Case Report

Delayed presentation of bilateral Morgagni's hernia in a child with Down's Syndrome

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ABSTRACT

This case report describes the delayed presentation of bilateral Morgagni's hernia in a 13-month-old girl with Down's Syndrome. The report emphasizes the fact that a previously normal chest x-ray should not preclude the diagnosis of Morgagni's hernia even when bilateral. The various presentations and the association between Morgagni's hernia and Down's Syndrome are also discussed.

Keywords: Congenital diaphragmatic hernia, Morgagni's hernia, Down's Syndrome.

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Congenital diaphragmatic hernia through the foramen of Bochdalek is a common well known clinical entity and patients with this type of hernia are usually symptomatic immediately after birth.[1] This is in contrast to Morgagni's hernia which is rare in the pediatric age group accounting for approximately 5% of all types of congenital diaphragmatic hernia and is usually asymptomatic discovered accidentally or by production of vague gastrointestinal or respiratory symptoms.[2,3] This is a report of a case of delayed presentation of bilateral Morgagni's hernia in a child with Down's Syndrome.

Case Report. A 13-month old girl with Down's Syndrome was admitted to the hospital with the diagnosis of Morgagni's hernia. She was a product of full term normal spontaneous vaginal delivery to a 40-year-old gravida 2 para one mother. Her birth weight was 2.6kg. She was found to have features of Down's Syndrome and chromosomal analysis showed aneuploidy of 47xx + 21. She was also found to have a small umbilical hernia and congenital heart disease in the form of patent ductus arteriosus and atrial septal defect. The atrial septal defect was hemodynamically irrelevant and the patent ductus arteriosus closed spontaneously. She was asymptomatic and at the age of 3 months on routine follow-up, her chest x-ray was normal **(Figure 1a).** At 6-months of age she was admitted with cough and fever and found to have bronchopneumonia. One month later she was re-admitted to the hospital with bronchopneumonia and gastroenteritis with moderate to severe dehydration. Her chest x-ray this time showed a right paracardiac opacity **(Figure 1b)** that was not investigated. At the age of 13 months she presented to the clinic complaining of recurrent attacks of difficulty in breathing. Her chest x-ray showed a heterogenous opacity with a well demarcated upper border in the right side with gas shadows within it **(Figure 1c).** The diagnosis of Morgagni's hernia was suspected and this was confirmed by barium enema **(Figure 2).** She was operated on through an upper midline incision and found to have bilateral Morgagni's hernia. There

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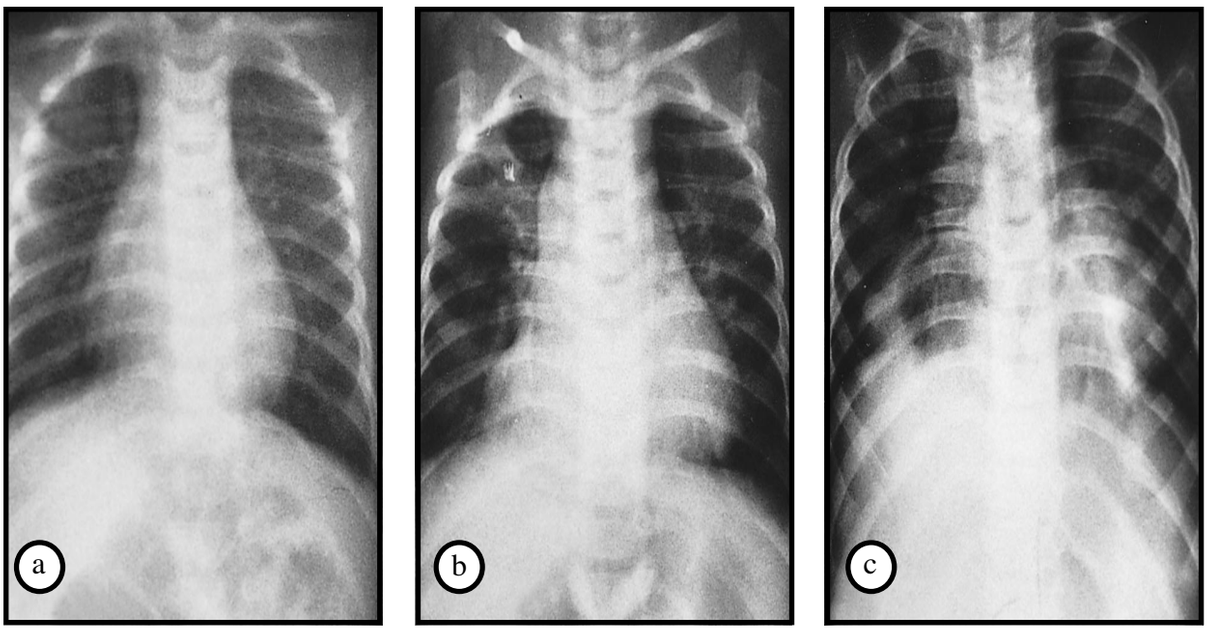

Figure 1 - Serial chest x-rays showing **a)** normal appearing chest x-ray **b)** a right paracardiac opacity **c)** a heterogenous opacity with a well demarcated upper border on the right side with gas shadow within it.

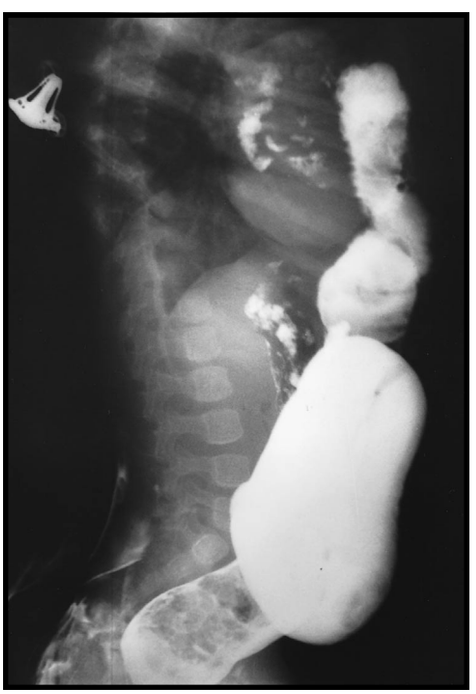

Figure 2 - Barium enema showing classic Morgagni's hernia.

were hernial sacs on both sides. The left one contained small and large bowel and the right one contained small bowel and liver. The contents were reduced and the sacs excised. Repair of both defects was carried out using silk sutures. Post-operatively, the patient did well and was discharged home on the 4th post operative day.

Discussion. Morgagni's hernia in the pediatric age group is considered to be rare when compared with other types of congenital diaphragmatic hernia representing only 1-6% of all types of congenital diaphragmatic hernias.[2] Berman et al reported only 18 cases of Morgagni's hernia over a period of 40 years, 15 of them presented in the last 20 years.[3] Pokornay et al found only 4 (5.4%) Morgagni's hernia among 74 patients with congenital diaphragmatic hernia.[4] A suprisingly high incidence (10.5%) of Morgagni's hernia was reported recently from the Kingdom of Saudi Arabia (KSA).[5] The reason for this high incidence of Morgagni's hernia among Saudi children with congenital diaphragmatic hernia is not exactly known. A high incidence of consanguinity in this part of the world may be a contributing factor. Morgagni's hernia was reported in identical twins with Down's Syndrome which

raises the possibility that it may result from an inherited defect.[6] This is supported by the frequent association of Morgagni's hernia with other congenital defects such as Down's Syndrome.[6,7] In a collective series of 46 children with Morgagni's hernia, 16 (34.8%) of them had Down's Syndrome.[7] Although this genetic evidence is only circumstantial, the high rate of consanguinity in KSA may contribute to the increased incidence of Morgagni's hernia in our patients. Add to this, the increased awareness of this condition among pediatricians and general practitioners which probably resulted in an increase in diagnosis.

The majority of Morgagni's hernia (90%) occur on the right side, the left is affected in only 8% of patients and is bilateral in 2%.[8] The rarity on the left side is attributed to the reinforcing and protective effect of the heart and pericardium. A hernial sac is found in the majority (95%) of these patients. In the pediatric age group the presentation of children with Morgagni's hernia is variable and although acute intestinal obstruction and colonic perforation have been reported in children with Morgagni's hernia, this is a rare occurrence.[9,10] Morgagni's hernia can be symptomatic discovered accidentally during the evaluation of other non-related symptoms or it can be precipitated by trauma,[6,7,11] but during infancy Morgagni's hernia can result in severe respiratory distress.[4] Although Morgagni's hernia is a congenital defect its appearance may be precipitated by trauma.[11] Not rarely, Morgagni's hernia can present with repeated attacks of chest infection or vague gastrointestinal symptoms.[2,3,5] The vagueness and non-specific symptoms in these children are a contributing factor for delayed and misdiagnosis especially if the child is not adequately investigated. Not only this, but the diagnosis can be confusing if the hernial sac contains omentum or part of the liver as this can be confused with an anteriorly placed chest mass.[12] On the other hand, when the hernial sac contains bowel loops, the diagnosis can be made by routine chest x-ray with a lateral film to show the anterior herniation of bowel loops. This can be confirmed by barium enema or barium meal and follow through. At times the hernial sac although congenitally present may be empty and so the presence of a previously normal chest x-ray as in our patient, should not preclude a diagnosis of Morgagni's hernia. In such situations and where there is confusion regarding the diagnosis, ultrasound (US) and computerized tomography (CT) scan proved useful in establishing the diagnosis. The operative approach to Morgagni's hernia can be either transthoracic or transabdominal, but for various reasons the majority advocate the transabdominal approach.[3,5] This approach is further supported by the fact that in those with bilateral hernia such as our patient, the diagnosis of bilaterality may not be recognized pre-operatively[5] and if a transthoracic approach was adapted, one hernial side will be missed. Add to this, the fact that bowel malrotation is present in 26% of these patients which must be recognized and corrected.[3] Once diagnosed, we like others advocate repair of Morgagni's hernia.[6] This is to obviate the risk of incarceration, strangulation and to alleviate the pressure effect of the herniated viscera on pulmonary parenchyma with its subsequent sequelae.

References

1. Simson JNL, Eckstein HB. Congenital diaphragmatic hernia: a 20-year experience. *Br J Surg* 1985, 72: 733-736.
2. Cullen ML, Klein MD, Philippart AI. Congenital diaphragmatic hernia. *Surg Clin North Am* 1985; 65: 1135-1138.
3. Berman L, Stringer D, Ein SH, Shandling B. The late presenting pediatric Morgagni' hernia: a benign condition. *J Pediatr Surg* 1989; 24: 970-972.
4. Pokornay WJ, McGill CW, Herberg FJ. Morgagni hernias during infancy: presentation and associated anomalies. *J Pediatr Surg* 1984; 19: 394-397.
5. Al-Salem AH. Congenital hernia of Morgagni in children. *Annals of Saudi Medicine* 1998; 18: 260-262.
6. Harris GJ, Soper RT, Kimura KK. Foramen of Morgagni hernia in identical twins: Is this an-inheritable defect? *J Pediatr Surg* 1993; 28: 177-178.
7. Kubiak R, Platen C, Schmid E, Gruber R, Ludwig KH, Rauh W. Delayed appearance of bilateral Morgagni herniae in a child with Down's Syndrome. *Pediatr Surg Int* 1998; 13: 600-601.
8. Comer TP, Clagett OT. Surgical treatment of hernia of the foramen of Morgagni. *J Thorac Cardiovasc Surg* 1966; 52: 461-468.
9. Kimmelstiel FM, Holgersen LO, Hilfer C. Retrosternal (Morgagni) hernia with a small bowel obstruction secondary to a Richter's incarceration. *J Pediatr Surg* 1987; 22: 998-1000.
10. Cakmak O, Pektas O, Baskin D. Retrosternal hernia (Morgagni) with colonic perforation due to incarceration. *Pediatr Surg Int* 1990; 5: 274-275.
11. Al-Salem AH. Incidental bilateral Morgagni hernia in a traumatized child. *Aust N Z J Surg* 1992; 62: 910-912.
12. Al-Salem AH, Al Faraj AA, Al-Dabbous I. Morgagni hernia simulating an anterior mediastinal mass. *Annals of Saudi Medicine* 1992; 12: 226-227.