

UNUSUAL APPEARANCE OF THE LIVER ON ULTRASONOGRAPHY AND COMPUTED TOMOGRAPHY IN A PATIENT WITH CYSTIC FIBROSIS*

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SUMMARY: Men S, Yüce A, Özmen M, Akhan O, Kale G. (Departments of Radiology and Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey). Unusual appearance of the liver on ultrasonography and computed tomography in a patient with cystic fibrosis. Turk J Pediatr 35: 135-140, 1993.

A seven-year-old boy with cystic fibrosis (CS) who presented with abdominal pain is reported. Ultrasonographic and computed tomographic studies of the upper abdomen revealed unusual liver findings. An ultrasound scan showed a liver that was exceedingly heterogenous and a mixed echo pattern with dominant hyperechogenicity. Computed tomography showed large, multiple hypodense cyst-like lesions in the liver. Using the ultrasound scan as a guide, a needle biopsy was performed. The pathological findings were in accord with the findings obtained from ultrasonography and computed tomography, and were consistent with pathological findings seen in CS cases. *Key words:* cystic fibrosis, ultrasound, computed tomography.

Cystic fibrosis (CS) is a complex hereditary disorder, and the most common lethal autosomal recessive disease of the Caucasian population. Defective ionic transport across epithelial surfaces appears to be the major pathogenic mechanism and is responsible for a wide and variable group of clinical manifestations and complications. In addition to abnormalities of salivary and sweat gland secretion and bronchogenic and pancreatic dysfunction, CF frequently involves the liver. The exact incidence is difficult to determine since most studies have employed different methods to detect hepatic involvement and have investigated different age groups. More recently, ultrasonography (US) has been used to assess parenchymal abnormalities, the incidence of abnormal scans ranging from 23 to 60 percent, depending on the age of group studied. All these studies showed that the incidence of liver involvement increases with age^{1,2}. The

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prevalence of frank biliary cirrhosis is probably five percent³. We present striking US and computed tomography (CT) findings in a CS patient with hepatic involvement.

Case Report

A seven-year-old boy with CF diagnosed at one year of age, was admitted to Hacettepe Children's Hospital for evaluation of abdominal pain. A diagnosis of CF had been established because the child had pulmonary infection, malabsorption and an elevated sweat chloride level (60, 80 mEq/L). He has been receiving pancreatic enzyme and fat-soluble vitamins. On this admission his height and weight measurements were below the 3rd percentile. Hepatomegaly was noted (three cm below the right costal margin and seven cm below the xiphoid).

Laboratory tests which included peripheral blood counts, liver function tests and prothrombin and partial thromboplastin times were within normal limits.

Abdominal ultrasound findings showed that the liver parenchyma was exceedingly heterogenous and had a nodular appearance. A mixed echo pattern with prominent hyperechogenicity, especially in the periportal areas, was observed. The liver was diffusely enlarged and the contours were irregular. The gall bladder was normal and the pancreas could not be visualized because of bowel gas which is a particular problem of patients with CF. The vertical length of the spleen was 107 mm and estimated to be at the upper limit (Figs. 1a,b). An abdominal CT scan showed hepatomegaly and multiple lobulated cyst-like lesions within the liver parenchyma which were several centimeters in size (Figs. 2a,b).

The attenuation values of -7, -8 and -14 Hounsfield units (HU) were obtained after measuring the centers of the lesions and they were consistent with the HU values of fat. Relatively hyperdense thin septa separated the hypodense lesions from each other. Intra and extrahepatic bile ducts were normal on calibration. The pancreas was replaced by fat. The US scan and CT findings revealed cyst-like lesions which were considered to be fatty infiltration of the liver parenchyma surrounded by thin bands of fibrosis.

A needle biopsy of the liver was performed using a 17 gauge Menghini needle and guided by the ultrasound scans. A pathological examination of the specimen exhibited extensive fatty infiltration in the hepatocytes, periportal fibrosis, bile duct proliferation with mucinous bile plugging in the lumen and pericholangitis consistent with the liver pathology of CF.

Discussion

Exocrine gland dysfunction occurs in patients with CF which is characterized by abnormal secretions from the mucous glands. Although organ involvement in CF

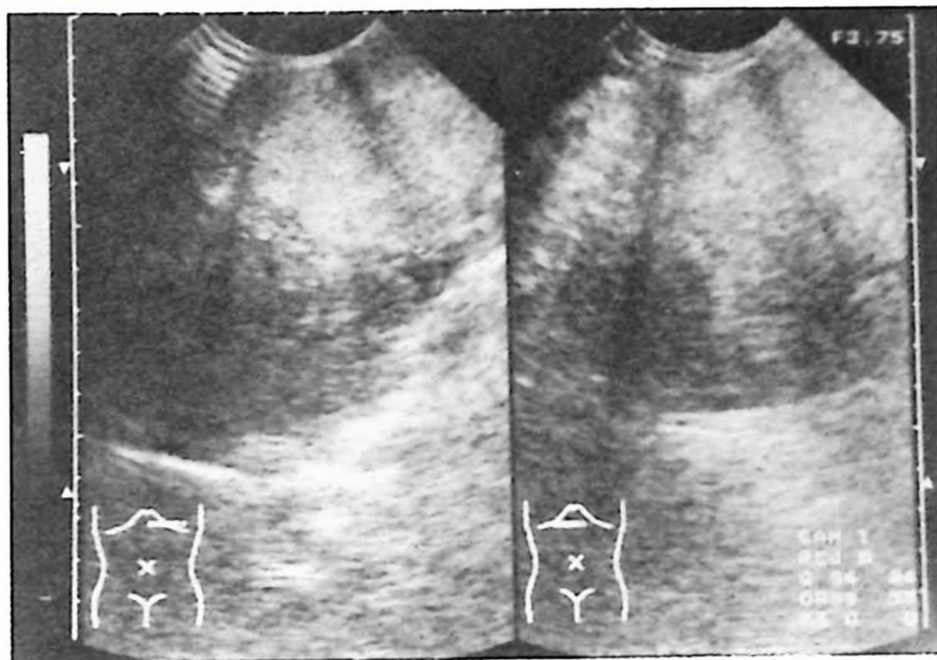


Fig. 1a

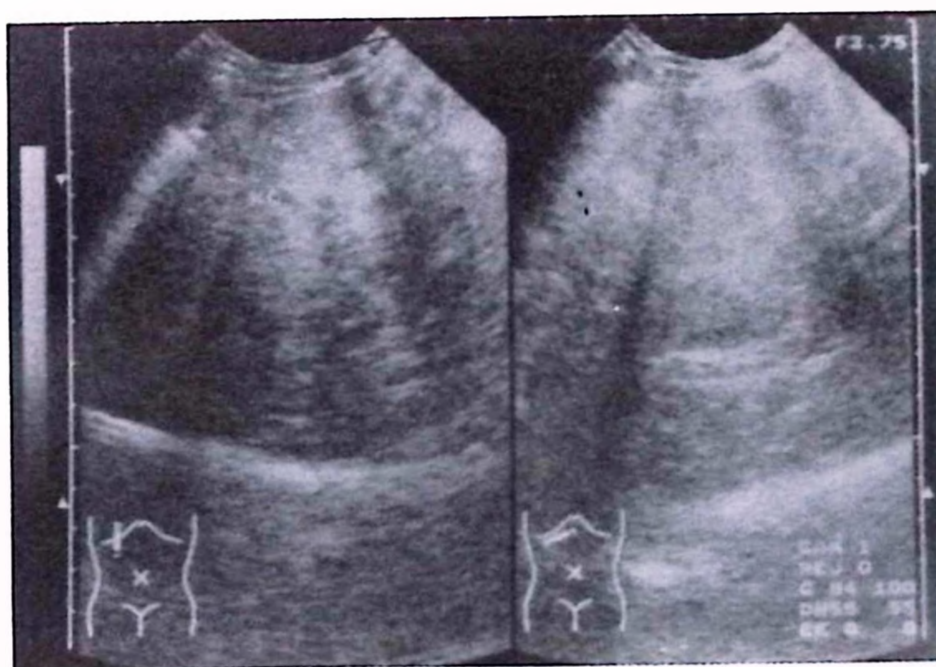


Fig. 1b

Figs. 1a, 1b: Subcostal axial and sagittal sections show increased echogenicity throughout the liver parenchyma and a heterogenous echo pattern with nodularity.

varies, hepatic involvement tends to increase with age. The presence of CF-related liver disease has been ascribed to obstruction of bile ductules with inspissated mucus, and the development of progressive cholangitis, periportal fibrosis and biliary cirrhosis².

The clinical and pathological spectrum of hepatic disease is reflected in a wide range of sonographic findings. Some patients have normal hepatic sonograms,



Fig. 2a



Fig. 2b

Figs. 2a, b: In the pre and post-contrast enhanced scans of the upper abdomen, multiple lobulated cyst-like lesions several centimeters in size and separated from each other by thin septa are seen. Note that there is no contrast enhancement within the lesions after administration of intravenous contrast material.

while others have irregular areas of increased echogenicity or alternating echogenicity and anechoicity and still others have rather prominent periportal echogenicity that corresponds to proliferation of bile ducts with or without biliary stasis, inflammatory changes, edema, fibrosis or a combination of these^{1,4,5}. Fatty infiltration of the liver is clearly demonstrated on computed tomography. The entire liver may show decreased density with bright portal venous areas.

However patchy infiltration of the liver may also be detected. Cyst-like changes represent the areas of fatty infiltration surrounded by fibrosis^{3,6}.

The CT scans of our patient revealed large, hypodense cyst-like lesions several centimeters in size in the liver, which simulated space-occupying lesions, while the sonograms showed a liver with a heterogenous multinodular appearance. To our knowledge, this is the first time such striking findings have been reported in a CF patient.

The lesions in our case mimic space-occupying lesions of the liver such as hepatocellular carcinoma, hepatoblastoma, multiple metastatic nodules, multiple abscesses, and polycystic disease of the liver.

The sonograms and CT scans did not demonstrate distortions or narrowing of the veins in the vicinity of the lesions in the liver as seen in hepatoblastoma, hepatocellular carcinoma or metastatic nodules⁷. In addition, the configuration of the cyst-like lesions observed in the liver of our patient was not consistent with the description of the cysts typical of polycystic disease^{7,8}.

Multiple liver abscesses may also mimic hepatic involvement of CF. On the CT scans of our patient there were thin linear bands surrounding the hypodense lesions which did not show the contrast enhancement expected in liver abscess⁸, and, therefore, were considered to be thin bands of fibrosis rather than granulation tissue.

The US and CT findings of our patient were consistent with the findings obtained following the pathological examination of the biopsy specimen. Extensive fatty infiltration in the hepatocytes was considered to be responsible for the increased echogenicity in the nodular lesions seen on US scans. We believed that the periportal fibrosis, cholangiolar proliferation, mucoid material in the bile ducts and cholangitis demonstrated on the pathological examination accounted for the increased periportal echogenicity on the US scans, and we assumed that the thin hyperdense septa surrounding and infiltrating the hypodense lesions on the CT scans represented thin bands of fibrosis.

In conclusion, we hope that CT and US findings in this patient may aid the radiologist in becoming more familiar with the various radiological patterns of liver involvement in CF patients.

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