A rare anatomic anomaly- Ectopic liver presenting as right atrial mass

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LIFE CHANGING MEDICINE

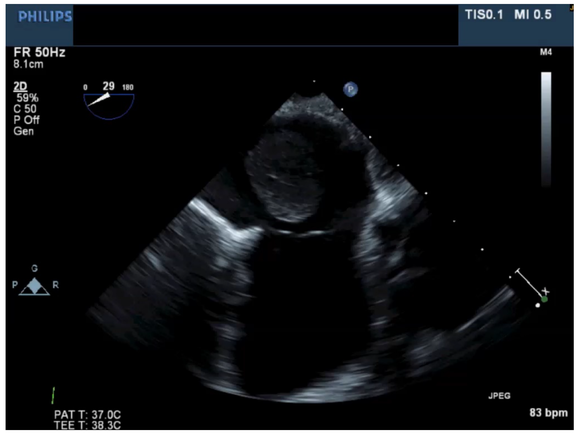
Introduction

- Ectopic Liver is generally observed intra-abdominally, its extension supra-diaphragmatically is a rare occurrence.
- There are only 23 such reported cases in literature with majority being reported in younger age groups as extension of caudate lobe of liver through a diaphragmatic hernia.
- However presentation of an ectopic liver as an atrial mass in adults is an extremely rare anomaly, with only 3 reported cases in Pubmed

Presentation

- Patient is a 45 year old obese Caucasian male
- Past medical history of obstructive sleep apnea presented with chief complaints of worsening of shortness of breath on exertion over a period of 8 months
- It is also associated with one episode of syncope in the remote past.
- He denied any orthopnea, chest pain, palpitations, recurrent syncope, paroxysmal nocturnal dyspnea or lower extremity edema
- Physical exam S1, S2 with a diastolic murmur was heard. All other exam were unremarkable
- Complete blood counts and metabolic panel were unremarkable.
- Initial troponin was 0.03 trended to a maximum value of 0.08, subsequently trended down to 0.05
- Electrocardiogram showed normal sinus rhythm
- Transthoracic Echocardiogram (TTE) revealed a 2.7x1.6 cm mass at the junction of right atrium (RA) and inferior vena cava (IVC) and a patent foramen ovale(PFO) and ejection fraction of 55-60%.
- Cardiac surgery performed right mini thoracotomy, resection of right atrial /inferior vena cava mass with PFO closure
- Firm, brown with rubbery consistency was resected followed by closure of PFO
- Histology confirmed partially encapsulated liver parenchyma.
- Post operative course was unremarkable, resolution of dyspnea noted at follow-up.

Imaging



TTE showing globular,mobile echo density that appeared to the posterolateral aspect of the right atriumright atrial mass 2.7 x 1.6cm in a 4 chamber view

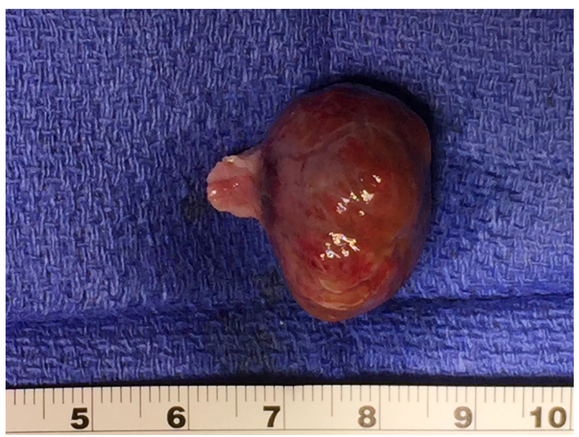
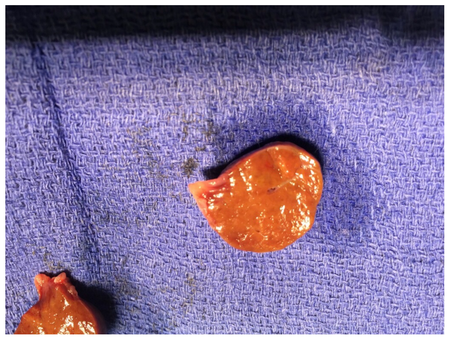
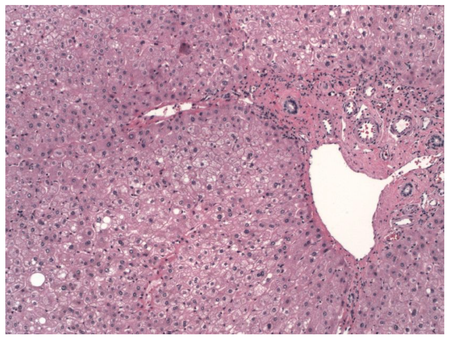


Figure of mass the size of a grape coming from the IVC wall below the diaphragm, the mass was resected along with its stalk

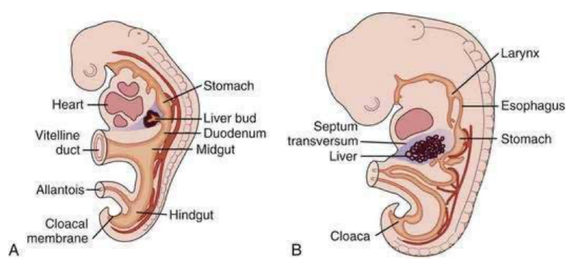
Imaging



Sectioning showing a brownish colored tissue with rubber consistency similiar to renal or hepatic tissue



Histology shows **hexagonal plates** of hepatocytes stacked on top of each other arranged similiar to the spokes of cartwheel



Close association of Liver Bud/ IVC/ Septum Transversum allows migration of hepatocytes into the IVC lumen During formation of the caudate lobe

Conclusion

- Right atrial masses are often an incidental discovery on echocardiography.
- 75 percent of primary cardiac tumors are benign,majority of benign lesions are myxomas; other common benign lesions include papillary fibroelastomas and lipomas. In children, rhabdomyomas and fibromas are the most common, however ectopic liver as a cardiac mass is extremely rare
- Regardless resection often remains the default choice considering the embolic and carcinogenic potential of ectopic masses, including sudden death
- However there exists no imaging or diagnostic modality that can yet differentiate a cardiac tumor from an ectopic mass.
- A possible clue in our patient could be location of the mass in the high IVC/RA junction.
- A plausible explanation to this anomaly could be the close association of liver bud, IVC and septum transversum that allows migration of hepatocytes into the IVC lumen during formation of the caudate lobe.

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