Incremental value of live/real time three-dimensional transesophageal echocardiography over the two-dimensional technique in the identification of accessory liver lobe presenting as a right atrial mass

Abstract

We describe an adult patient with a large right atrial mass which was found to have the same echotexture as adjacent liver tissue on two-dimensional transesophageal echocardiography (2DTEE). By using live/real time three-dimensional transesophageal echocardiography (3DTEE), we were able to examine the mass by meticulous cropping and confirm the echotexture was similar to liver tissue throughout its extent. Three-dimensional cropping also redemonstrated in a definitive manner that the mass was in continuity with liver. Thus, in our patient 3DTEE served to increase the confidence level of making a definitive diagnosis of a supernumerary lobe of liver. To the best of our knowledge, the incremental value of 3DTEE over 2DTEE in diagnosing a supernumerary liver lobe has not been reported previously. The mass was surgically resected and the findings confirmed as liver tissue by pathological examination.

Keywords: Right atrial mass, inferior vena cava, accessory liver lobe, three-dimensional transesophageal echocardiography, intracardiac mass, supernumerary liver lobe, two-dimensional transesophageal echocardiography

Introduction

Accessory liver lobe is a congenital abnormality of hepatic tissue overgrowth. It is related to embryonic heteroplasia or rarely may occur secondary to trauma or surgery.[1] When in direct continuity with liver it is termed a supernumerary liver lobe and when completely separate from liver is described as an ectopic liver lobe.[2] The reported incidence of accessory liver lobes, including Riedel's lobe, which is the most well-known type seen as a tongue like protrusion of hepatic segments V and VI, is variably stated as being 3.3 to 31% in the general population.[3,4] Accessory liver lobe can be found at many sites such as the gallbladder, spleen, retroperitoneum, pancreas, adrenal gland, portal vein, diaphragm, thorax, gastric serosa, inferior vena cava (IVC), testes, and umbilical vein. Accessory liver lobe presenting as a right atrial (RA) mass is a very rare occurrence and there are only 15 cases reported in the literature. Ansari-Gilani et al. in their case report of supernumerary liver lobe have commented on the echo texture of the RA mass being similar to liver tissue on two-dimensional transesophageal echocardiography (2D TEE), but in the remainder 14 cases echo texture was not reported. Also, three-dimensional transesophageal echocardiography (3DTEE) was not performed in any case. We report a case of a supernumerary liver lobe which extended from intrahepatic inferior vena cava (IVC) into the RA and the utility of live/real time 3DTEE in making a more confident definitive diagnosis of this lesion. To the best of our knowledge, this has not been reported previously.

Case Report

This was a 53-year-old African American female who was referred to our tertiary care university hospital for surgical management of a RA mass which was thought to be a tumor. She presented to the outside hospital with polydipsia, polyuria, dyspnea and blurring of vision and was found to have diabetes mellitus with ketoacidosis. Auscultation revealed a loud murmur and she underwent 2DTTE which found a mobile RA mass about 4 x 3 cm in size. No other abnormalities were detected and both left and right ventricular function were normal. To evaluate the mass further 2DTEE was performed which showed the mass to be pedunculated and arising from the intrahepatic portion of IVC. The mass did not obstruct the tricuspid valve or IVC. Despite optimal management of her diabetes and ketoacidosis, the patient’s dyspnea worsened with increased oxygen requirements. A computed tomography angiography of the chest was performed which showed sub massive pulmonary embolism. She was begun on low molecular weight heparin infusion and transferred to our hospital for surgical excision of the RA mass. Repeat 2DTTE performed in our institution with a Philips (Philips Healthcare, Andover, MA) EPIQ 7 ultrasound system and a X5-1 transducer showed no changes from the previous study. Three-dimensional (3D) TTE was also performed but the images were suboptimal in quality and did not add to the information provided by 2DTTE. The mass measured about 3.79 cm x 2.5 cm on 2DTTE and the attachment of the mass to the intrahepatic IVC was not seen on both 2DTTE and 3DTTE. In preparation for cardiac surgery left heart catherization was performed which demonstrated only mild non-obstructive coronary artery disease.

During surgery, intraoperative 2DTEE performed with similar equipment as 2DTTE and a X7-2t transducer showed a large pedunculated mass measuring 4.5 cm x 3.1 cm which was echodense, homogenous in appearance with no calcification and noted to be arising from the intrahepatic portion of the IVC (Figure 1 and Movie 1). It was attached to the liver with a pedicle measuring 1.1 cm mediolaterally and on 2DTEE (Movie 2), the mass had uniform sponge-like texture with medium echogenicity indistinguishable from the adjoining liver parenchyma, but we did not notice any tubular echolucencies in the mass suggestive of vasculature. No other abnormalities were noted except for moderate tricuspid regurgitation. 3DTEE was also performed as described in the literature before [5] and the acquired 3D data sets carefully and sequentially cropped using multiple plane angulations in both full volume and multiple plane reconstruction (MPR) modes (Figure 2). Cropping using QLAB software (Philips Healthcare, Andover, MA) demonstrated the texture of the mass throughout its entire extent to be strikingly similar to liver, a portion of which was also acquired in the 3D data sets (Movie 3 and 4). Cropping of 3D data sets also showed no evidence of very bright reflective areas suggestive of calcification or echolucencies which signify extensive vasculature/ hemorrhage or lysis seen in a thrombus. The mass with its pedicle was noted to be continuous with adjacent liver on 2DTEE, but cropping by 3DTEE helped us visualize the pedicle in axial, coronal and sagittal planes to be in continuity with liver (Movie 5). Thus, a diagnosis of supernumerary liver lobe rather than ectopic liver could be definitely made.

At surgery the RA mass was freed up and resected from the intrahepatic IVC. Due to co-existent moderate tricuspid regurgitation and annular dilation noted on TEE, the surgeon repaired the tricuspid valve using a 25 mm ATS Simulus ™ FLX-C band (Medtronic, Minneapolis, MN). The patient did well following surgery and was discharged in a satisfactory condition. Histological examination of the resected tissue (Figure 3) with reticulin stain confirmed the 3D TEE findings of normal liver tissue with no pathological abnormality.

DISCUSSION

In our patient 3D TEE increased the confidence level of 2DTEE in making a definitive diagnosis of a supernumerary liver lobe which presented as a mass in the IVC and RA initially thought to be a tumor. Firstly, 3DTEE showed the texture of the mass exactly similar to liver tissue not only in a few sections as in 2DTEE but throughout the whole extent of the mass acquired in the 3D data set increasing the confidence level of the diagnosis. Secondly, it helped us confirm the continuity of the mass to liver imaged adjacent to the intrahepatic IVC by visualization in multiple planes. During evaluation of this accessory hepatic tissue, we thought it is of great value for all the echocardiographers to have a knowledge of the ultrasound characteristics of the normal liver and some commonly encountered pathological lesions (Table I).

Table I. Ultrasound characteristics of normal liver compared with pathological lesions

	Gray scale and Doppler ultrasonography findings
Normal Liver	Homogenous with low-level gray, is mildly hyperechoic or isoechoic compared to renal cortex and is hypoechoic compared to spleen. Branching tubular echolucencies that can be traced towards the porta or the IVC. [6]
Fatty liver	Based on the severity minimal to marked increase in echogenicity and normal visualization of the diaphragm to poor or no visualization of the hepatic vessels and diaphragm. [7]
Hepatitis	Diffusely decreased echogenicity with accentuated brightness of the portal triads or increased thickness and echogenicity of the soft tissues surrounding the portal vein branch, called periportal cuffing. Normal color Doppler
Liver cyst	Anechoic with a thin, well-demarcated wall and posterior acoustic enhancement and no internal vascularity on color Doppler. [8]
Hemangioma	Typically, small lesion (< 3 cm), well defined, homogenous and hyperechoic. Color Doppler may show peripheral feeding vessels. [9]
Focal Nodular Hyperplasia	Mass may appear similar to liver tissue, slightly inhomogeneous scar in the center is hypoechoic. Blood vessels we visualized in the center and periphery on Doppler. [10]
Hepatocellula	Variable appearance. Most small HCCs (<5cm) are hypoechoic with a thin,

r carcinoma (HCC)	peripheral hypoechoic halo (corresponding to fibrous capsule). With time, masses tend to become more complex, echogenic and inhomogeneous as a result of necrosis and fibrosis. [11]
Metastasis	Can be single, but usually multiple well-defined rounded lesions of varying size with positive mass effect causing distortion of adjacent vasculature and a hypoechoic halo (target or bull's eye sign) surrounding the liver mass. [12]

3D echocardiography (3DE) has been found useful and superior to 2D echocardiography (Table II) in characterizing intracardiac masses [13-19] but so far there are no reports of its usefulness in diagnosing an accessory liver lobe. As 3D echocardiography evolved over time, several authors have reported the advantages of using 3DTTE and 3DTEE in the assessment of cardiac masses. [13,18,20] Full volume Images can be obtained by 3DE and sectioned in multiple planes to examine and assess the intracardiac masses in terms of their homogenous or heterogeneous nature, point of attachment, vascularity and calcifications. The tissue characteristics of a few cardiac masses are peculiar on 3DE, which guides in the diagnosis of their etiology.

Table II. Three-dimensional echocardiography (3D Echo) characteristics of various cardiac masses

Mass	Real time 3D Echo characteristic findings
Myxoma	Discrete Echolucencies concerned with necrosis/hemorrhage. Calcification presents as hyperechoic reflective areas
Hemangioma	Echolucencies consistent with vasculature involving the entire mass with minimal solid tissue
Sarcoma/ chordoma	Highly reflective rings surrounding echolucencies (necrosis/vasculature), resembling a “doughnut”
Thrombi	Central area of echolucencies (due to lysis) and association with low flow state

On review of literature only 15 reported cases of accessory hepatic tissue presenting as a right atrial mass were found which are included in the following table (Table III).

Table III. Review of literature of cases with accessory hepatic tissue presenting as right atrial mass

Author	Age/Sex	Presentation	Ectopic or Supernumerary
Doshi et al. [21] May 2019	46/F	Paroxysmal atrial fibrillation	Supernumerary and pedunculated
D'Angelo et al.[22] April 2019	53/M	Incidentally diagnosed	Supernumerary and pedunculated
Soliman et al. [23] Jan 2019	37/F	Left popliteal artery occlusion	Supernumerary
Yeh et al. [24] Feb 2018	69/F	Palpitations	Supernumerary and pedunculated
Giritharan et al. [25] Jan 2018	30/F	Chest pain	Supernumerary and pedunculated
Izzo et al. [26] Dec 2017	43/F	Syncope	Supernumerary and sessile
Peake et al. [27] March 2017	66/F	Incidentally diagnosed	Supernumerary
Forest et al. [28]	58 /F	Syncope	Supernumerary and sessile

Aug 2016			
Moody et al. [29] April 2016	37/F	Palpitations (Atrial fibrillation)	Supernumerary and pedunculated
Ansari-Gilani et al. [30] Feb 2014	40/F	Recurrent chest pain	Supernumerary and pedunculated
Xu et al. [31] June 2012	52/F	Fatigue and chest discomfort	Ectopic
Sarsam et al. [32] April 2012	40/F	Dyspnea	Supernumerary
Trocciola et al. [33] Jan 2011	42/F	Palpitations and dyspnea	Supernumerary and pedunculated
Chapman- Fredricks et al. [34] Nov 2010	12/M	Seizure	Supernumerary and pedunculated
Brustmann et al. [35]	20/M	Autopsy finding	Ectopic

Aug 2002			

Conclusion

Accessory liver lobe should be considered as a part of the differential diagnosis of right atrial masses, particularly in females of age group 30 - 70 years. Our case demonstrates the role of 3DTEE and its additional value over the 2DTEE in making a more confident definitive diagnosis of a supernumerary liver lobe presenting as a mass in the IVC and RA.

References

1. Wang C, Cheng L, Zhang Z, et al. Accessory Lobes of the Liver: A Report of 3 Cases and Review of the Literature. *Intractable Rare Dis Res*. 2012;1(2):86-91.
2. Glenisson M, Salloum C, Lim C, et al. Accessory Liver Lobes: Anatomical Description and Clinical Implications. *J Visc Surg*. 2014;151(6):451-455.
3. Sham R, Sain A, Silver L. Hypertrophic Riedel's Lobe of the Liver. *Clin Nucl Med*. 1978;3(3):79-81.
4. Gillard JH, Patel MC, Abrahams PH, et al. Riedel's Lobe of the Liver: Fact or Fiction? *Clin Anat*. 1998;11(1):47-49.
5. Sinha A, Nanda NC, Khanna D, et al. Morphological Assessment of Left Ventricular Thrombus by Live Three-Dimensional Transthoracic Echocardiography. *Echocardiography*. 2004;21(7):649-655.
6. Wilson S, Withers C. The Liver In: Rumack C, Levine D, eds. *Diagnostic Ultrasound*. Fifth ed.: Elsevier, inc.; 2018:74w-138.
7. Wilson SR, Rosen IE, Chin-Sang HB, et al. Fatty Infiltration of the Liver--an Imaging Challenge. *J Can Assoc Radiol*. 1982;33(4):227-232.
8. Gaines PA, Sampson MA. The Prevalence and Characterization of Simple Hepatic Cysts by Ultrasound Examination. *The British Journal of Radiology*. 1989;62(736):335-337.
9. Bree RL, Schwab RE, Neiman HL. Solitary Echogenic Spot in the Liver: Is It Diagnostic of a Hemangioma? *American Journal of Roentgenology*. 1983;140(1):41-45.
10. Buetow PC, Pantongrag-Brown L, Buck JL, et al. Focal Nodular Hyperplasia of the Liver: Radiologic-Pathologic Correlation. *Radiographics*. 1996;16(2):369-388.
11. Tanaka S, Kitamura T, Imaoka S, et al. Hepatocellular Carcinoma: Sonographic and Histologic Correlation. *AJR Am J Roentgenol*. 1983;140(4):701-707.
12. Wernecke K, Vassallo P, Bick U, et al. The Distinction between Benign and Malignant Liver Tumors on Sonography: Value of a Hypoechoic Halo. *AJR Am J Roentgenol*. 1992;159(5):1005-1009.
13. Asch FM, Bieganski SP, Panza JA, et al. Real-Time 3-Dimensional Echocardiography Evaluation of Intracardiac Masses. *Echocardiography*. 2006;23(3):218-224.
14. Meng Q, Lai H, Lima J, et al. Echocardiographic and Pathologic Characteristics of Primary Cardiac Tumors: A Study of 149 Cases. *Int J Cardiol*. 2002;84(1):69-75.
15. Singh A, Miller AP, Nanda NC, et al. Papillary Fibroelastoma of the Pulmonary Valve: Assessment by Live/Real Time Three-Dimensional Transthoracic Echocardiography. *Echocardiography*. 2006;23(10):880-883.
16. Dod HS, Burri MV, Hooda D, et al. Two- and Three-Dimensional Transthoracic and Transesophageal Echocardiographic Findings in Epithelioid Hemangioma Involving the Mitral Valve. *Echocardiography*. 2008;25(4):443-445.
17. Mehmood F, Nanda NC, Vengala S, et al. Live Three-Dimensional Transthoracic Echocardiographic Assessment of Left Atrial Tumors. *Echocardiography*. 2005;22(2):137-143.

18. Reddy VK, Faulkner M, Bandarupalli N, et al. Incremental Value of Live/Real Time Three-Dimensional Transthoracic Echocardiography in the Assessment of Right Ventricular Masses. *Echocardiography*. 2009;26(5):598-609.
19. Suwanjutah T, Singh H, Plaisance BR, et al. Live/Real Time Three-Dimensional Transthoracic Echocardiographic Findings in Primary Left Atrial Leiomyosarcoma. *Echocardiography*. 2008;25(3):337-339.
20. Zaragoza-Macias E, Chen MA, Gill EA. Real Time Three-Dimensional Echocardiography Evaluation of Intracardiac Masses. *Echocardiography*. 2012;29(2):207-219.
21. Doshi A, Shah D, Gupta S, et al. A Rare Case of an Ectopic Liver Presenting as Right Atrial Mass. *Case Rep Cardiol*. 2019;2019:4103827.
22. D'Angelo AM, van Boxtel BS, Wu IY, et al. Deep Hypothermic Circulatory Arrest for Resection of a Right Atrial Accessory Hepatic Lobe. *J Thorac Cardiovasc Surg*. 2019;158(3):e101-e102.
23. Soliman M, Akanbi O, Salem A, et al. Ectopic Liver Tissue Mistakenly Diagnosed as a Right Atrial Myxoma. *Cureus*. 2019;11(1):e3862.
24. Yeh B, Hudson AJ, Reoma JL, et al. Intracardiac Heterotopic Liver Presenting as a Right Atrial Myxoma: A Case Report. *Mil Med*. 2018;183(1-2):e175-e178.
25. Giritharan S, Salhiyyah K, Tsang G. Surgical Excision of Ectopic Hepatic Tissue in the Right Atrium. *J Thorac Cardiovasc Surg*. 2018;155(1):e47-e48.
26. Izzo BA, Stoudenmire CJ, Byrd BF, 3rd. Ectopic Liver Tissue Presenting as a Right Atrial Mass on Echocardiography. *CASE (Phila)*. 2017;1(6):212-215.
27. Peake B, Seevanayagam S, Vazquez M, et al. Aberrant Non-Neoplastic Liver Tissue Presenting as an Intra-Atrial Mass. *BMJ Case Rep*. 2017;2017.
28. Forest SJ, Kaplan KC, Michler RE. Accessory Liver in the Right Atrium: A Rare Cause of Syncope. *Ann Thorac Surg*. 2016;102(3):e229-e231.
29. Moody WE, Hubscher SG, Rooney SJ, et al. Intracardiac Ectopic Liver Mimicking Atrial Myxoma--an Unusual Cause for a Right Atrial Mass. *Int J Cardiol*. 2016;209:210-212.
30. Ansari-Gilani K, Jenkins T, Landeras L, et al. Multimodality Imaging of an Unusual Case of an Obstructive Intracaval Mass by an Aberrant Liver. *Circulation*. 2014;129(5):e310-312.
31. Xu L, Jeudy J, Burke AP. Ectopic Hepatic Tissue Presenting as Right Atrial Mass. *Hum Pathol*. 2012;43(6):958-960.
32. Sarsam S, Arsene C, Abu-Rashed K, et al. Heterotopic Hepatic Polyp Identified in the Right Atrium. *J Med Cases*. 2012.
33. Trocciola SM, Balsam LB, Yee H, et al. Ectopic Liver: An Unexpected Finding in a Right Atrial Mass. *Ann Thorac Surg*. 2011;92(2):715-718.
34. Chapman-Fredricks J, Birusingh R, Ricci M, et al. Intracaval Liver with Cardiac Extension. A New Developmental Anomaly? *Fetal Pediatr Pathol*. 2010;29(6):401-406.
35. Brustmann H. Heterotopic Liver in the Right Cardiac Auricle. *Ann Diagn Pathol*. 2002;6(4):248-249.