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Case Report

Fetal Supradiaphragmatic Accessory Liver Lobe

Report of a Case and Review of the Literature

J. Vercelli-Retta

Department of Morbid Anatomy, Hospital de Clínicas, School of Medicine, Montevideo, Uruguay

Summary. A right thoracic accessory liver lobe with a normal diaphragm is reported in a stillborn fetus. A review of the literature reveals seven cases affecting adult patients.

Key words: Liver – Accessory lobe – Supradiaphragmatic – Fetus.

Introduction

Ectopic liver lobes are infrequent malformations, almost all of them being described in the abdominal cavity (Cullen, 1925; Willis, 1958). Thoracic heterotopic liver tissue without diaphragmatic malformation is a real curiosity. In the available literature there are only seven well documented human cases, all of them affecting adults. This paper reports the finding of a supradiaphragmatic accessory liver lobe in a stillborn fetus.

Case Report

A twenty five year old white pregnant female with a history of two previous spontaneous abortions (1973, 1974) presented in August 1975 at our Hospital. Her last normal menstrual period was on June 23, 1975. No history of exposure to any known teratogens, viral agents or drugs was elicited. After the first consultation, observation continued at the Obstetric Out-Patient Department every fifteen days. At the 22nd week of pregnancy the physical examination revealed no leg edema, the blood pressure was 120/65 mm Hg, the uterus measured 18 cm and the fetal heart rate was normal. She had no proteinuria and blood glucose was 95 mg/dl. On Dec 25th she was hospitalized with a 12 h history of spontaneous rupture of the membranes, the fetal heart was not heard. Twenty four hours later delivery of a dead fetus occurred following an oxytocin infusion.

Necropsy Findings

A white male fetus, placenta and membranes were received for pathological study. The weight of the fetus was 630 g, its measurements were as follows: crown-heel length, 290 mm; crown-rump

Send offprint requests to: Jorge Vercelli-Retta, M.D., Dpto. de Anatomía Patológica, Hospital de Clínicas, P.B., Avda. Italia s/n., Montevideo, Uruguay, South America

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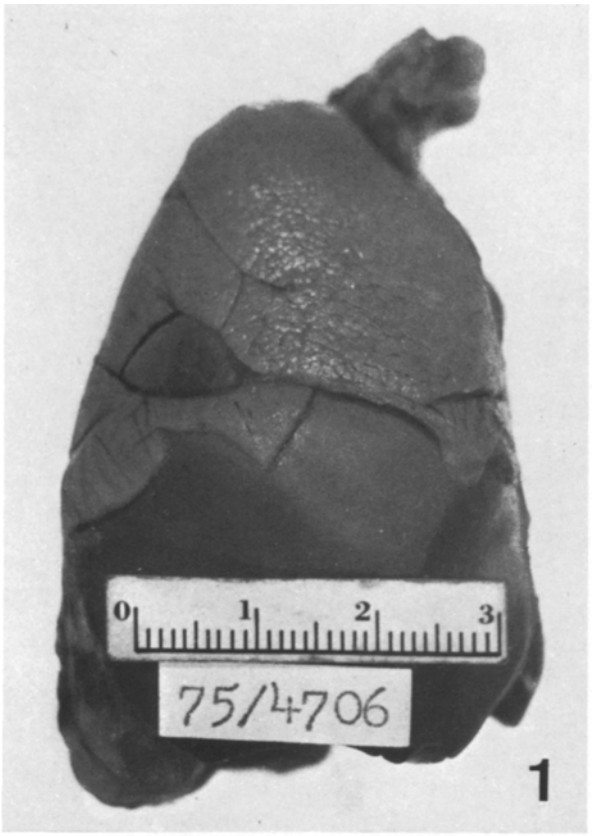

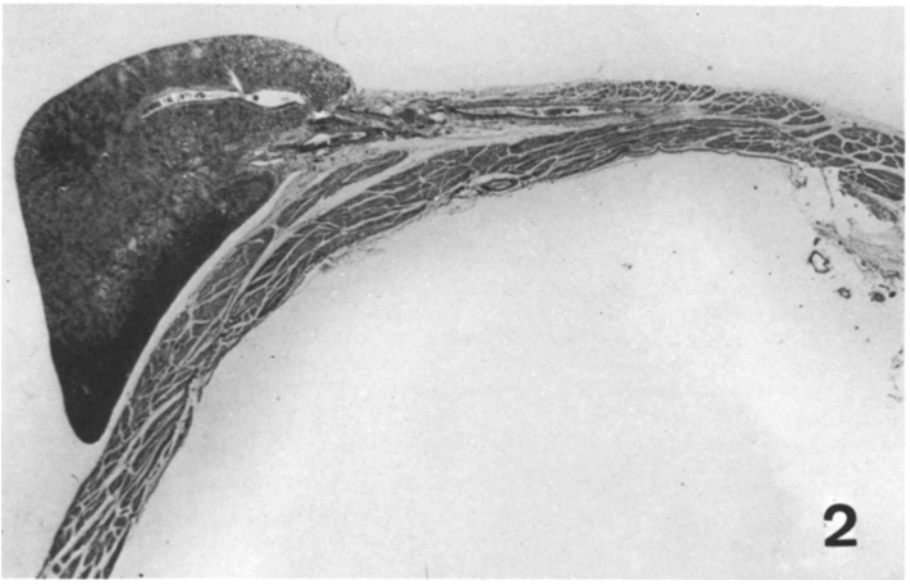

Fig. 1. Fetal visceral block showing the right lung and the abdominal liver. Among these organs and lying over the diaphragmatic fragments, there is a triangular, lateral mass corresponding to the accessory thoracic liver lobe

Fig. 2. Histological section of the hepatic accessory lobe with its joining pedicle entering the normal diaphragm. Some vessels cross the peduncle and disappear among the muscle bundles. A cross section of the falciform ligament showing some vessels is seen near the ending of the accessory pedicle. (H and E stain, $\times 9$)

length, 175 mm; occipitofrontal circumference, 210 mm. Gross examination revealed some degree of maceration without external malformation. Dissection of the central nervous system showed extensive ventricular haemorrhage. In the thoracic cavity a triangular mass beneath the anterior border of the inferior lobe of the right lung was found lying above the respective hemidiaphragm (Fig. 1). It was of pyramidal shape measuring $9 \times 5 \times 4$ mm. The mass had a sharply encapsulated smooth surface and a firm consistency that clearly differentiated it from the lung. The inner inferior angle of this tumor-like formation adhered to the diaphragmatic convexity by a 3 mm long and 2 mm thick pedicle, which apparently entered the muscle. The diaphragm was normal, having neither hernias nor any hiatus in its surface. In the abdominal cavity the liver was normally developed and its weight was 35.7 g.

The remaining viscera showed no abnormal features. The placenta weighed 256 g. The umbilical cord was 36 cm long and had three vessels.

Histological examination of the thoracic malformation showed typical hepatic tissue. Broad portal spaces contain bile ducts with very narrow lumina, some venules and occasionally an arteriole within the abundant connective tissue stroma. The hepatic lobules were of normal form and had well preserved cellular trabeculeae around sinusoids containing numerous nests of hemopoietic cells.

The study of the aberrant hepatic peduncle showed an artery, a large vein and several transverse and longitudinal sections of bile ducts. These elements disappeared within the diaphragm. Serial sections (one of each five sections of 4 μm thick were mounted and studied) did not show the subsequent course of these vascular and canalicular elements. Some veins, lymphatic vessels and nerve fibres were shown to cross the falciform ligament. The histological appearance of the diaphragmatic muscle was normal (Fig. 2). The surface of the liver mass and the pedicle were lined with flat cells similar to those of the pleura. Microscopic study of the orthotopic liver did not show abnormalities.

Apart from the ventricular haemorrhage all other organs, the placenta and the umbilical cord were histologically normal and their development was in accordance with the fetal age.

Comments

This is an exceedingly rare finding. There are no cases of supradiaphragmatic ectopic liver lobes reported in fetuses in the literature (Wacker, 1963; Potter and Craig, 1976). Seven cases of well documented thoracic accessory liver lobes have been reported in adults (Rodríguez-Pérez, 1955; Hansbrough and Lipin, 1957; Kaufman and Madoff, 1960; Le Roux, 1961; Hudson and Brown, 1962; Caron et al., 1970; Sehdeva and Logan, 1971) (Table 1). The malformation has no sex predilection. Three typical sites for the anomaly have been described, the right costophrenic variety being the most frequent. A left supradiaphragmatic liver lobe and another case located close to the inferior vena cava have been reported. All these observations were made incidentally by radiography and clinically a neoplasm was usually diagnosed.

The present case was an incidental autopsy finding. The lack of bile accumulation in the liver sections suggests that a communication between the ectopic pedicle and the biliary tree of the abdominal liver may have existed. From microscopic study it may be suggested that this communication was via the falciform ligament. This was not demonstrated macroscopically and the ligament was cut during the dissection of the abdominal liver, thus histological examination was incomplete.

Although this type of hepatic malformation has not been described in association with human chromosome abnormalities (Carr, 1975), it is not possible to discard a cytogenetic defect in the present case.

Table 1. Thoracic accessory liver lobes: review of the literature[a]

Author	Year reported	Patient age and sex		Clinical data	Surgical or pathology findings
Rodriguez-Pérez	1955	15	F	Routine chest X-ray: right supradiaphragmatic mass, 6 cm diameter	Pedunculated hepatic lobe
Hansbrough and Lipin	1957	26	M	Ulcerative colitis. Chest X-ray: right posterior costophrenic tumor	Liver lobe (137 g). Pedicle crossing a diaphragmatic hiatus. Hepatic fibrosis
Kaufman and Madoff	1960	48	F	Rheumatic heart disease. Recent cerebral embolus. Chest X-ray shows right costophrenic mass	Liver lobe placed in the neighborhood of inf. vena cava. Diaphragmatic hiatus with joining pedicle to the abdominal liver
Le Roux	1961	18	M	Routine chest X-ray, tumor of the right costophrenic angle	Liver lobe with a pedicle joined with the abdominal liver
Hudson and Brown	1962	21	M	Routine chest X-ray, right supradiaphragmatic tumor	Pedunculated liver lobe
Caron et al.	1970	26	F	Chest X-ray: right costodiaphragmatic mass	Pedunculated liver lobe of 8×6 cm, slightly nodular surface
Sehdeva and Logan	1971	21	F	Intermittent left lower chest pain. Chest X-ray, left supradiaphragmatic mass	Pedunculated, 4 cm, left thoracic liver lobe. No diaphragmatic defects

[a] Only cases with a normal diaphragmatic muscle are included

There will be no satisfactory embryological explanation of this liver malformation until an early stage has been found in an embryo. Some authors (Hansbrough and Lipin, 1957) have postulated that the diaphragmatic closure pinches a liver bud which then develops above the muscle. Chouke (1932) suggested that islands of hepatic tissue can be carried along the primitive veins. This hypothesis appears to be more convincing since the complexity and characteristic anatomic variations of veins could also explain the variety of sites of hepatic thoracic and extrathoracic heterotopic lobes.

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