Case Report

Report of an incidental finding of a congenital intradiaphragmatic cyst in an adult and a critical review of the literature

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Abstract

An incidental chest x-ray finding of an oval soft tissue opacity in the right costophrenic recess in a 55-year-old man prompted further investigation by unenhanced CT which demonstrated a 3.9 cm diameter lesion of fluid density intimately related to the lateral aspect of the right hemidiaphragm and right lobe of the liver. It was not possible on CT to determine whether this was intrapulmonary, an unusual exophytic hepatic simple cyst, or a diaphragmatic cystic abnormality. Subsequent ultrasound, however, allowed the diagnosis of a diaphragmatic mesothelial cyst by confirming an anechoic, thin-walled cystic structure separate from the liver capsule, from which it moved independently, with a fixed relationship to the diaphragm and splitting of muscular fibres of the right hemidiaphragm around its periphery. This case serves to highlight a rare congenital abnormality of the hemidiaphragm as a cause of a pulmonary nodule visible on a chest radiograph and to illustrate the usefulness of ultrasound in confirming the diagnosis. It also allows a critical review of the literature on intradiaphragmatic mesothelial cysts in adults and children.

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Case report

A 55-year-old male was referred by his general practitioner for chest radiography because of mild exertional dyspnoea following an upper respiratory tract infection. The chest radiograph (Fig. 1) demonstrated a subtle ovoid opacity projected over the right lateral costophrenic recess adjacent to the right hemidiaphragm. There was no previous imaging available at the time of reporting to see if this was a new or long-standing abnormality. A subsequent lateral chest radiograph (Fig. 1) showed this lesion as an oval density projected
Fig. 1 – PA and left lateral chest radiographs demonstrate a subtle ovoid opacity (arrowed) projected over the right lateral hemidiaphragm.

Fig. 2 – Axial (a), coronal (b) and sagittal (c) unenhanced CT images in both lung and soft tissue windows show a homogenous cystic lesion in the right lateral costophrenic recess. Its site of origin is unclear.

over the cardiac silhouette with its long axis lying vertically, which was inseparable from the silhouette of the right hemidiaphragm inferiorly but was clearly defined by air-filled lung around the remainder of its circumference. An intrapulmonary lesion abutting a pleural surface was suspected and the patient was referred for an unenhanced CT which demonstrated a well-defined 36 × 28 × 39 mm oval abnormality of fluid density (mean attenuation of 1 Hounsfield Unit (HU), range from −41 to 46 HU) which appeared intimately related both to the hemidiaphragm and the capsule of the adjacent liver (Fig. 2). This subtended both acute and obtuse angles with the hemidiaphragm on both coronal and sagittal reconstructions. The former, together with the fact that more than 50% of the lesion’s circumference was surrounded by lung, suggested a pulmonary cystic lesion, whilst the latter was more consistent with it being diaphragmatic or, less likely, hepatic origin. It was not felt that CT allowed a confident diagnosis and an ultrasound study was requested. This study demonstrated a unilocular oval thin-walled anechoic cyst which was clearly separate from the liver capsule and moved with the diaphragm. Muscular fibres of the hemidiaphragm could be seen to split around the lower end of the cyst confirming its intradiaphragmatic location (Fig. 3). The lesion demonstrated no evidence of flow on colour Doppler imaging. A chest radiograph performed 4 years previously at an outside hospital, which had been reported as normal, was subsequently obtained for review. This demonstrated the same lesion of similar size (Fig. 4). A diagnosis of a mesothelial cyst of the diaphragm was made based upon these characteristic imaging findings and, in view of the longstanding nature of this lesion which had not significantly changed in size over 4 years, the patient was reassured and discharged from follow-up.

Discussion

Intrathoracic mesothelial cysts are recognized as being congenital defects thought to arise within the pleuro-peritoneal membrane during embryological development of the lung bud
Fig. 3 – Ultrasound image of right lateral thorax in a para-coronal plane clearly demonstrates it is separate from the liver capsule (arrowheads). Splitting of fibres of the right diaphragm (arrows) around the cyst confirms its intradiaphragmatic location.

Fig. 4 – Coned frontal radiograph performed 4 years previously demonstrates same ovoid soft tissue opacity in the right lateral costophrenic angle.

The literature would suggest that mesothelial cysts are much more common in children and there have been 3 case series from single centres [7–9] describing this entity in 9, 11, and 30 patients respectively. In only the first of these articles [7], however, was there histologic evidence of the mesothelial nature of the cysts as 4 children aged between 7 and 14 years were treated surgically. The authors described further 5 children aged 0–12 months who were found incidentally to have peripherally cysts during ultrasound with similar sonographic features and which they concluded, therefore, were also mesothelial. This is despite the fact that they were in a much younger age group and were all significantly smaller than those in their surgically treated patients. The authors of the subsequent paper describing 11 children with mesothelial cysts [8] based their diagnosis on imaging findings alone, namely an oval or bilobed cyst of water density with a thin wall involving the right posterolateral right hemidiaphragm. It would appear that their confidence in these characteristic imaging findings was based upon the article by Estaun et al. [7]. Five of the eleven children were treated by percutaneous aspiration and sclerotherapy with ethanol and in only one of these were mesothelial cells found in the aspirated fluid; the other 5 contained histiocytes and macrophages. Kahriman et al. [9] used these same characteristic imaging findings to make the diagnosis in 30 children in 2016. Seventeen of these 30 children were treated by percutaneous aspiration and ethanol sclerotherapy. The results of cytologic examination of the aspirated fluid were only available in 8 patients and in none of these were mesothelial cells demonstrated. Six contained neutrophils, macrophages, and lymphocytes and 2 proteinaceous fluid. It is also noteworthy that both these papers were from centres in areas where hydatid disease is endemic although important to point out that in none of those in whom aspiration was performed was there evidence of hydatid disease. Taking all these points into account, one must wonder how confident one can be that all these cysts were truly intradiaphragmatic mesothelial cysts as opposed to, for example, acquired extrahepatic cystic lesions fixed to the undersurface of the hemidiaphragm. Regardless of these criticisms, the take-home messages from these 3 articles can be summarized as follows:

- Although uncommon, mesothelial cysts of the diaphragm are more prevalent in children than previously thought;
- They have characteristic ultrasound findings that allow a confident diagnosis;
- Conservative management is recommended with follow-up ultrasound imaging which should show stability or a decrease in cyst size over time;
- and management by percutaneous aspiration and sclerotherapy should be reserved for those children in whom there is a convincing history of attributable symptoms.

There are several aspects of these 3 articles that are worthy of brief discussion:

**Side and site of diaphragmatic involvement**

The fact that no left diaphragmatic mesothelial cysts were demonstrated in any of these articles was not discussed despite the fact that left-sided mesothelial cysts are described [1,2] and most commonly occur in the right or left anterior cardiophrenic angle although they may occur elsewhere [2]. An intradiaphragmatic location is exceptionally rare and there are only a few reports in the literature most of which are descriptions of 1 or 2 patients in whom surgical resection was performed because of concerns regarding the nature of the abnormality [1,3–6]. The majority of these reports conclude that imaging cannot safely exclude a malignant lesion and that resection is mandatory. Most of these reports are more than 15 years old, however, and we would argue that current imaging might allow a more conservative approach in some individuals.
in adults. What is more, there is no embryologic reason why they should only occur on one side [1,2]. Perhaps the most likely explanation for the apparent exclusive right diaphragmatic involvement is that cysts occur with equal frequency on both sides but are more commonly visualized on the right because the hemidiaphragm on this side is easily seen during ultrasound due to its intimate relationship to the liver whilst that on the left is poorly visualized due to the adjacent stomach. An incidental cyst is much more likely, therefore, to be seen on the right. It is less easy to explain why the cysts are described in children as more commonly involving the posterolateral aspect of the hemidiaphragm as opposed to other sites particularly as this position does not appear to be more common in adults [1,2,10].

**Bilobulated shape**

Akinci et al. [8] described mesothelial cysts in 11 children all of which were “bilobulated” and suggested that this was a characteristic sign that could aid in diagnosis. It is not entirely clear why this sign, which is a common appearance of intrahepatic simple cysts, was considered by them to be helpful in making the diagnosis particularly given the fact that none of the cysts in the 9 patients reported by Estau et al. [7] exhibited this appearance. It is also not a sign that has been described as being characteristic of histologically confirmed diaphragmatic mesothelial cysts in other paediatric or adult patients in the literature. Kahriman et al. [9], however, found this a common appearance in their group in whom 26 of 30 were bilobed. This “characteristic” sign is, therefore, based upon 2 papers in neither of which was there any histologic confirmation of the diagnosis of a mesothelial cyst. Both Akinci et al. [8] and Kahriman [9] suggested that this shape is “due to the complex embryologic development of the hemidiaphragm” but did not expand upon their reason for suggesting why this might be the cause or try to explain what it is about diaphragmatic development that might account for this. If this shape is indeed a sign of an intradiaphragmatic location, we suggest that it is more simply explained by these relatively soft cysts being pinched between muscle fibres of the diaphragm.

**Is it reasonable to apply the management recommendations of these three articles regarding paediatric cases to adults?**

There has, to our knowledge, been no report of an adult with a mesothelial cyst being treated conservatively or by aspiration and sclerotherapy and the conventional teaching is that any cystic lesion involving the diaphragm in an adult patient should be resected regardless of whether it is the cause of symptoms or an incidental finding on imaging performed for another reason. This recommendation, as mentioned above, is largely based upon historical case reports, the authors of which concluded that surgical resection was essential because serious pathology could not be excluded on imaging alone. When considering whether this recommendation should now be changed it is necessary to review the pathologies that might involve the hemidiaphragm, all of which are rare, and determine if it is possible to be confident in differentiating them from mesothelial cysts on the basis of imaging findings. These will be considered in turn:  

**Intradiaphragmatic bronchogenic cysts**

These lesions, which are also extremely rare, typically involve the posteromedial hemidiaphragm adjacent to the spine. They almost invariably contain soft tissue elements or proteinaceous fluid that is echogenic on ultrasound and are, therefore, usually easily differentiated from mesothelial cysts [11–13].

**Malignant primary diaphragmatic sarcomas and malignant hepatic and pulmonary neoplasms infiltrating the diaphragm**

These are invariably soft tissue tumours and will not be confused with thin-walled mesothelial cysts on ultrasound even if complicated by necrosis. Contrast-enhanced CT and MR will demonstrate pathologic enhancement [14].

**Hydatid disease**

This is the pathology that is most likely to cause diagnostic confusion and should certainly be considered in endemic regions. It should be possible to differentiate hepatic subcapsular hydatid cysts from extrahepatic, subdiaphragmatic disease due to peritoneal spread but the differentiation of the latter from intradiaphragmatic hydatid cysts or mesothelial cysts is considerably more difficult. The presence of multiple cysts and the ultrasound, CT, or MR demonstration of daughter cysts would clearly support hydatid disease. Calcification has been described in both hydatid disease and mesothelial cysts [5,8,18] and is not, therefore, an especially useful discriminatory finding. Serological tests supporting hydatid disease are clearly helpful but are not positive in all cases [15–19].

The case that we have described here is a useful addition to the literature on intradiaphragmatic mesothelial cysts for several reasons. Firstly, it has described the ultrasound features of these lesions, including splitting of muscular fibres of the hemidiaphragm by the cyst, which confidently demonstrate an intradiaphragmatic location without surgical confirmation; secondly, it shows the importance of a review of previous imaging which has allowed us to confirm a lack of change in size over 4 years; thirdly, it has allowed a critical review of the literature in both adult and paediatric groups.

**Teaching point**

Mesothelial cysts of the diaphragm are extremely rare and may mimic hepatic or pulmonary pathology. Ultrasound may provide the best non-invasive way of confirming their nature and allow conservative management.

**REFERENCES**

[1] Cruickshank G, Cruickshank DB. Intradiaphragmatic mesothelial cysts. Thorax 1951;61:145–53.
[2] Mouroux J, Venissac N, Leo F, Guillot F, Padovani B, Hofman P. Usual and unusual locations of intrathoracic mesothelial cysts. Is endoscopic resection always possible? Eur J Cardio-thorac Surg 2003;24:684–8.
[3] Greenberg M, Madan V, Ataie EO, Rao AK. Intradiaphragmatic cyst: a diagnostic challenge. JAMA 1974;230(8):1176.
[4] Ueda H, Andoh K, Kusano T, Iwasaki A, Inutsuka S. Diaphragmatic cyst with elevated level of serum tissue polypeptide antigen. Thorac Cardiovasc Surg 1992;40:195–7.

[5] Sans N, Giron J, Bloom E, Daste G, Padovani B, Bernard JL, et al. Congenital mesothelial cyst of the diaphragm: imaging aspects. Apropos of 2 cases in adults and review of the literature. J Radiol 1999;80(6):593–6.

[6] Martino G, Braccioni A, Vergine M, Calvitti M, Cariati S, Veneroso S, et al. Mesothelial cyst of the diaphragm. Report of a case and review of the literature. G Chir 2000;21(6-7):290–6.

[7] Estaun JE, Alfagerme AG, Banuelos JS. Radiological appearance of diaphragmatic mesothelial cysts. Pediatr Radiol 2003;33:855–8.

[8] Akinci D, Akhan O, Ozmen M, Ozkan OS, Karcaaltincaba M. Diaphragmatic mesothelial cysts in children: radiologic findings and percutaneous ethanol sclerotherapy. AJR 2005;185(4):873–7.

[9] Kahriman G, Ozcan N, Dogan S, Bayram A. Imaging findings and management of diaphragmatic cysts in children. Pediatr Radiol 2016;46:1546–51.

[10] Oncel M, Sunam GS, Ozbek S. Congenital cyst in a rare localisation. BMJ Case Rep June 2013. doi:10.1136/bcr-2013-009298.

[11] Dagenais F, Nassif E, Dery R, Lapointe R. Bronchogenic cyst of the right hemidiaphragm. Ann Thorac Surg 1995;59:1235–7.

[12] Westphal FL, Menezes AQ, Guimaraes RAG. Intradiaphragmatic bronchogenic cyst. J Pneumol 2003;29(3):148–50.

[13] Mubang R, Brady JJ, Mao M, Burfeind W, Puc M. Intradiaphragmatic bronchogenic cysts: case report and systematic review. J Cardiothorac Surg 2016;11(1):79.

[14] Thapar S, Ahuja A, Rastogi A. Rare diaphragmatic tumor mimicking liver mass. World J Gastrointest Surg 2014;6(2):33–7.

[15] Eren S, Ulku R, Tanrikulu AC, Eren MN. Primary giant hydatid cyst of the diaphragm. Ann Thorac Cardiovasc Surg 2004;10(2):118–19.

[16] Aydin Y, Ozgokce M, Naldan ME, Turkyilmaz A, Ergolu A. Diaphragmatic hydatid cyst: report of three cases. Turk Gogus Kalp Damar 2014;22(3):672–5.

[17] Kumar VKDP, Shetty S, Saxena R. Primary hydatid cyst of the diaphragm mimicking diaphragmatic tumour: a case report. J Clin Diagn Res 2015;9(8):TD03–4.

[18] Salih AM, Kkamad FH, Rauf GM. Isolated hydatid cyst of the diaphragm, a case report. Int J Surg Case Rep 2016;29:130–2.

[19] Dumitrescu M, Bolca C, Cordos I. Primary hydatid cyst of the diaphragm: a case report. J Surg 2014;10(3):253–4.