CASE REPORT

A huge Morgagni hernia with compression of the right ventricle

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A 21 year old male with no relevant medical history presented to our Institution for further assessments of a right paracardiac mass founded on a chest X-ray. Chest computed tomography revealed a wide median defect of the diaphragm at the level of xiphoid process of the sternum, with the herniation of omental fat tissue in the mediastinum. Cardiac magnetic resonance confirmed the presence of a huge hernia originating from the foramen of Morgagni (sterno-costal hiatus), displacing the heart leftwards and posteriorly and compressing the right ventricle (RV), giving to it a tubular shape.The signal characteristics were typical of fat tissue, with hyperintense signal in T1 and T2 weighted black blood images and homogeneus signal suppression on STIR T2 black blood images. Short axis real time cine images, performed during deep inspiration, showed an early diastolic ventricular septal bounce, with flattening of the interventricular septum during mid-late diastole: they represented signs of diastolic dysfunction of the right ventricle, resembling a sort of "pseudo-constrictive" pathophysiological model. The patient was thus referred to surgical repair of the diaphragmatic defect.

Keywords: Morgagni hernia, Heart compression, Diastolic dysfunction, Cardiac magnetic resonance, Computed tomography

A 21-year-old man with no relevant medical history presented at our institution for further assessments of a right paracardiac mass found on chest X-ray. He complained, during the last year, of vague thoracic and abdominal discomfort with mild dyspnea during effort. Plain

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chest X-ray performed at another institution revealed right paracardiac opacity on the posteroanterior view, while the right lateral view showed opacity in the peridiaphragmatic area of the anterior mediastinum (Fig. 1A and B). We performed chest computed tomography (CT) that revealed a wide median defect of the diaphragm at the level of the xiphoid process of the sternum, with herniation of omental fat tissue in the mediastinum. It contained some linear soft-tissue opacities that represented omental vessels (Fig. 1C and D). These findings were consistent with a large Morgagni hernia (MH). The bulk shifted the heart to the left. We decided to perform cardiac magnetic resonance (CMR) to better evaluate the mass, its tissue characteristics, and the anatomical and functional relationships with the heart. CMR confirmed the presence of a large

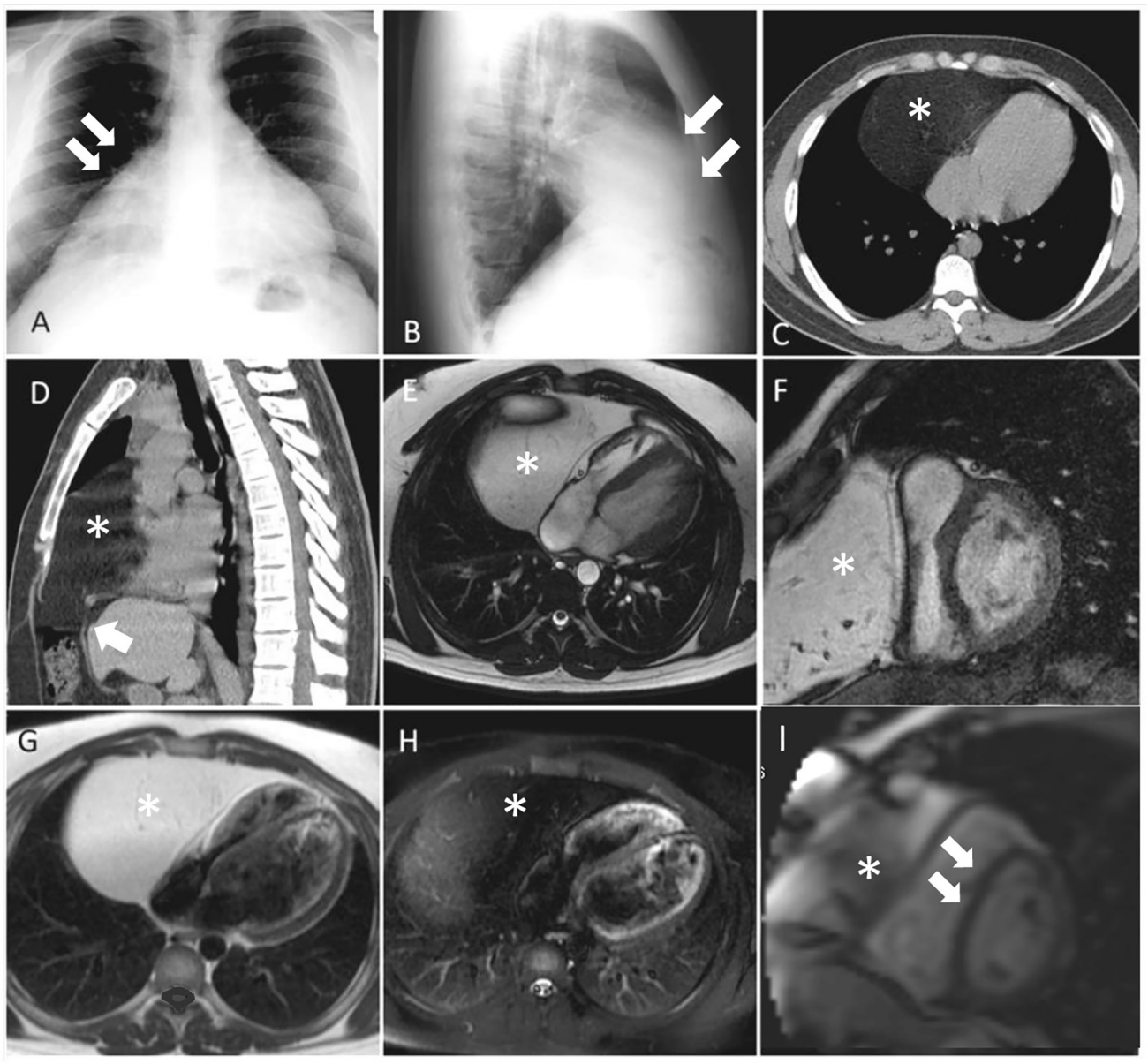

Figure 1. (A, B) Chest X-ray posteroanterior and lateral views showing a large right paracardiac opacity localized in the anterior mediastinum (arrows). (C, D) Axial and sagittal computed tomography images depicting a large diaphragmatic defect (arrow), with omental tissue in the retrosternal space (*); these are characteristics features of MH (*). (E, F) Axial and short axis steady-state free precession showing the heart dislocated leftward with compression of the right ventricle free wall from the MH (*). (G) Turbo spin echo black blood proton density axial image showing the hyperintense signal of fat tissue (*), with vascular structures inside. (H) Homogeneous signal suppression (*) on short T2 inversion recovery T2-weighted axial image. (I) Real-time cine short axis image emphasizing the pseudoconstrictive setting, with ventricular septal bounce and flattening during inspiration (arrows), compromising left ventricular filling. MH = Morgagni hernia.

MH originating from the foramen of Morgagni (sternocostal hiatus), displacing the heart leftwards and posteriorly, and compressing the right ventricle (RV), giving it a tubular shape. The effects of compression on the RV free wall were clearly showed on the steady state free precession cine images (Fig. 1E and F). The signal characteristics were typical of fat tissue: hyperintense signal in proton density and T2-weighted black blood spin echo sequences with homogeneous signal suppression on short tau inversion recovery T2 spin echo black blood images (Fig. 1G and H). Short axis real time cine images, performed during deep inspiration, showed an early diastolic ventricular septal bounce, with flattening of the interventricular septum during mid to late diastole (Fig. 1I): they represented signs of diastolic dysfunction of the RV, resembling a sort of pseudoconstrictive pathophysiological model. The patient was thus referred for surgical repair of the diaphragmatic defect. This case depicts a curious manifestation of a Morgagni diaphragmatic hernia, with relevant effects on cardiac hemodynamics. MH is the least common form of congenital diaphragmatic hernia, accounting only for 2–3% of cases; in most cases they occur on the right side (90%), while the remaining arise from the left or bilaterally [1]. The hernia typically contains omentum, followed by transverse colon and small bowel, and less frequently it contains liver and stomach [2]. MH are typically asymptomatic and incidentally discovered during unrelated examinations. When they are symptomatic, they usually present with unspecific abdominal discomfort or thoracic distress [3,4]. MH was first described in 1769 by Giovanni Battista Morgagni. He described an anterior retrosternal diaphragmatic defect between the xiphoid process of the sternum and costochondral attachments of the diaphragm [5], as result of failure of muscle tissue to spread to the area during embryologic development [6,7]. MH can be asymptomatic until adult life, or it can simulate other diseases such a large intrathoracic lipoma, or tuberculosis [8]. The most frequent features include respiratory symptoms (43%) that usually occur in lesions located on the right side (from recurrent chest infections [7] to respiratory failure [9]), gastrointestinal symptoms (gastrointestinal occlusion) (33%), or both (13%) for left-sided lesions [10,11]. It is probable that additional factors operating in later life (e.g., obesity, ascites, chronic constipation, ileus, and trauma) may result in increased intra-abdominal pressure and lead to visceral herniation through the defect and production of symptoms [10]. Surgical correction is considered mandatory because complications, such as bowel obstruction, ischemia, and bowel necrosis [12] could be life threatening. It can first occur with complications such as gastric volvulus, splenic rupture, gastric or other intestinal obstruction, and/or perforations [11]. Observation is reserved only for debilitated patients. However, because the prevalence of MH in adults has not been well reported, it is difficult to compare operative and nonoperative management. Hunter [13] described successful nonoperative management of an MH, but this management strategy failed in several cases [14]. Surgical repair is the only definitive treatment. Standard steps are: dissection of the hernia sac, reduction of the contents, and repair of the defect in the diaphragm (directly or by the use of surgical mesh). Laparotomy is the commonly used approach and allows for easier reduction of the hernia contents and evaluation of the contralateral diaphragm (for additional defects). Now, this technique has been replaced by laparoscopic repair, with the advantage of reduced surgical trauma, lower morbidity, and shorter hospital stay. Thoracotomy is a more appropriate approach to deal with recurrent cases or when there is associated intrathoracic disease requiring surgical intervention. Thoracotomy may, however, miss bilateral cases when appropriate imaging techniques are not used before surgery and will not be appropriate for repair of larger midline defects [10]. Transternal repair of the hernia is preferred in patients undergoing concomitant open heart surgery [15]. In this patient with no cardiac or intrathoracic disease a laparoscopic approach represented the best option. The patient underwent laparoscopic repair of the hernia with no complications. Early (1 week) and late (1 month) follow-up with echocardiography and CT showed restoration of normal shape and function of the RV. The patient's symptoms (effort dyspnea above all) disappeared promptly after surgery. CT and MR play a crucial role in the diagnostic evaluation of MH and in the differential diagnosis of fat-containing lesions. With their high anatomical and contrast resolution, they are useful to demonstrate the extent and content of the hernia, its anatomical location, together with the presence and width of the diaphragmatic defect. They help to differentiate a fatty omental mass from prominent epicardial fat pad, lipoma, liposarcoma, and thymolipoma [4]. CMR also has potential because it can merge anatomical and functional information and we recommend its use in every case of paracardiac mediastinal mass.

Conflict of interest

None.

References

[1] Hoyos DA. Foramen of Morgagni hernia. In: Shields TW, LoCicero J, Reed CE, Feins RH, editors. General thoracic surgery. Philadelphia: Lippincott and William Wilkins; 2009. p. 719–24.
[2] Ambrogi V, Forcella D, Gatti A, Vanni G, Mineo TC. Transthoracic repair of Morgagni's hernia: a 20-year experience from open to video-assisted approach. Surg Endosc 2007;21:587–91.
[3] Rodriguez Hermosa JI, Tuca Rodriguez F, Ruiz Feliu B, Girones Vila J, Roig Garcia J, Codina Cazador A, et al.. Diaphragmatic hernia of Morgagni-Larrey in adults: analysis of 10 cases. J Gastroenterol Hepatol 2003;26:535–40.
[4] Pineda V, Andreu J, Cáceres J, Merino X, Varona D, Domínguez-Oronoz R. Lesions of the cardiophrenic space: findings at cross-sectional imaging. Radiographics 2007;27:19–32.
[5] Anraku M, Shargall Y. Surgical conditions of the diaphragm: anatomy and physiology. Thorac Surg Clin 2009;19:419–29.
[6] Nasr A, Fecteau A. Foramen of Morgagni hernia: presentation and treatment. Thorac Surg Clin 2009;19:463–8.
[7] Al-Salem AH. Congenital hernia of Morgagni in infants and children. J Pediatr Surg 2007;42:1539–43.
[8] Pinto CS, Bernardo J, Eugénio L, Antunes MJ. Morgagni hernia mimicking intrathoracic lipomatous tumor. Rev Port Circ Cardiothorac Vasc 2013;20:135–7.
[9] Rodríguez Hermosa JI, Tuca Rodríguez F, Ruiz Feliu B, Gironès Vilà J, García Roig J, Codina Cazador A, et al.. Diaphragmatic hernia of Morgagni-Larrey in adults: analysis of 10 cases. Gastroenterol Hepatol 2003;26:535–40.
[10] Ahmad M, Al-Arifi A, Najm HK. Giant hernia of Morgagni with acute coronary syndrome: a rare case report and review of literature. Heart Lung Circ 2015;24:e144–7.
[11] Kesieme EB, Kesieme CN. Congenital diaphragmatic hernia: review of current concept in surgical management. ISRN Surg 2011;2011:974041.
[12] Pober BR, Russell MK, Ackerman KG. Congenital diaphragmatic hernia overview. In: Pagon RA, Bird TC, Dolan CR, et al., editors. GeneReviews. Seattle (WA): University of Washington, Seattle; 1993–2006, p. 469–72.
[13] Hunter WR. Herniation through the foramen of Morgagni. Br J Surg 1959;47:22–7.
[14] Horton JD, Hofmann LJ, Hetz SP. Presentation and management of Morgagni hernias in adults: a review of 298 cases. Surg Endosc 2008;22:1413–20.
[15] Nenekidis I, Anagnostakou V, Zisis C, Prokakis C, Koletsis EN, Apostolakis E, et al.. Transternal repair of a giant Morgagni hernia causing cardiac tamponade in a patient with coexisting severe aortic valve stenosis. J Cardiothorac Surg 2011;6:30.