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Accessory Hepatic Lobe-A 'not so Rare' Entity

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ABSTRACT

Introduction: Accessory Hepatic Lobe (AHL) is an uncommon developmental anomaly usually asymptomatic and reported as an incidental finding on cross-sectional imaging, laparotomy or autopsy. With increasing number of cases undergoing cross-sectional imaging the detection rate of AHL has increased.

Aim: The aim of this study was to assess the incidence and describe the imaging features of AHL using Computed Tomography.

Materials and Methods: A retrospective review of CT images of patients who underwent abdominal CT for diverse clinical indications was undertaken for detection of accessory hepatic tissue within the abdomen and pelvis and the data so obtained was analysed.

One thousand abdominal CT-scans were retrospectively

reviewed for the presence of AHL and when identified, were further characterized based on their location, volume and blood supply, wherever possible using existing set of images.

Results: A total of seven cases with AHLs were detected during review of 1000 case studies, with an incidence of 0.7% in our study group. All AHLs were perihepatic in location. Five of the accessory lobes were connected to the left lobe of liver by a band of hepatic tissue or a vascular pedicle and two AHLs were seen arising from the caudate lobe.

Conclusion: AHL is not as uncommon an entity as was earlier believed. Awareness regarding this entity and its clinical significance is important in order to be able to identify and manage complications arising from it.

Keywords: Anomalies, Congenital, Imaging, Liver

INTRODUCTION

AHL was considered as a rare developmental anomaly. Most cases with an AHL are asymptomatic and reported as incidental finding on cross-sectional imaging, laparotomy or autopsy. Very rarely an AHL may become symptomatic and present with abdominal pain or torsion. With the ever increasing use of cross-sectional imaging modalities the incidence of detection of AHL has also increased. Accessory hepatic tissue can be easily identified and characterized when detected by CT or MRI of the abdomen is performed for related or unrelated clinical conditions. Awareness of existence of this anomaly enables early diagnosis by imaging modalities like ultrasound, CT and MRI.

This study aims to discuss the incidence and imaging findings in cases with AHLs, detected at imaging, with a brief review of literature pertaining to this anomaly.

MATERIALS AND METHODS

A total of 1000 abdominal CT-scans performed in the Department of Radiology of Nizam's Institute of Medical

Sciences, Telangana, India, over a period of one year (May 2015 - April 2016), were retrospectively reviewed by random selection. These included patients of all age groups, both males and females, who underwent plain and contrast enhanced CT-scans of the abdomen for various indications, referred by different medical and surgical specialties of the hospital.

All scans were performed on Philips 16 slice MDCT Brilliance scanner using standard techniques after informed consent obtained from all patients prior to the examination. The abdominal CT-scans were retrospectively evaluated by two experienced radiologists (with 22 and 8 years of experience in body imaging respectively). All images were reviewed for the presence of any accessory hepatic tissue and the observations were recorded and subsequently analysed.

STATISTICAL ANALYSIS

Since this was an observational study, no statistical methods were employed for analysis.

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RESULTS

A total of 1000 abdominal CT-scans performed for varied indications were reviewed retrospectively for the presence of accessory hepatic tissue /lobe. There were 676 male and 324 female subjects in the study group with their ages ranging from 18 to 85 years.

In our study including 1000 cases, a total of seven cases with of these cases of AHL, five were males and two females with the youngest subject being 36 years of age and the oldest 85 years.

On reviewing the images, seven cases with AHLs were

detected. The location and morphological features of the accessory lobes and their blood supply was identified and recorded [Table/Fig-1-7].

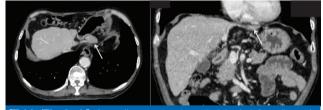
DISCUSSION

AHL is considered as a rare developmental anomaly. The exact incidence in general population has not been extensively reported in literature.

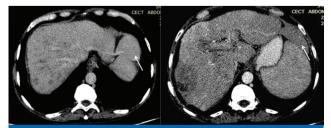
AHL may present as accessory hepatic tissue lying close and connected to the liver or may occur in ectopic locations within the abdomen pelvis or thorax. An accessory lobe of liver is usually asymptomatic and of little clinical significance. It may

S. No.	Age (in years)	Sex	Clinical Details	Location of AHL	Size of AHL (in cm)	Blood Supply
1	60	М	A case of Carcinoma Urinary Bladder for post treatment evaluation of abdomen	Sub phrenic/ perihepatic	3x1.5x1 separate pedicle	From left branch of hepatic artery and left portal vein
2	58	Μ	Chronic Liver Disease with Hepatocellular Carcinoma	Left perihepatic	7x3x2.5 separate pedicle	From left lobe branches
3	36	М	Suspected Renal Calculi	Perihepatic extension from caudate	6x3x3 tongue like projection from caudate lobe	From branches of caudate lobe
4	43	F	Bilateral Renal Calculi	Perihepatosplenic	4x3.5x2.5 separate pedicle	From left lobe
5	85	М	Abdominal Aortogram for Atherosclerotic Disease	Perihepatic	3.2x2.7x2.5 separate pedicle and accessory fissure with separate capsule	From left lobe
6	50	М	Hepatic Hemangiomas	Perihepatic extension of caudate	1.8x1.6x1.5 sessile extension	From caudate lobe
7	53	F	Debulked Pseudomyxoma Peritonei	Perihepatic	1.2x1.1x1 short pedicle	Left lobe segment 4

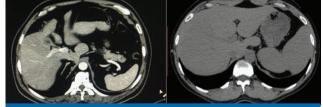
[Table/Fig-1]: Clinical and imaging features of accessory hepatic lobes.



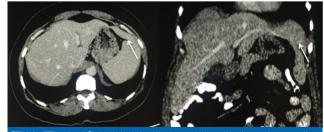
[Table/Fig-2a-b]: A small accessory hepatic lobe in left perihepatic subphrenic location (arrows) with its vascular pedicle connected to left hepatic lobe vessels.



[Table/Fig-3a-b]: A large AHL in left upper abdomen perihepatosplenic location (arrows) with its vascular pedicle arising from left lobe of liver (segment 2).



[Table/Fig-4a-b]: Two cases with AHL showing a tongue like projection of accessory hepatic tissue arising from the caudate lobe (arrows).



[Table/Fig-5a-b]: AHL in left subphrenic location (arrow) extending around the spleen with a vascular pedicle arising from left hepatic lobe vessels.



[Table/Fig-6]: Accessory lobe of liver lying to the left of left lobe of liver (arrow) with a complete fissure separating the AHL from segments 2 and 3 of liver.



[Table/Fig-7a-b]: A small accessory hepatic lobe connected to segment 4 of liver (arrow). An incomplete accessory fissure is also noted between segment 4 and right lobe of liver.

be detected incidentally at cross sectional imaging or during abdominal surgery. Rarely symptomatic presentation of AHL has been reported when the accessory lobe undergoes complications like torsion or tumour development. This anomaly might be misinterpreted as a space occupying lesion or neoplasm especially when occurring in ectopic locations. An awareness regarding the existence of AHL and its varied appearances is important in order to accurately diagnose this rare condition.

Theories of Development of AHL

This rare anatomical entity is believed to result from embryonic heteroplasia. It occurs from an error in the formation of the endodermal caudal foregut in the third gestational week and segmentation of the hepatic bud [1]. Rarely, an AHL may result following trauma or surgery [2]. Some authors suggest association of AHL with an autosomal recessive gene as observed by anatomical research based on necropsies in rats [3].

Presently, there are two hypotheses regarding the development of an AHL. One suggests that the embryonic liver curls outwards and forms an accessory lobe during early stage of development [4] and according to the other theory, an accessory lobe results from increased intra-abdominal pressure due to the development of the tunica muscularis recti and the enlarging liver [5].

Classification of Types of AHL

There is very little existing literature about the classification

of AHL. AHL has been classified into two types according to Stattaus - an accessory lobe joined to normal hepatic tissue or a completely separate accessory lobe. Alternately, AHL can also be classified as pedunculated or sessile [6].

Some authors have classified AHL by volume and weight into different types [2,7,8] -

(1) A bulky AHL (> 31 g) connected to the liver via a stalk of tissue or wide base in the subphrenic or perihepatic zone.

(2) A small AHL (11–30 g) connected to the liver via a wide base on the surface of the liver or around the right posterior lobe.

(3) A completely separate AHL with no connection to normal liver tissue that is most often seen in the thorax or pelvic cavity and;

(4) A pinpoint ectopic AHL (< 10 g) that is most often located at the margins of the liver or even gall bladder wall.

Ectopic location of AHL has also been reported in a few cases where the AHL was located in the thorax or pelvic cavity [7,9-12].

Yet another method of classification of AHL based on its biliary drainage and the presence or absence of a common capsule has also been proposed by some authors. They describe three types of AHL based on the above findings-

Type I - The separate accessory lobe duct drains into an intrahepatic bile duct of the normal liver.

Type II - The separate accessory lobe duct drains into an extrahepatic bile duct of the normal liver.

Type III - The accessory lobe and the normal liver have a common capsule; the bile duct of the accessory lobe drains into an extrahepatic duct [13].

All AHLs in our study were located in perihepatic location and were easily identified at CT. Of the seven cases reported here, AHL was sessile and connected to normal liver tissue in two cases and pedunculated with a separate vascular pedicle in other five cases. Classified by volume and weight, three of our cases had a bulky AHL while four cases had a small AHL.

None of the AHLs in our study showed any parenchymal abnormality or complication on imaging.

Clinical Presentation

AHL is usually asymptomatic and reported incidentally on cross sectional imaging, during laparotomy for related or unrelated conditions or at autopsy.

Symptomatic AHLs have been infrequently reported in literature at all ages from infancy to adulthood [11,12,14].

The clinical manifestations of AHL depend on the complications, such as torsion, traumatic rupture, or infarction [15-17].

Torsion of AHL is most frequent and may lead to severe complication. Cases with torsion present with acute abdomen

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or right upper quadrant pain with or without tenderness or a palpable mass [18]. Clinically, it may mimic many other causes of acute abdomen including acute cholecystitis, pancreatitis, perforated peptic ulcer or intussusception.

Other differential diagnoses include duodenal hematoma, intussusception, perforated peptic ulcer, ovarian torsion, wandering spleen, retrocaecal appendicitis and an intraabdominal tumour [8]. Umbilical hernia and bile duct cysts have also been reported as associations with this condition [1,16].

Ectopic AHL within the thorax may at be misdiagnosed as pulmonary, pleural or diaphragmatic tumour depending on its location [19,20].

With advances in cross-sectional imaging and the popularity of their use, the rate of AHL detection is increasing as more cases are being incidentally picked up during examinations performed for a related or unrelated cause.

All patients with AHL in our study presented with unrelated symptoms or disease conditions and AHL was detected incidentally in all these cases. None of the cases had symptoms or imaging features of complications related to the anomaly.

Diagnosis of AHL

Cross-sectional imaging modalities like ultrasonography, CT and MRI enable easy and accurate diagnosis of AHL, including its size, shape, location and blood supply.

An uncomplicated AHL incidentally detected by imaging has the same echogenicity, density or signal as normal liver tissue with distinct margins and may be connected to normal liver tissue by a stalk of tissue or a vascular pedicle [12,21]. The presence of independent biliary drainage as well as arterial, venous, and portal branches also helps to diagnose this rare anomaly. This subtype of AHL is more common and can be easily and confidently diagnosed at imaging.

In all seven of our cases, the AHLs were easily detected and characterized by CT due to their proximity to the liver and similar attenuation and enhancement pattern as the normal liver tissue on CT. Of these, five were pedunculated and were connected to the liver by a stalk of hepatic tissue in four and a vascular pedicle in two cases. Five out of seven cases of AHL was seen arising from or receiving vascular supply from left lobe of liver and in two cases from the caudate lobe.

The diagnosis of a complicated or ectopically located AHL may be difficult as it may present as an indeterminate soft tissue mass on imaging. The diagnosis of AHL may be challenging in such a situation and at times may even be missed. But careful evaluation of vascular connections might give a clue to the presence of AHL. None of our cases had ectopic location of AHL, as all the seven AHLs detected in our study were perihepatic in location.

Complications of AHL

A number of cases with complications related to AHL have been described in literature. These include torsion, infarction, haemorrhage and fracture of the accessory lobe. AHL has also been reported in association with biliary atresia, diaphragm defects, and acromphalus. Reports of gallbladder torsion, hepatic dysfunction and hemangioma occurring in AHL are also present in literature [15,22-24].

A pedunculated AHL carries a higher risk of acute complications like torsion, hemorrhage and rupture [4]. In addition to this any disease affecting the liver can also involve the accessory hepatic tissue including infections and neoplasms.

None of our cases presented with any complications of AHL.

Management of AHL

Asymptomatic patients with AHL discovered incidentally do not require any treatment. Some workers suggest that if a patient is diagnosed with a pedunculated AHL, surgery should be performed as soon as possible in order to avoid unexpected complications. However, this is a point of debate and cannot be generalized.

Rarely, emergency situations may arise and require urgent laparotomy or even surgical intervention in cases with acute torsion of the AHL. Resection of the AHL should be performed in adults with complications like torsion or rupture. These patients are reported to have a good prognosis following surgery.

It is also suggested to perform a liver transplant or resection of the AHL in patients with serious complications at birth, such as acromphalus, biliary atresia, or gallbladder torsion [25,26].

LIMITATIONS

The study includes a sample size of 1000 and hence, although representative, may not reflect the exact incidence of AHL in general population for which larger studies may be required.

Secondly, being a retrospective study, the scans reviewed were evaluated as available. Although, this did not affect the detection and morphological evaluation of AHLs but detailed evaluation of the vascular and ductal anatomy of the lobes could not be performed in all cases. A prospective study with arterial portal and hepatic venous phase imaging can further improve the information obtained in such cases.

CONCLUSION

It is important for clinicians and radiologists to be aware of this not so rare anatomic entity as it can present with acute or recurrent right upper quadrant pain. The routine check list while reading abdominal CT and MRI studies should include inspection of segmental anatomy including vascular and biliary drainage of the liver as this aids the radiologist in prompting the diagnosis of an accessory hepatic lobe.

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