Hernia (2000) 4: 37-39

Hernia

Morgagni hernia in a patient with Morquio syndrome

T.Z. Nursal, M. Atli and V. Kaynaroglu

Department of General Surgery, Hacettepe University Medical School, Sihhiye, 06100, Ankara, Turkey

Summary: Morgagni hernia is a type of congenital diaphragmatic hernia which constitutes 2-6% of congenital diaphragmatic hernia cases. Morquio syndrome, a type of lysosomal hydrolase deficiency, results in keratan sulphate accumulation and skeletal dystrophy. Other anomalies such as cardiac anomalies and trisomy 21 may accompany the Morquio syndrome; however Morgagni hernia has not been previously reported with this syndrome. A patient with Morquio syndrome that presented with bilateral Morgagni hernias is described. During surgery primary repair is preferred, however when there are large defects synthetic mesh grafts may also be used.

Key words: Morgagni hernia — Congenital diaphragmatic hernia — Morquio syndrome

Correspondence to: T. Z. Nursal

Received June 24, 1999
Accepted in final form September 29, 1999

Morgagni hernia is a rare congenital retrosternal hernia which is usually detected incidentally. Congenital diaphragmatic hernia has been reported to occur with a prevalence of 3.13 per 10,000 live births [Torfs 1992] with Morgagni hernias constituting only 2.1-6% of congenital diaphragmatic hernias [Kheradpir 1988, Torfs 1992, Bragg 1996].

The Morquio syndrome, first described in 1929, is a type of familial skeletal dystrophy [Wiedemann 1992]. The disease is also known as mucopolysaccharidosis type IV in which two lysosomal hydrolases, N-acetylgalactosamine-6-sulphatase (type IV A) and β-galactosidase (type IV B) have reduced activity or are absent [Doman 1990]. The enzyme deficiencies result in impaired degradation of keratan sulphate, a cartilage constituent, resulting in retarded bone growth.

In this report, a patient with Morquio syndrome that presented with bilateral Morgagni hernias is described.

Case report

A 124 cm tall, 20 year old patient first noticed that he could not walk well at 1,5 years of age. The patient was later found to have skeletal deformities and be of short stature, and was admitted to the Hacettepe University pediatrics department at 10 years of age. Although urine excretion of keratan sulphate was within normal limits (1 mg/dl), anterior flame-shaped protrusions of the vertebral bodies were observed on the lateral spine radiograph.

During the 1.5 months before being admitted to the general surgery department with the prediagnosis of diaphragmatic hernia, the patient had recurrent respiratory infections. On initial evaluation loops of intestines were observed in the right hemithorax

(Fig. 1). This was confirmed by a computerized tomography scan (Fig. 2). The patient also had mild aortic and mitral valve regurgitation diagnosed on the echocardiogram.

The patient was operated on via an upper midline abdominal incision. There was malrotation of the intestines with the terminal ileum and caecum in a sac protruding through the right Morgagni foramen retrosternally into the right hemithorax (Fig. 3). In addition, a small left Morgagni hernia was observed on the left side, but contained no viscera. Both defects were repaired by interrupted nonabsorbable sutures. The patient made an uneventful recovery and was discharged on the 8th postoperative day.

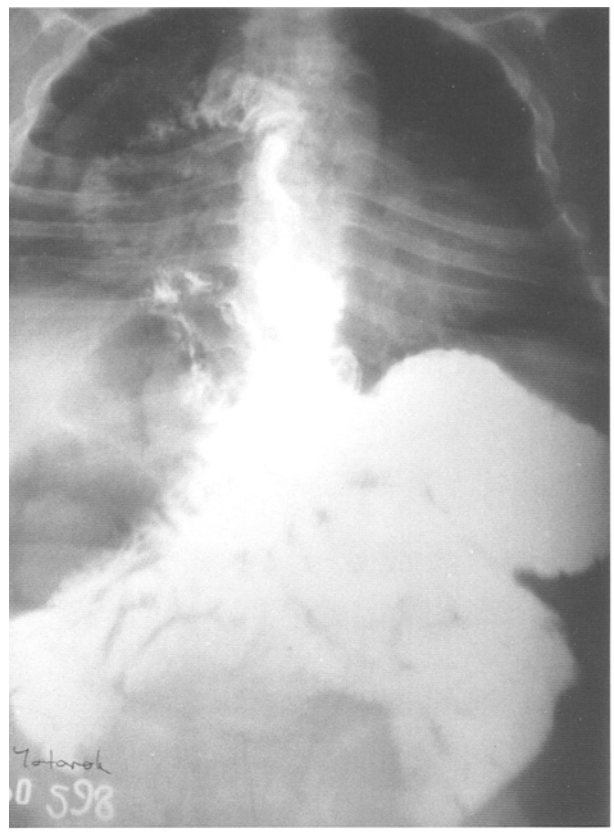

Fig. 1
Supine upper gastrointestinal system barium radiogram of the patient. A loop of intestine could be seen traversing the right hemithorax

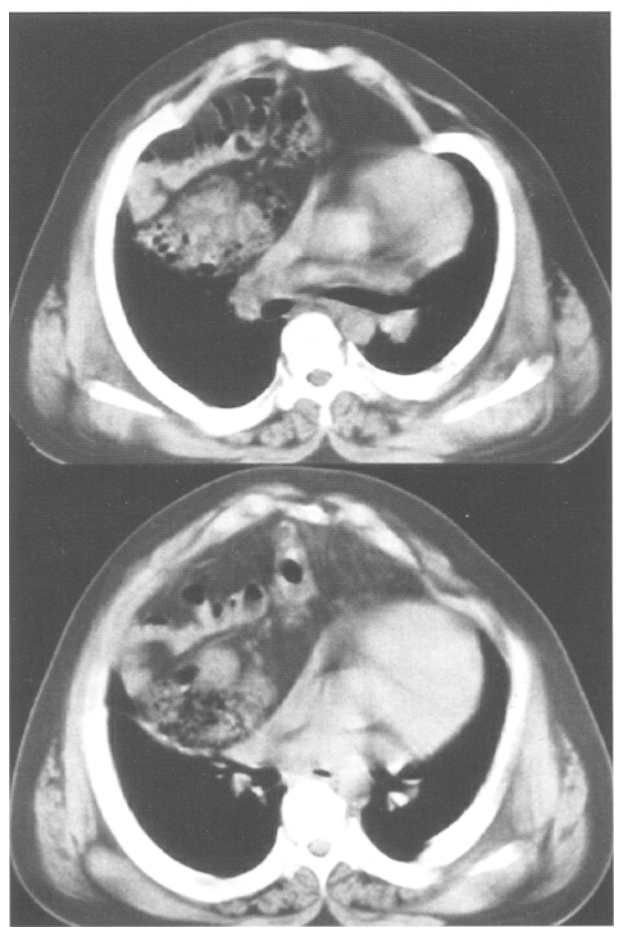

Fig. 2
Loops of intestines can be seen on the CT scan

Discussion

Morquio syndrome is a skeletal dystrophy characterized by short stature, coarse features, dysplastic teeth and corneal opacities [Doman 1990]. It is a lysosomal storage disease inherited in an autosomally recessive fashion with a resultant accumulation of keratan sulphate in cartilage which subsequently impairs the growth of bones [Scully 1991]. The excess keratan sulphate is excreted in the urine, which can assist in the diagnosis, however it has been recorded that the excretion of keratan sulphate may diminish during adolescence therefore increased excretion of keratan sulphate is not considered essential for the diagnosis [Doman 1990]. The urine keratan sulphate level was normal in our patient but the physical stigma were present. Also there were anterior flame-like protrusions of the vertebral bodies, which is almost pathognomonic for Morquio syndrome [Doman 1990]. Although there are multiple anomalies which accompany Morquio syndrome, congenital diaphragmatic hernia has not been previously reported.

Congenital diaphragmatic hernias are rare anomalies. Four distinct clinical types have been described [Torfs 1992]; 1) Anterolateral hernia 2) Posterolateral hernia 3) Pars sternalis hernia

Fig. 3
Perioperative photograph of the patient showing the defect in the right hemidiaphragm

and 4) Morgagni hernia. Posterolateral is the most common hernia type occurring in 95% of patients. The other 3 types are reported to occur with an incidence of about 2% each [Torfs 1992]. The general etiology of congenital diaphragmatic hernia is proposed to result from a fusion anomaly between different portions of the septum transversum during development. In Morgagni hernia, the proposed theory is failure of fusion of the muscular components of the diaphragm with the tendinous portion of the pars sternalis around the foramen of Morgagni (hiatus for epigastric vessels) [Torfs 1992, Steiner 1993]. Others have proposed that, due to the presence of a hernial sac with most patients, the etiology is failure of the muscular elements from cervical myotomes to reinforce the diaphragm [Kheradpir 1988].

Most Morgagni hernias are on the right side with the presence of the pericardium being proposed as a barrier against the occurrence of the rarer left sided hernia [Kimmelstiel 1987, Berman 1989]. Our patient had bilateral Morgagni hernias. Bilateral hernias were reported in 3 out of 15 patients in one series [Berman 1989]; while others have reported bilateral hernia to be the most common type [Bragg 1996]. There are multiple anomalies associated with Morgagni hernias, including cardiac defects, trisomy 21 and genitourinary anomalies [Pokorny 1984, Berman 1989], however Pokorny reported no intra-abdominal anomalies [Pokorny 1984]. In contrast, one quarter of Berman's patients had intestinal malrotation [Berman 1989]. Our patient also had malrotation of the intestines in which the caecum and terminal ileum were in the hernial sac in the right upper quadrant.

The clinical presentation is usually late in childhood in contrast to posterolateral hernias in which respiratory insufficiency and hypoplastic lung cause early recognition [Berman 1989]. Morgagni hernias are usually asymptomatic until respiratory infection or gastrointestinal disturbances appear [Fotter 1992]. The diagnosis is usually straightforward from a chest radiograph although normal chest radiographs have also been reported [Groff 1990, Fotter 1992]. An upper gastrointestinal barium series and computerized tomography are other excellent tools for diagnosis.

The best treatment is surgery, there being little morbidity and mortality [Bragg 1996]. Although repair through a thoracotomy is possible, the abdominal approach is preferable with the aim of investigating for associated abdominal anomalies and the possible presence of bilateral hernias as observed in our patient. Recently, with the advent of video-endoscopic surgery, laparoscopic and thoracoscopic Morgagni hernia repairs have been successfully performed [Newman 1995, Hussong 1997]. The suggested repair is primary closure by interrupted sutures, however in cases with large defects synthetic grafts may also be needed.

Acknowledgement. We wish to thank Dr. W. Criss for his help on preparing the English manuscript.

References

Berman L, Stringer D, Ein SH, Shandling B (1989) The late-presenting pediatric Morgagni hernia: A benign condition. J Pediat Surg 24: 970-972

Bragg WD, Bumpers H, Flynn W, Hsu HK, Hoover EL (1996) Morgagni hernias: an uncommon cause of chest masses in adults. Am Family Phys 54: 2021-2024

Doman AN, Maroteaux P, Lyne D (1990) Spondyloepiphyseal dysplasia of Maroteaux. J Bone and Joint Surg 72-A: 1364-1369

Fotter R, Schimpl G, Sorantin E, Fritz K, Landler U (1992) Delayed presentation of congenital diaphragmatic hernia. Pediat Radiol 22: 187-191

Groff DB (1990) Diagnosis of a Morgagni hernia complicated by a previous normal chest X-ray. J Pediat Surg 25: 556-557

Hussong RL Jr, Landreneau RJ, Cole FH Jr (1997) Diagnosis and repair of a Morgagni hernia with video-assisted thoracic surgery. Ann Thorac Surg 63: 1474-5

Kheradpir MH, Ahmadi J (1988). Morgagni hernias during infancy. Int Surg 73: 257-259

Kimmelstiel FM, Holgersen LO, Hilfer C (1987) Retrosternal (Morgagni) hernia with small bowel obstruction secondary to a Richter's incarceration. J Pediat Surg 22: 998-1000

Newman L 3rd, Eubanks S, Bridges WM 2nd, Lucas G (1995) Laparoscopic diagnosis and treatment of Morgagni hernia. Surg Laparosc Endosc 5: 27-31

Pokorny WJ, McGill CW, Harberg FJ (1984) Morgagni hernias during infancy: Presentation and associated anomalies. J Pediat Surg 19:394-397

Steiner Z, Mares AJ (1993) Anterolateral diaphragmatic hernia: is it a Morgagni hernia? Eur J Pediat Surg 3: 112-114

Torfs CP, Curry C, Bateson TF, Honore LH (1992) A population-based study of congenital diaphragmatic hernia. Teratology 46: 555-565

Wiedemann HR (1992) Luis Morquio. Eur J Pediat 151: 549