Case Report

An unusual anterior mediastinal mass in a child with B-Thalassemia major

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ABSTRACT

This case report describes the delayed appearance of Morgagni's hernia in a 5 year old child with B-Thalassemia major to present as an anterior mediastinal mass. The progressive enlargement of the liver resulted in herniation of the left lobe of the liver through the already congenitally present Morgagni's hernia leading to its enlargement. The report also emphasizes the fact that a previously normal chest x-ray should not preclude the diagnosis of Morgagni's hernia. Morgagni's hernia should also be included in the differential diagnosis of anterior mediastinal masses in children.

Keywords: Congenital diaphragmatic hernia, Morgagni's hernia, B-Thalassemia major.

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Congenital herniation through the foramen of Bochdalek is common, and patients with this type of hernia are usually symptomatic soon after birth.[1] This is in contrast to herniation through the foramen of Morgagni's which is rare in the pediatric age group accounting for about 5% of all types of congenital diaphragmatic hernias. The diagnosis in some of these patients is made accidentally on chest x-ray obtained for other reasons as they are usually asymptomatic or produce vague gastrointestinal or respiratory symptoms.[2,3] At times the diagnosis can be difficult or delayed if the hernial sac contains omentum or part of the liver confusing it with a low anteriorly placed mediastinal mass.

This is a report of a case of delayed and unusual presentation of Morgagni's hernia in a child with B-Thalassemia major. This report is the first of such an association between Morgagni's hernia and B-Thalassemia major.

Case Report. A 5 year old boy with B-Thalassemia major was admitted to the hospital because of recurrent attacks of cough and chest infection of 2 months duration. He was a product of full term normal spontaneous vaginal delivery. His birth weight was 2.1kg. He was admitted to the hospital at the age of 2 days because of sepsis, but all his cultures were negative. He was also found to have a right inguinal hernia for which he had right inguinal herniotomy at the age of 5 months. At the age of $2^1/_2$ years he was diagnosed to have B-Thalassemia major. His hemoglobin electrophoresis showed fetal hemoglobin of 99% and hemoglobin A_2 of 1%. His complete blood cell count at this time showed hemoglobin of 5.9g/dl, hematocrit of 18.5%, red blood cell count of 2.67 x 10^{12}/l, platelets of 424,000/ml and white blood cell count of 11.9 x 10^9/l. He was started on regular blood transfusions every month. At the age of 4 years, he developed autoantibodies and evidence of hypersplenism. His blood transfusion requirement increased to almost once every 10-14 days. He underwent splenectomy, following which his blood transfusions decreased to once every 6-8 weeks. At the age of 5 years he was admitted to the hospital with a 2 month history of repeated attacks of cough and chest infection. His previous chest x-ray at the age of 3 years was normal

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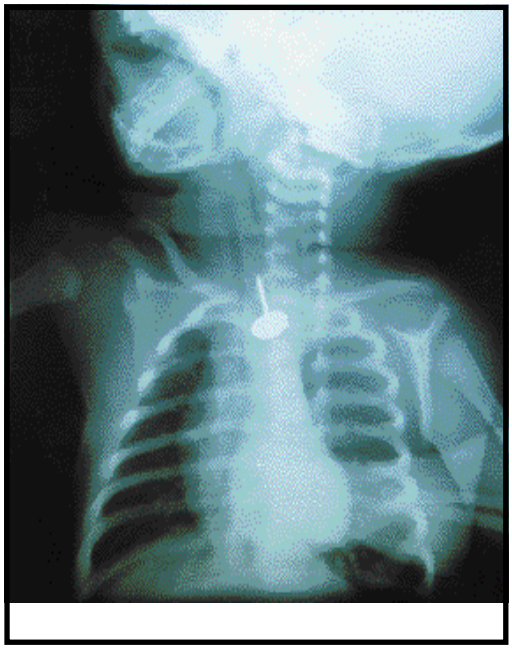

Figure 1 - Normal appearing chest x-ray.

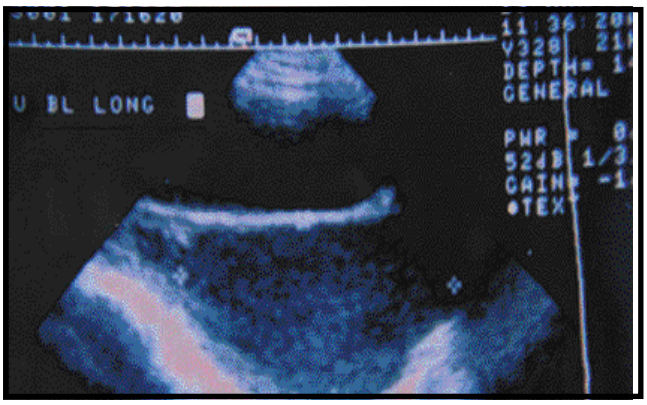

Figure 2 - Antero-posterior chest x-rays showing a well demarcated opacity in the antior mediastinum.

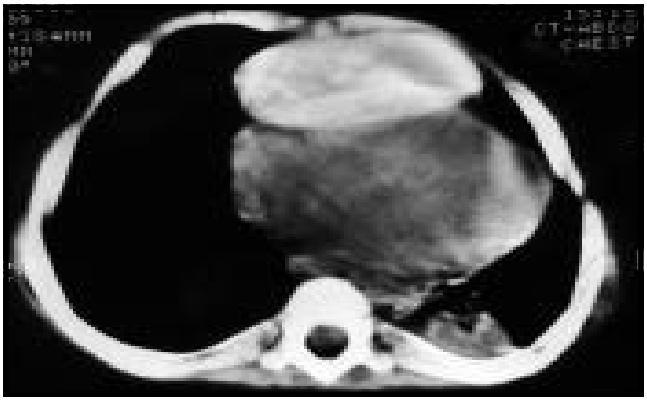

Figure 3 - CT-scan of the chest showing an anterior mediastinal mass.

(Figure 1). His anteroposterior and lateral chest x-ray (Figure 2) during this admission showed a well defined opacity in the anterior mediastinum. His abdominal ultrasound and computerized tomography (CT) scan (Figure 3) showed anterior herniation of the left lower lobe of the liver. He underwent an upper midline laparotomy which revealed a large left sided Morgagni's hernia with a hernial sac containing part of the left lobe of liver. This was reduced, the sac was excised and the defect was closed using non-absorbable sutures. Post-operatively, he did well and was discharged home on the 5th 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 about 5% 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.[2] Pokornay et al found only 4 (5%) Morgagni's hernia among 74 patients with congenital diaphragmatic hernia.[5] 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%.[6] 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. It is well known that Morgagni's hernia can be associated with other anomalies, congenital heart disease being the most common in up to 80% of children with Morgagni's hernia.[3] The association between Morgagni's hernia and Down's syndrome is well established. In a collective series of 46 children with Morgagni's hernia, 16 (35%) of them had Down's syndrome.[7] Our patient had B-Thalassemia major. Such an association between Morgagni's hernia and B-Thalassemia major to our knowledge has not been reported previously.

In the pediatric age group, Morgagni's hernia can be asymptomatic, discovered accidentally during the evaluation of other non related symptoms, but during infancy Morgagni's hernia can result in severe respiratory distress.[5] Commonly, Morgagni's hernia can present with repeated attacks of chest infection or vague gastrointestinal symptoms.[2,3,8] 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 mediastinal mass.[4] At times the hernial sac, although congenitally present, may be empty and so the presence of a previously normal chest x-ray, such as in our patient should not

preclude a diagnosis of Morgagni's hernia. In a previous report Morgagni's hernia, although congenitally present, its appearance was precipitated by a trauma in a child.[9] In our patient, who had a previous normal chest x-ray, the progressive enlargement of the liver as a result of the associated Beta-Thalassemia resulted in herniation of the left lobe of the liver through the already congenitally present Morgagni's hernia. The herniated left lobe of the liver into the chest simulated an anteriorly placed mediastinal mass with pressure effect leading to repeated attacks of cough and chest infection. Only when the hernial sac contains bowel loops can the diagnosis of Morgagni's hernia can be made by routine chest x-ray with a lateral film to show the anterior herniation of the bowel loops. This can be confirmed by barium enema or barium meal and follow-through. In situations such as in our patient, when there is confusion regarding the diagnosis, we found ultrasound and CT-scan useful in establishing the diagnosis. So, Morgagni's hernia with herniation of part of the liver should be included in the differential diagnosis of anterior mediastinal masses in children.

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. Al-Salem AH, Al Faraj AA, Al-Dabbous I. Morgagni hernia simulating an anterior mediastinal mass. Annals of Saudi Medicine 1992; 12: 226-227.
5. Pokornay WJ, McGill CW, Herberg FJ. Morgagni hernias during infancy: presentation and associated anomalies. J Pediatr Surg 1984; 19: 394-397.
6. Comer TP, Clagett OT. Surgical treatment of hernia of the foramen of Morgagni. J Thorac Cardiovasc Surg 1966; 52: 461-468.
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. Al-Salem AH. Congenital hernia of Morgagni in children. Annals of Saudi Medicine 1998; 18: 260-262.
9. Al-Salem AH. Incidental bilateral Morgagni hernia in a traumatized child. Aust NZJ Surg 1992; 62: 910-912.