Bilateral Bochdalek hernias associated with severe aortic tortuosity and aberrant right subclavian artery

Sinan Akay1, MD, Bilal Battalı1, MD, Kemal Kara1, MD, Ugur Bozlar1, MD

INTRODUCTION
The most common congenital diaphragmatic hernia (CDH) observed in adults is Bochdalek hernia (BH), with a prevalence of 0.17%–6%.(1,2) Although it has been described to occur more commonly on the left side of the body, other series have found a higher incidence on the right.(1,3,4) While there are many reports(2,3) on the incidence of BH, the side of the body in which BH occurs, the demographics of patients with BH, and its associated pathologies are not well described. Also, bilaterality of BH has seldom been mentioned in the literature,(1,3,4) although its incidence has been reported to be 3%–6% by Gale.(2) In this report, we describe a case of bilateral BHs, which were incidentally discovered and found to be associated with severe aortic elongation and an aberrant right subclavian artery. To the best of our knowledge, this is the first case report that suggests a link between vascular anomalies and BH.

CASE REPORT
A 27-year-old man presented to the pulmonary medicine department of our hospital with complaints of shortness of breath following exercise and claimed that he had been suffering from this distressing reaction since childhood. Chest radiography showed an increased cardiothoracic ratio, an abnormally prominent aortic knuckle, decreased volume of the left lung, and a leftward deviation of the mediastinal structures and heart. In addition, lobulated diaphragmatic contours, an obliterated left costodiaphragmatic recess and a loop formation that was superimposed on the heart shadow at the distal segment of the descending thoracic aorta, were observed (Fig. 1). Computed tomography angiography (CTA) was performed to rule out aortic aneurysm; it revealed a severely elongated thoracic aorta, with two loop formations at the level of the proximal and distal segments of the descending aorta, and an associated aberrant right subclavian artery. Bilateral BHs were also detected with CTA. The following were also observed: herniated left kidney; abdominal fat tissue that extends into the left hemithorax; and herniated segments 2 and 4A of the liver that extends into the right hemithorax through the bilateral diaphragmatic defects (Fig. 2). The pulmonary venous system, inferior and superior vena cava, and other arterial structures were normal.

1 Gulhane Military Medical Faculty, Department of Radiology, Ankara, Turkey
Correspondence Dr Sinan Akay, Consultant, Gulhane Military Medical Faculty, Department of Radiology, Etilk 06018, Ankara, Turkey akaysinan04@yahoo.com
Fig. 2 (a) Axial, (b) coronal, (c) sagittal, (d) oblique-sagittal, and maximum-intensity-projection and volume-rendered (e) coronal and (f) oblique-sagittal computed tomography angiography images demonstrate the herniating part of segments 2 and 4A of the liver (thin white arrows), herniating left kidney (white arrowheads), and a small portion of abdominal fat tissue from the bilateral diaphragmatic defects (black arrows). A severely elongated thoracic aorta, with two loop formations at the level of the proximal and distal segments of the descending aorta (thick white arrows), and an aberrant right subclavian artery (short white arrows) are also seen. S: spleen
**DISCUSSION**

BH is the leading form of CDH, with a prevalence in the adult population ranging from 0.17% to 6%.\(^{(1,2)}\) BH is usually detected in newborns, where it presents as acute respiratory distress, or later in infancy, where it presents as respiratory insufficiency.\(^{(3)}\) The patient’s prognosis is poor if the hernia is large, with mortality rates as high as 30% being the norm; death results from hypoplasia of the underlying lung and pulmonary arterial hypertension.\(^{(4)}\) However, as small hernias are usually asymptomatic, they can be missed and diagnosed only in adulthood.\(^{(5)}\) Most BH cases in adults are discovered incidentally via the use of imaging modalities during investigations for other pathologies. Such cases of BH are likely to be asymptomatic.\(^{(6)}\) Although hernias in the posterior part of the diaphragm are thought to be congenital, such hernias may also be acquired or exacerbated through trauma or physical exercise.\(^{(7)}\)

BHs can present on radiographic imaging as a focal bulge in the hemidiaphragm or a mass-like opacity adjacent to the posteromedial aspect of either hemidiaphragm. The warning signs are a typical location and a mass that has a lower density than soft tissue (due to its fat content).\(^{(8)}\) In our case, we detected an increased cardiothoracic index, decreased left lung volume, and deviation of the heart and larger mediastinal structures. No mass-like lesion was seen on the patient’s chest radiograph. The typical imaging appearance of BH is one of discontinuity of the posterior or posteromedial diaphragm, with protrusion of the peritoneal or retroperitoneal fat through the defect. Although less common, the colon, small bowel, liver, spleen, or kidneys may also herniate into the thoracic cavity.\(^{(9)}\)

Herniating organs and the discontinuity of the diaphragm can be better detected with multidetector CT imaging, due to its multiplanar reformating (MPR) and maximum-intensity-projection (MIP) capabilities. As mentioned, the diagnosis of BH for our patient was made based on the CTA results. The coronal, axial, and sagittal reformatted CTA images provided us with detailed anatomical knowledge of the diaphragmatic defects and herniated structures in our patient.

BH may be associated with various congenital anomalies, which are frequently gastrointestinal in nature. These include oesophageal duplication cysts, malrotation of the intestine, Meckel’s diverticulum, hepatic hypoplasia, and omphalocele. Pathologies that are cardiac in nature, such as patent ductus arteriosus, patent foramen ovale, ventricular septal defect, preductal aortic coarctation and tetralogy of Fallot, may also be seen. Other disorders that may coexist with BH are vesicoureteral reflux, undescended testicle, pulmonary hypoplasia, and midline defects like cleft palate and spina bifida.\(^{(10-12)}\) With the use of CTA in our case, we identified two loop formations in an elongated descending thoracic aorta and an aberrant right subclavian artery, in addition to the bilateral BHs. Although cardiovascular anomalies associated with BH have been reported,\(^{(11)}\) to the best of our knowledge, this is the first case in the medical literature that describes the occurrence of a severely elongated aorta with loop formations and an aberrant subclavian artery, and concomitant BHs. Moreover, there does not appear to be any case report in the literature regarding the concurrent occurrence of an aortic loop and an aberrant subclavian artery.

In the differential diagnosis of our patient, we considered diseases that could give rise to the aortic tortuosity seen in our patient, including arterial tortuosity syndrome and Loeys-Dietz syndrome. Both of these syndromes are connective tissue disorders similar to Marfan syndrome and in that all of them have a genetic base that is generally detected at an early age. The most striking features of these syndromes are typical craniofacial anomaly that results in a distinctive facial appearance, and elongation, aneurysm and tortuosity of the aorta and supra-aortic arteries.\(^{(11,14)}\) As our patient did not have craniofacial anomalies and was of a relatively older age, these syndromes were ruled out as causative factors.

In conclusion, BH may be associated with a number of congenital anomalies that can be symptomatic and/or highly complex, as shown in our case, in which bilateral BHs were found to be associated with a severe aortic elongation (with a double loop formation) and an aberrant right subclavian artery. In our opinion, multidetector CT and CTA with MPR and MIP imaging capabilities are reliable imaging modalities that improve the detection of BHs and any associated anomalies, whether for the purpose of diagnosis or presurgical evaluation.

**REFERENCES**