophagus was inflamed and a small hiatal hernia was found. A repair of the hernia and a Nissen fundoplication were performed. The patient had a good recovery. There was a rapid gain in weight and the emesis ceased. Intestinal bleeding stopped post-operatively. Torticollis began within four months to gradually disappear almost completely without surgical intervention. Preoperatively the patient could only tolerate fluids, whereas at present, she heartily eats all kinds of food. A follow-up ten months later revealed that the patient was symptom-free. The barium swallow showed no evidence of hiatal herniation, GER, or esophagitis (Fig. 3). The head and neck postures were normal.

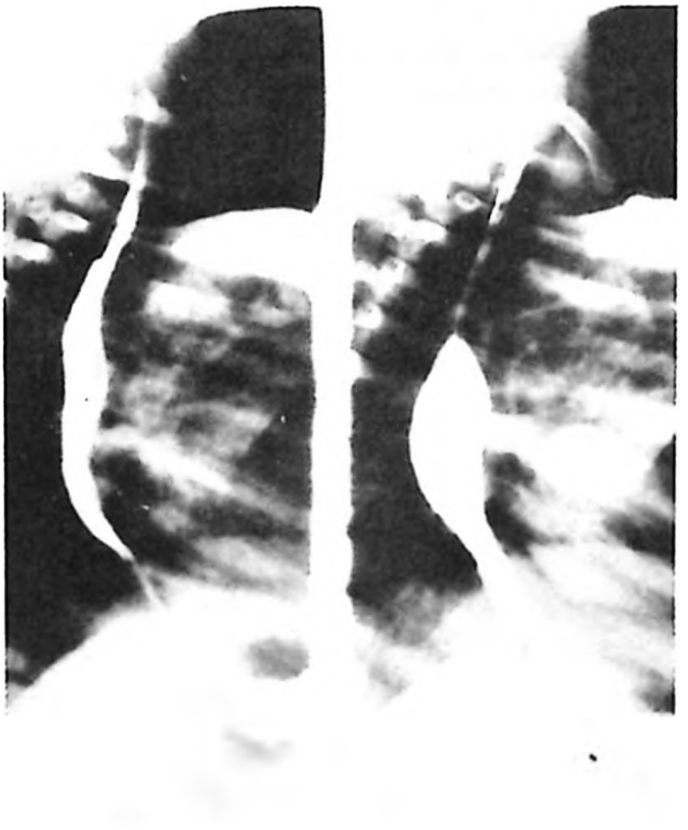

Fig. 3: Postoperative esophagogram showing normal passage of barium into the stomach without evidence of hiatus hernia or GER.

Discussion

The exact incidence of GER in the general pediatric population is unknown. The initial symptom of GER in infancy is vomiting, and Carré reported it to be present in 90 percent of his patients by the sixth week of life[1].

The recognition of the unusual association of GER, with or without hiatus hernia, with torticollis and abnormal posturing of the upper part of the trunk has been

attributed to Dr. Paul Sandifer[2]. The original description of five patients with this syndrome was by Kinsbourne in 1964[3]. Since then additional cases have been reported[2,4-8]. This exceedingly rare syndrome was seen only in one of our patients during the last twenty years.

The presence of head tilt distinguishes this syndrome from other cases of hiatal hernia with reflux esophagitis and iron deficiency anemia. The cause of torticollis is still unknown. The disappearance of torticollis after successful repair of hiatus hernia and / or GER indicates that it occurs due to an attempt to relieve the pain of esophagitis resulting from GER[8]. We believe that torticollis causes the adoption of an abnormal posture because of repeated reflexive head tilting secondary to cronically occuring GER over a long period of time.

Since torticollis is a common problem in infancy, evaluation for GER should be made if none of the other causes listed in Table I can be documented. However, the normal sternocleidomastoid, muscle a and normal cervical roentgenographic examination should alert the physician to the possibility of GER and/or hiatus hernia as the underlying causes of torticollis. An esophagogram, radionuclide studies with Tc 99m sulfur colloid in feedings, 24-hour esophageal pH monitoring and / or esophageal manometric studies are sufficient to confirm the diagnosis.

Table I: Causes of Torticollis

Abnormalities of the sternocleidomastoid muscle	(hematoma and / or fibrosis, congenital shortening)
Congenital anomalies of cervical vertebrae	(hemivertebrae, dysplasias, fusion anomalies)
Traumatic abnormalities of cervical vertebrae (rotatory subluxation)	
Inflammatory conditions of the neck (tuberculosis, rheumatoid arthritis)	
Tumors of the spinal canal or posterior fossa	
Abnormalities of extraocular muscles	
Abnormalities of the vestibular apparatus	

Most patients reported in the literature with Sandifer syndrome have been treated surgically. After repair of the hiatus hernia and the GER, the gastrointestinal bleeding ceases immediately, and the iron deficiency anemia disappears within about six weeks. As mentioned above, the torticollis gradually disappears with complete and permanent return of normal motion and appearance of the head and neck by six months.

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