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Chest

Bifid sternum in a young woman: Multimodality imaging features

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ABSTRACT

Bifid sternum is a rare fusion anomaly of the chest wall that accounts for 0.15% of all chest deformities and may be associated with cardiac or vascular anomalies. It is usually diagnosed and surgically corrected at birth or within the first month of life. Being a diagnosis made during the neonatal period, computed tomography scan and magnetic resonance imaging are not often performed; not so many cases in literature have been studied with II level diagnostic imaging, such as computed tomography or magnetic resonance.

We describe a case of bifid sternum, rarely diagnosed in adults, discovered in a 21-year-old woman who came to our Diagnostic Imaging Department to perform a chest magnetic resonance after a chest X-ray.

Case report

A 21-year-old woman came to our Department of Diagnostic Imaging with a bony defect in the central upper part of her chest wall, which had been evident since birth and more noticeable during inspiration.

There were no alterations in laboratory tests, echocardiography, and electrocardiogram.

Chest X-ray, in the posteroanterior and latero-lateral projections, showed an abnormal radiolucent area in the upper part of the thoracic wall (Fig. 1). A general practitioner requested a magnetic resonance imaging (MRI) to rule out possibility of mediastinal pathology.

Chest MR was performed with axial and coronal planes by using a protocol that included T1-weighted turbo spin echo (repetition time/echo time: 613/8) (Figs. 2-4) and T2-weighted turbo spin echo (repetition time/echo time: 3500/100) sequences (Fig. 6). MRI showed a sternal fusion defect with an incomplete superior cleft of the manubrium and the body of the sternum, which was "U-shaped"; the right ventricle was directly in contact with the chest wall, but pulsations were not clinically evident because the bone defect was tiny.

The patient underwent a chest computed tomography (CT) (helical scan; 0.7-s rotation time; pitch 0.9; 120 kV; 200 mA; image thickness of 1.25 mm; bone plus filter) integrated with multiplanar and volume rendering reconstructions (Figs. 5 and 6). This last step confirmed the diagnosis of bifid

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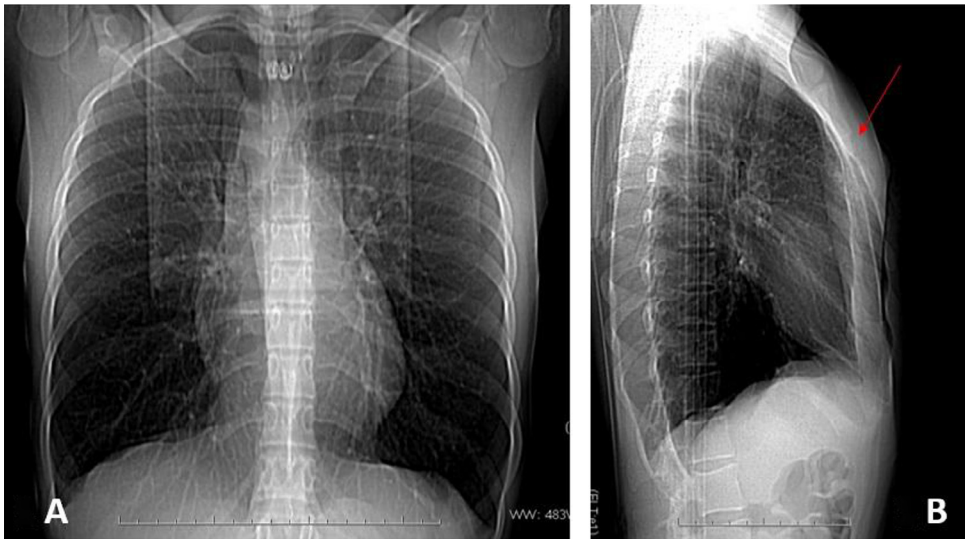

Fig. 1 – Twenty-one-year-old woman with bifid sternum. Findings: Double-projection chest X-ray. (A) Figure demonstrates no pathologic findings in posteroanterior (PA) projection. (B) Figure demonstrates an abnormal radiolucent area (red arrow) in the upper part of the thoracic wall in latero-lateral (L-L) projection. The patient shows a bony defect in the central upper part of her chest wall, more noticeable during inspiration, without alterations in the laboratory tests, echocardiography, and electrocardiogram. Technique: PA and L-L projections of chest X-ray.

sternum and, according to Ravitch classification, it was an isolated defect of the upper sternum.

The patient refused to undergo surgery after a cardiothoracic counseling, and was lost to follow-up.

Discussion

The sternum develops over a long period, with different times of maturation in each person. It begins to form during the sixth week of intrauterine life from paired mesoderm plates located in the anterior chest wall on each side of the midline; the sternal plates first chondrify, fuse with the ribs, and then undergo endochondral ossification [1]. The fusion of the ossification centers of the manubrium and sternal body (segments 1-4) starts at birth, and it is completed during the sixth year. The timing of ossification of the xiphoid (segment 5), instead, is more variable. Besides, it is possible to find an asynchronous ossification of segments 1-4, most common in the superior and inferior body, which may be an expression of a normal variant or suggestive of an

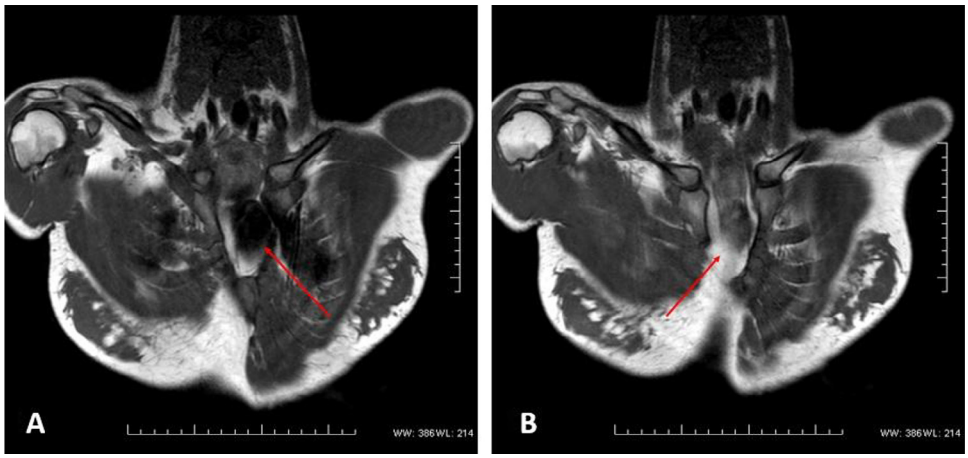

Fig. 2 – Twenty-one-year-old woman with bifid sternum. Findings: Noncontrast chest coronal magnetic resonance imaging (MRI). (A and B) Figures demonstrate the sternal fusion defect with an incomplete superior cleft of the manubrium and the body of the sternum (red arrows). This is a rare fusion anomaly of the chest wall that occurs in 0.15% of all chest deformities. It is usually diagnosed and corrected surgically at birth or within the first month of life, so it is rarely diagnosed in adulthood. Technique: 1.5 Tesla General Electric (GE) software version 15 (GE Healthcare, Milwaukee, WI); noncontrast chest coronal T1-weighted (W) turbo spin echo (TSE) (TR 613 ms, TE 8 ms, slice thickness 3 mm, skip 1 mm).

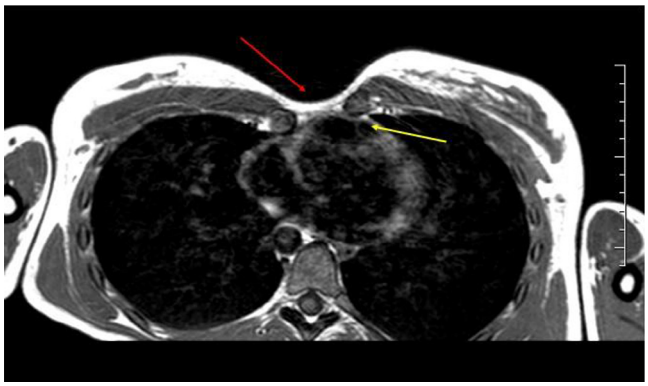

Fig. 3 – Twenty-one-year-old woman with bifid sternum. Findings: Noncontrast chest axial MRI demonstrates lack of fusion of the sternal margins (red arrow), with the right ventricle directly in contact with the chest wall (yellow arrow). The bony defect in the central upper part of the chest was evident since birth and more noticeable during inspiration, but heart pulsations were not clinically evident on the chest wall. There were no alterations in the laboratory tests, echocardiography, and electrocardiogram. Technique: 1.5 Tesla General Electric (GE) software version 15 (GE Healthcare, Milwaukee, WI); noncontrast chest axial T1-weighted (W) turbo spin echo (TSE) (TR 613 ms, TE 8 ms, slice thickness 3 mm, skip 1 mm).

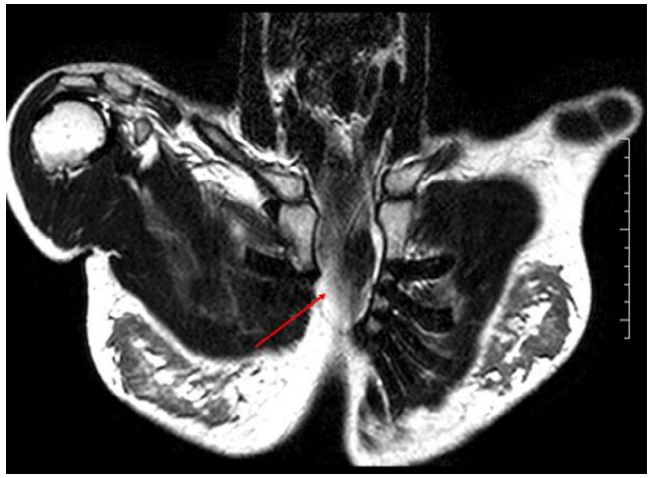

Fig. 4 – Twenty-one-year-old woman with bifid sternum. Findings: Noncontrast chest T2-weighted magnetic resonance imaging (MRI) on coronal plane shows bifid sternum with the incomplete superior cleft of the manubrium and the body of the sternum (red arrow). Technique: 1.5 Tesla General Electric (GE) software version 15 (GE Healthcare, Milwaukee, WI); noncontrast chest coronal T2-weighted (W) turbo spin echo (TSE) (TR 3500 ms, TE 100 ms, slice thickness 3 mm, skip 1 mm).

underlying disease [2]. Fusion failure results in congenital sternal clefts, constituted by a spectrum of abnormalities generally asymptomatic (74%) and associated with others defects in 72% of cases [2].

Bifid sternum is a rare fusion anomaly, and represents 0.15% of all chest deformities [3]. According to Ravitch classification of sternal clefts, there are 3 main groups of sternal anomalies depending on the associated defects: the first

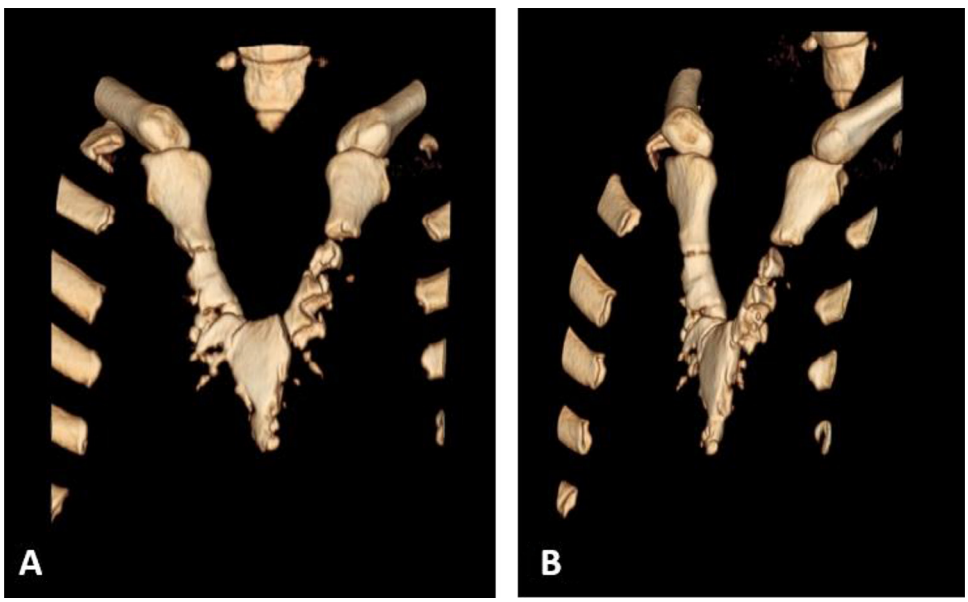

Fig. 5 – Twenty-one-year-old woman with bifid sternum. Findings: volume rendering reconstruction of noncontrast chest computed tomography (CT). (A and B) Figures show 2 views of cleft of the upper part of the sternum with "*U-shape*" variant. The superior sternal cleft may be "*V-shaped*" when the cleft reaches the xiphoid process, or "*U-shaped*," with a bony bridge joining the 2 edges ending at the third or fourth costal cartilages, as seen in this case. Technique: CT GE (General Electric Company) LIGHTSPEED volumetric computed tomography 64 slices; noncontrast CT with volume rendering reconstruction (helical scan; 0.7-s rotation time; pitch 0.9; 120 kV; 200 mA; image thickness of 1.25 mm; bone plus filter).

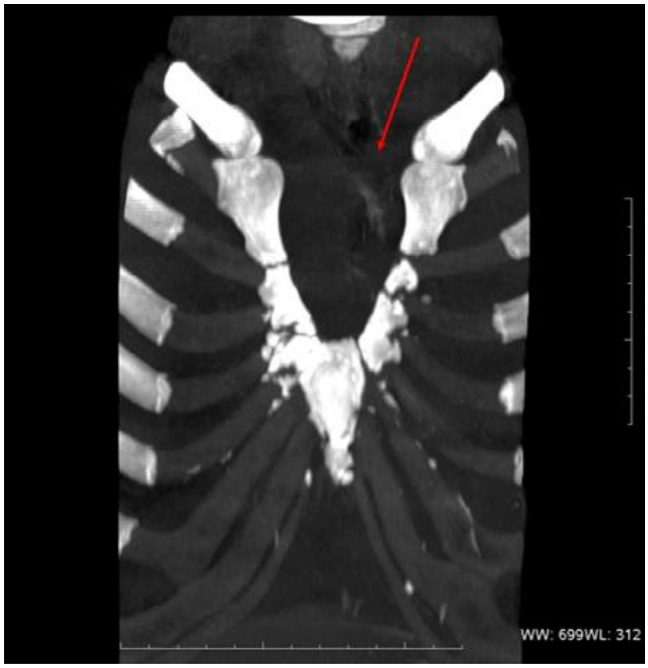

Fig. 6 – Twenty-one-year-old woman with bifid sternum. Findings: maximum intensity projection-3-dimensional reconstruction of noncontrast chest computed tomography (CT) demonstrates the cleft of the upper part of the sternum (red arrow). This is a rare fusion anomaly of the chest wall that occurs in the 0.15% of all chest deformities. Technique: CT GE (General Electric Company) LIGHTSPEED volumetric computed tomography 64 slices; non-contrast CT with maximum intensity projection-3D reconstruction (helical scan; 0.7-s rotation time; pitch 0.9; 120 kV; 200 mA; image thickness of 1.25 mm; bone plus filter).

includes isolated sternal defects, the second group is constituted by cleft sternum with abdominal wall and diaphragm malformations (ectopia cordis), and the third is known as Cantrell pentalogy. Pentalogy of Cantrell (or "thoraco-abdominal syndrome") is a rare congenital syndrome characterized by multiple malformation that affect in particular the diaphragm, the abdominal wall, pericardium, heart, and the lower sternum. The syndrome has 5 characteristic findings: sternal cleft, omphalocele, anterior diaphragmatic hernia, ectopia cordis, and a variety of intracardiac defect [4].

The cleft may involve the manubrium and upper sternebrae, the lower sternum saving the xiphoid process, but also the entire sternum. Therefore, sternal clefts are classified in complete and partial forms, which can be superior or inferior [5].

Even if the fusion of the sternum occurs craniocaudally, the most common defect is the partial superior form, which affects the upper sternum and the manubrium with normal fusion of the lower part [6]; this kind of anomaly is often isolated. The superior sternal cleft may be "V-shaped," when the cleft reaches the xiphoid process, or "U-shaped," with a bony bridge joining the 2 edges ending at the third or fourth costal cartilages [7], as seen in our case. When the cleft involves only the manubrium, with a regular body, the anomaly is called bifid manubrium. The partial cleft of the lower sternum, the third type for frequency, is usually associated with complex syndromes as Cantrell pentalogy, thoracic, and thoraco-abdominal ectopia cordis [2]. Another anomaly, which is the least common, is sternal foramen, characterized by a fusion defect in the middle portion of the sternum. When there is no bone between the hyoid and the pubis on the midline, the second type of cleft for frequency, the defect is complete [8]. Sternal aplasia is rare and incompatible with life. All patients should be evaluated to rule out any associated anomalies at the earliest, so that prognosis may be improved; besides, it is important to consider that sternal clefts determine a major exposure of the mediastinal structures and vessel to injuries.

Multidetector CT is considered the best technique for evaluating sternum anatomy and its defects, offering the possibility to perform 3-dimensional evaluation, which can help the surgeon, too. MRI can provide additional information about the bone marrow and soft tissues around the sternum by better defining their relationship. However, it cannot give a clear definition of the bone and its morphology, which is crucial for surgical repair [9]. Imaging techniques are also essential for the diagnosis of associated malformations.

In the neonatal period, the first-line treatment is the surgical correction of the defect using autologous graft [7,10,11]; however, primary closure of the bony defect is also reported in literature [12]. As the infant grows, the cartilage becomes less malleable, and the thoracic contents move anteriorly into the defect; besides, adults with a large cleft can develop respiratory symptoms with impaired gas exchange. Repairs performed after the age of 3 months have always required more supportive postoperative care, with higher incidence of cardiac complication. Beyond 1 year of age, autologous rib cartilage grafts [13] or prosthetic materials like Marlex mesh, Teflon mesh, silicon elastomer, and acrylic plate are required. Good mobilization of the 2 sternal edges is the key to a secure approximation. The risk of complication decreases when defects are repaired in the earlier age [2,14].

Differential diagnosis includes all chest anomalies that appear clinically as a depression or an asymmetry of the central-anterior thoracic wall. In particular, the most frequent sternal anomaly is pectus excavatum, which is a sternal depression that can be isolated or associated with complex syndromes and is often associated with restrictive pulmonary symptoms and cardiac functional anomalies. Depression of the central thoracic wall can be clinically evident in both pathologies; however, the diagnostic imaging findings allow for making the distinction between sternal depression (pectus excavatum) and the cleft [15]. Sternal clefts should be distinguished from pseudo-clefts, characterized by the incomplete fusion between the manubrium and the sternal body or between the body and the xiphoid process. Pseudo-clefts in fact have no clinical significance, unlike sternal clefts [16].

When the diagnosis is made in young children, non-ossified or "missing" sternal ossification centers should be considered in differential diagnosis because they may be normal variants and not a sign of underlying congenital heart disease or bone dysplasia [17].

Poland syndrome appears as an asymmetric chest wall, owing to the unilateral absence of the pectoralis major and minor muscles; it can mimic sternal cleft because the affected hemithorax is hyperlucent at X-ray. CT examination shows no

Table 1 – Summary table of bifid sternum.

Etiology	• Idiopathic—Failure fusion of the ossification centers of the sternal body.
Synonyms	• Sternal cleft.
Incidence	• 0.15% of all chest deformities.
Gender ratio	• No specific ratio.
Age predilection	• Neonatal diagnosis. Few cases discovered in adult age.
Risk factors	• No definite risk factor.
Treatment	• Neonatal period: surgical correction using autologous graft. • Beyond 1 year of age: surgical correction using autologous rib cartilage grafts or prosthetic materials like Marlex mesh, Teflon mesh, silicon elastomer, and acrylic plate.
Prognosis	• Good, but if repair is done after the age of 3 months, requires more supportive postoperative care with higher incidence of cardiac complication.
Findings on diagnostic imaging	• Chest X-ray: abnormal radiolucent area in the upper part of the thoracic wall. • Chest CT-MR imaging: sternal fusion defect with an incomplete superior cleft of the manubrium and the body of the sternum, which may be "V-shaped," when the cleft reaches the xiphoid process, or "U-shaped," with a bony bridge joining the 2 edges ending at the third or fourth costal cartilages.

Table 2 – Differential diagnosis.

Disease	Diagnostic clues
Pectus excavatum	Sternal depression, which differs from the incomplete fusion on the midline.
Pseudo-cleft	Incomplete fusion between the manubrium and the sternal body or between the body and the xiphoid process.
Asynchronous non-ossified sternal segments	Non-ossified or "missing" sternal ossification centers can be normal variants and should be distinguished from pathologic conditions.
Posttraumatic deformities	Positive history for trauma.
Tumors	Anamnestic data, together with the morphologic appearance, are suggestive for a benign or a malignant lesion. Biopsy is often required.
Poland syndrome	Asymmetry of the chest wall for unilateral absence of the pectoralis major and minor muscles; the affected hemithorax is hyperlucent at X-ray and can mimic a sternal cleft.
Genetic syndromes (camptomelic dysplasia, Noonan syndrome, trisomy 17, and 18)	Sternal clefts may occur as a large spectrum of anomalies (sternal deformity in a shield chest, short sternum with a cleft, delayed sternal ossification) associated with other musculoskeletal malformations.

sternal abnormalities but aplasia or hypoplasia of other structures of the chest (muscles, ribs, breast) [15]. Sternal fractures should be considered in differential diagnosis of sternal foramen, but they can be excluded with a clinical history negative for trauma. The latter should be distinguished also from lytic lesion in primitive bone tumor, secondary localization, or direct invasion of an adjacent tumor. The anamnestic data are essential and the morphologic appearance can be suggestive of a benign or malignant lesion; however, biopsy is often required for the diagnosis, in particular, for soft tissue masses [18]. Even different primary rib anomalies may cause a deformity of the chest wall; however, an attentive clinical examination is sufficient for a differential diagnosis. Sternal defects are also frequent in rare genetic syndromes, such as Noonan syndrome, trisomy 17, and trisomy 18, in which they occur as a large spectrum of anomalies (sternal deformity in a shield chest, short sternum with a cleft, delayed sternal ossification) associated with other musculoskeletal malformations [17,19,20] (Tables 1 and 2).

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