MESENCHYMAL HAMARTOMA OF THE LIVER IN ADULTS – CASE REPORT

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Introduction
Mesenchymal hamartoma of the liver is a benign lesion presenting as an enlarging abdominal mass in children less than 2 years of age. Fewer than 5% cases are present in individuals over 5 years of age, and this lesion is extremely rare in adults. It may affect the left or the right lobe of liver as a cystic or solid mass or both components may be present. The pathogenesis remains incompletely understood, but these lesions have generally been considered to represent a development abnormality in the bile duct plate formation. Case Report. In this report, we present a case of a 44-year-old man who was surgically treated at the Department of Abdominal, Endocrine and Transplantation Surgery of the Clinical Center of Vojvodina due to cystic lesion in the liver segment IV that had been verified by computed tomography imaging diagnostics. The patient was sent from a smaller health center with the diagnosis of echinococcosis. After the adequate preparation of the patient, surgical excision of the liver cystic lesion was done. Once a thorough histological examination had been performed, the diagnosis of mesenchymal hamartoma was made. Conclusion. Mesenchymal hamartoma of the liver is a benign tumor resulting from abnormal, intra-uterine development of bile ducts and has a delayed clinical manifestation, thus this lesion appears to be related to the processes of maturation. It is potentially premalignant lesion presenting as a solid and/or cystic neoplasm. Symptoms, laboratory results and radiographic imaging are nonspecific and inconclusive, so surgical excision of the whole lesion is the imperative for the definitive diagnosis.

Key words: Liver Neoplasms; Hamartoma; Adult; Diagnosis; Tomography, X-Ray Computed; Hepatectomy

Summary
Introduction. Mesenchymal hamartoma of the liver is a benign lesion presenting as an enlarging abdominal mass in children less than 2 years of age. Fewer than 5% cases are present in individuals over 5 years of age, and this lesion is extremely rare in adults. It may affect the left or the right lobe of liver as a cystic or solid mass or both components may be present. The pathogenesis remains incompletely understood, but these lesions have generally been considered to represent a development abnormality in the bile duct plate formation. Case Report. In this report, we present a case of a 44-year-old man who was surgically treated at the Department of Abdominal, Endocrine and Transplantation Surgery of the Clinical Center of Vojvodina due to cystic lesion in the liver segment IV that had been verified by computed tomography imaging diagnostics. The patient was sent from a smaller health center with the diagnosis of echinococcosis. After the adequate preparation of the patient, surgical excision of the liver cystic lesion was done. Once a thorough histological examination had been performed, the diagnosis of mesenchymal hamartoma was made. Conclusion. Mesenchymal hamartoma of the liver is a benign tumor resulting from abnormal, intra-uterine development of bile ducts and has a delayed clinical manifestation, thus this lesion appears to be related to the processes of maturation. It is potentially premalignant lesion presenting as a solid and/or cystic neoplasm. Symptoms, laboratory results and radiographic imaging are nonspecific and inconclusive, so surgical excision of the whole lesion is the imperative for the definitive diagnosis. Key words: Liver Neoplasms; Hamartoma; Adult; Diagnosis; Tomography, X-Ray Computed; Hepatectomy

Sažetak
Mondson was the first to use this name of the tumor in 1956 [4]. Mesenchymal hamartoma of the liver can be found in the left or right lobe either as a cystic or solid mass, although both components may be present. Concerning localization and structure, pediatric and adult populations have different characteristics. Mesenchymal hamartoma of the liver is more common in the left liver lobe in children, while in adults it is equally common in the left (about 40%) and the right lobe of liver (40%), and in about 20% of cases, tumor is in both lobes [5]. It has been noticed that mesenchymal hamartoma of the liver in male patients is more common in the right lobe, while in women it is more common in the left lobe of liver, and cases when the tumor is in both lobes is more frequent in women. From the histological point of view, mesenchymal hamartoma of the liver consists of epithelial and stromal components. The epithelial component consists of hepatocytes looking relatively normally and bile ducts, both being surrounded by myxoid and fibrous stroma. Hepatocytes are arranged in groups of different sizes maintaining architectonics of cell plates as in a normal liver. Bile ducts have typical branching layout and they are frequently surrounded by acute inflammatory infiltrate in the wall of bile duct. Cystic spaces, if there are any, can be coated by thinned to cuboidal epithelium or they can be nonepithelialized and surrounded by loose or dense fibrous stroma. Stroma contains a large number of vascular spaces, spindle-shaped and inflammatory cells. Normal ports are not present. In adults, stroma is more fibrous and more densely hyalinized by only focal myxoid areas. In some cases, mesenchymal component can be dominant in a lesion with infrequent ductal elements [6]. The pathogenesis of mesenchymal hamartoma of the liver remains incompletely understood, but one can say that these lesions have generally been considered to represent a development abnormality in the bile duct plate formation. Several cytogenetic studies have suggested that this tumor can appear with the creation of embryonic sarcoma of the liver [7].

Case Report

A 44-year old male patient was admitted to the Department of Abdominal, Endocrine and Transplantation Surgery of the Clinical Center of Vojvodina for the surgical treatment of a change in liver that had been verified by computed tomography (CT) imaging diagnostic in another institution. The CT image attached hereby showed that there was a relatively sharply encapsulated focal hypodense lesion in the liver segment IV, 29 x 31 x 35 mm in size with internally localized hyperdense zone 5 mm thick. Right next to the described change, subcapsularly localized, there was a hypodense area 13 x 19 mm in size, hence the patient was diagnosed with echinococcus cyst.

**Figure 1.** A change on the IV segment of the liver - hypodense lesion 29x31x35 mm in size with ring hyperdense zone and subcapsular hypodense zone

**Slika 1.** Promena IV segmenta jetre - hipodenzna leziija dimenzija 29x31x35 mm sa prstenastom hiperdenznom i subkapsularnom hipodenznom zonom

**Figure 2.** A change on the IV segment of the liver – hypodense lesion 29x31x35 mm in size with ring hyperdense zone and subcapsular hypodense zone

**Slika 2.** Promena IV segmenta jetre - hipodenzna leziija dimenzija 29x31x35 mm sa prstenastom hiperdenznom i subkapsularnom hipodenznom zonom
The patient claimed to have had a numb pain in the stomach region under the right costral arch for the previous six months, but denied any other discomforts as well as any other hereditary diseases. After the clinical examination, laboratory tests were conducted which indicated serological negativity, therefore the referral diagnosis was rejected and the working diagnosis of tumor in the liver segment IV with indicated surgical treatment was made.

The laboratory affiliated to the Institute of Pathology and Histology of the Clinical Center of Vojvodina received operative material, a fragment of liver parenchyma, triangular in shape, 6.5 x 3.5 x 3 cm in size with dark pink, blurred capsule. A cystic formation 3.5 cm in diameter with yellowish, smooth inner surface surrounded by the liver parenchyma about 0.2 cm wide was seen on the intersection subcapsularly.

After classical material processing, histological preparations of cyst formations were obtained. Detailed pathohistological examination verified the existence of cystically dilated bile duct which was coated by focally multiplied cylindrical epithelial whose lumen contained a large amount of bile. Around the cystically dilated bile duct, there was a prominent connective tissue containing multiplied and partly dilated bile ducts (Figure 3). In addition to the described bile ducts, there were intersections of blood vessels with thickened wall and peripheral nerves (figures 4, 5, and 6). The proliferated connective tissue was partly and unclearly separated from the surrounding liver parenchyma which had appropriate histomorphological characteristics. The diagnosis of mesenchymal hamartoma of the liver was based on the typical morphological appearance of the change. In addition to the operative material,
the gallbladder, whose pathohistological finding indicated chronic cholecystitis, was received.

The postoperative period and hospital treatment were uneventful so the patient was discharged after five days for home care and recovery with advice to have examinations and monitoring regularly.

Discussion

Mesenchymal hamartoma of the liver accounts for about 8% of all tumor changes in pediatric age group. The majority (about 80%) of cases are diagnosed before 2 years of age. Mesenchymal hamartoma of the liver can rarely appear in an adult liver, but there are only a few case reports in adults, more frequently in women [7]. The clinical picture is different and it depends on the age of patients. The most common clinical picture of pediatric patients is painless enlargement of the abdomen usually noticed by parents. If the enlargement of the abdomen is significant or liver function is compromised, there could be ascites and/or jaundice. In some cases, there could be compression of the diaphragm or lungs with consequential respiratory disorders. Adult patients are usually asymptomatic, although some authors have given case reports of patients who complained about discomforts such as diffuse abdominal pain [7], and some have presented patients who, in addition to diffuse abdominal pain, had clinical signs and symptoms of hepatomegaly, pain in the right hypochondrium and in the left upper quadrant as well [8]. Since the appearance of tumor can vary from cystic to completely solid mass as well as a combination of both components, there is no consensus on which form of tumor is more common. Some authors have reported results which show higher frequency of cystic form in children [2], whereas others claim quite the opposite, that is they have found a higher frequency of solid mass tumor in pediatric age group [5]. The situation with adult patients is similar. Chau et al. [5] have reported higher frequency of cystic tumors, and Hernandez et al. [9] have recorded higher frequency of cystic tumors in both adult and pediatric patients. It is believed that women are more prone to developing cystic tumors, and for men there is no tendency to any kind of tumor. However, it has been noticed that if there is a case of tumor which has both cystic and solid component, then it is more frequent in women.

Because of its non-specificity, mesenchymal hamartoma of the liver is extremely hard to be diagnosed by laboratory or other methods. In laboratory tests, alkaline phosphatase, β-human chorionic gonadotropin (HCG), serum transaminases as well as the α-fetoprotein are mostly within normal range in these patients. Radiograph of mesenchymal hamartoma can vary from predominantly cystic to completely solid change. If it is a cystic mass, differential diagnosis suggests echinococcus cyst and hepatic abscess in the first place [10]. Ultrasonography is usually the first in line of the radiological evaluation of the change to show the suspected liver mass to be either cystic or solid as well as its dimensions. CT imaging is the next step to show anechoogenic cystic change with thin partitions. If a contrast is applied, a solid change will show; however, a cystic component does not show on contrast CT [11]. Thus, all imaging methods provide a nonspecific finding of mesenchymal hamartoma of the liver. A differential diagnosis allows various pathological processes in the liver such as a simple liver cyst, hydatid cyst, biliary cystadenocarcinoma or cystic metastases. If it is a solid mass, what follows is differential diagnosis of focal nodular hyperplasia, hepatic adenoma, cavernous hemangioma, angiomylipoma as well as hepatocellular carcinoma. One should also take into consideration Von Meyenburgov complex (biliary hamartoma) which is histologically manifested by multiplying bile ducts surrounded by a more or less connective tissue which can be found as tiny nods 1-2 mm in diameter scattered on the liver parenchyma [12]. In most cases, the definitive diagnosis of mesenchymal hamartoma of the liver is made after surgical excision of the lesion, and extremely rarely by needle biopsy.

Since the tumor has the potential for massive growth as well as the potential for malignant alteration into embryonal sarcoma or angiosarcoma, the treatment of mesenchymal hamartoma of the liver is surgical excision of the tumor in operable patients. Although we live in an era of highly developed laparoscopic surgery, the gold standard in the treatment of mesenchymal hamartoma of the liver is liver resection with negative margins [1].

Conclusion

Mesenchymal hamartoma of the liver is a benign tumor resulting from abnormal, intra-uterine development of bile ducts and has a delayed clinical manifestation, thus this lesion seems to be related to the processes of maturation. It is potentially premalignant lesion presenting as a solid and/or cystic neoplasm. It predominantly appears in children before 2 years of age and it is extremely rare in adult population, which happened to be in our case. The clinical picture as well as laboratory and radiological findings are nonspecific. Needle biopsy is not a diagnostic method, therefore, surgical excision of the lesion and detailed pathohistological analysis are the imperative for the therapy and the definitive diagnosis.
References