

Sandifer's syndrome: a new cause

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Abnormal movements of the head and neck associated with gastro-oesophageal reflux were first described in five children by Kinsbourne in 1964.¹ Two of these patients were under the care of the neurologist Paul Sandifer, whose name the syndrome bears. Since then sporadic reports have appeared in the literature, always in children with reflux oesophagitis. We present a case of typical "Sandifer" movements caused not by reflux, but by the pain of a chronic, penetrating oesophageal foreign body.

Case report

A two-year-old boy presented urgently to the paediatric department with a six-month history of recurrent vomiting and behavioural problems. The vomiting was of undigested food, occurring shortly after meals, and though initially it had been a weekly occurrence it had increased to about once a day by the time of presentation. The child was on the 10th percentile for both height and weight. The behavioural disturbance included a change from a sunny to an irritable disposition and a tendency to roll his head from side to side and to adopt peculiar positions on the floor, with an extended or laterally flexed neck and an arched back. These postures disappeared during sleep.

There was no significant medical history, though there had been considerable parental disharmony culminating in divorce during the period before the child came to the department, and as no organic cause for his symptoms was apparent they were initially explained on the basis of emotional disturbance. During investigation, however, a barium swallow showed a star-shaped, opaque foreign body immediately behind the mid-oesophagus, with no sign of contrast hold-up or extravasation at this point (fig). He was referred to the cardiothoracic surgical unit for further management. On being questioned the mother said that she had discarded a vase a year previously, the metallic star decorations of which had begun to fall off.

Oesophagoscopy showed mucosal thickening just distal to the aortic arch, with a bead of pus expressible posteriorly. The foreign body was not visible. The mediastinum was therefore approached through a left thoracotomy and a tarnished star 1.5 cm in diameter was removed from a chronic abscess cavity behind the oesophagus, at the level of the carina. A 1 cm communication was seen between the cavity and the oesophageal lumen; it was closed directly and reinforced with a pleural flap. After operation the child developed a leak at this point, which closed in three weeks with antibiotics, intercostal tube drainage, and a feeding gastrostomy.

At the time the leak healed the abnormal postures were noted to be diminished, but they were not completely abolished until the tube was removed two weeks later. The mother confirmed that the star was one of the vase decorations. On follow-up at nine months there were no feeding difficulties, the bizarre posturing had disappeared, and the child's general behaviour was again normal.

Comment

With the original description¹ and subsequent reports²⁻⁷ 21 children have been described with the typical history of abnormal postures and movements, most commonly neck extension or lateral flexion and occasionally opisthotonus.



Barium swallow showing impacted star-shaped foreign body.

These are frequently worse immediately after meals and are always absent during sleep. All the patients had reflux oesophagitis, generally in association with a demonstrable hiatal hernia, and the age range was from 3 weeks to 14 years. A further eight patients have had either generalised body spasms or apnoeic attacks.^{4,7} Perhaps significantly, the diagnosis in this latter group has always been made in the first three months of life, and reflux has been suggested as a possible cause of sudden infant death. In the typical cases the delay between onset of symptoms and diagnosis has sometimes been quite long and in the older age group the initial diagnosis has been psychiatric disturbance.

Eleven patients underwent surgical correction while the remainder, mostly infants, were managed conservatively. In those cases where treatment abolished the oesophagitis the movements also disappeared. In surgical cases, including our own, this occurred a few weeks after the operation, presumably the time taken for the pre-existing inflammation to resolve. Of particular interest was one patient in whom breakdown of a previously successful repair after a year was followed by a return not only of the oesophagitis but also of the abnormal movements.⁴ Both again resolved after a further repair. We have been unable to find any report of similar symptoms in adults with oesophageal pain and in none of the patients reported have the postures persisted beyond the age of 14 years. There is no apparent explanation of this feature.

It has been suggested that these movements are intended to relieve the pain, but in what way they may achieve this is not certain. On a purely mechanical basis they would appear to lengthen the oesophagus, so that reduction in tension is not the answer. This case shows that any cause of oesophageal pain can induce the movements, so their effect is not to reduce reflux of acid. Indeed, screening has shown that they actually increase this.² Perhaps the answer

lies in the innervation of the area, though the connection is not obvious and the absence of movements during sleep suggests a voluntary basis. The upper third of the oesophagus is supplied by the vagus via its recurrent laryngeal branches and sympathetically from the second cervical ganglion. The lower two-thirds receive parasympathetic branches direct from the vagus in the chest and sympathetic branches from T4-6 ganglia.⁸ For the present therefore the movements evidently must await further explanation.

References

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