Abstract

Hepatic hemangiomas are the most common hepatic lesion. They have a female predominance in all age groups, are often asymptomatic, and are increasingly found incidentally on abdominal ultrasound (US) or computer tomographic (CT) examinations. Hepatic hemangiomas can be single or multiple and are vascular malformations that grow by ectasia rather than neoplasia. Large hemangiomas can produce a variety of symptoms from discomfort to life-threatening complications. The complications are rare but include compression of adjacent organs, rupture, and Kasabach-Merritt syndrome. Magnetic resonance imaging (MRI) is the best imaging method to establish the diagnosis. Treatment modalities for giant hepatic hemangiomas, such as arterial ligation, radiotherapy, and corticosteroid and interferon therapy, have been implemented without any convincing success. Surgical resection is so far the only consistently curative method of treatment. Indications for performing surgery of hepatic hemangiomas should be severe unacceptable symptoms, growth of the tumor, inability to exclude malignancy, persons with a high risk of hepatic injury, and complications. If surgery is indicated, different techniques can be used, including enucleation, hepatic resection, and laparoscopic techniques. When the diagnosis of hepatic lesions is uncertain despite thorough radiological evaluation and biochemical analysis, percutaneous biopsy is recommended, and the latest open laparotomy with biopsy and/or resection. The authors believe that considered hepatic resection of benign lesions in the liver in appropriately chosen patients is a safe procedure. A report of a special case of a giant liver hemangioma and a literature review on the diagnosis and treatment of hemangiomas of the liver are presented (Adv Clin Exp Med 2009, 18, 1, 7–12).

Key words: hepatic hemangioma, giant, surgical treatment, complications.
can mimic other benign and malignant hepatic lesions such as focal nodular hyperplasia (FNH), hepatic adenoma (HA), hepatocellular carcinoma (HCC), and metastasis [6]. This article presents a special case report of a giant liver hemangioma and provides a literature review on the diagnosis and treatment of hemangiomas of the liver.

Case Report

A 42-year-old female presented with four months of feeling discomfort in the right subcostal region and radiating lumbar pain. Physical examination revealed an enlarged liver. All laboratory findings were normal. Percutaneous needle biopsy did not establish a diagnosis. Later that year exploratory laparotomy at the local hospital was performed and a huge reddish tumor was found in the right liver lobe. Peroperative biopsies showed no malignancy, thus no conclusive diagnosis was reached. The tumor was at the time considered inoperable because of its size and location. External radiation downsized the tumor somewhat, though the tumor still disabled the patient severely.

Almost twelve years later, a CT scan and US with biopsy and hepatic angiography established the diagnosis of cavernous hemangioma. Since the appearance of symptoms the liver had grown substantially and the patient’s symptoms had worsened. She was transferred to the present authors’ surgical department, a reference center for hepatic surgery, where the patient complained of a large, protruding, painful, tender abdominal mass as well as dyspnea and severe immobility due to varicose veins on edematous legs. The patient also had developed cardiac problems due to reduced backflow from the inferior vena cava. Physical examination revealed a large knotty mass in the the abdomen extending into the right lower quadrant. Blood chemistry showed unspcific elevated liver tests. Esophagogastroduodenoscopy showed a discrete esophageal varix. US showed the liver monstrously enlarged and the porta hepatis displaced 10 cm to the left. Liver segments 5–8 were replaced by a tumor of varying echogenicity. Sequential CT scans confirmed the US findings and also showed small areas of calcification, a compressed vena cava, and a recanalized umbilical vein due to high portal venous pressure. Tc-99m colloid and Tc-99m red blood cell angiography was typical of a hemangioma.

At operation the abdomen was opened with a wide bilateral subcostal incision which later was extended into the right chest. A 40-cm-large tumor was found in the right liver lobe. The tumor was densely adherent to adjacent structures and organs. The left liver lobe was compensatively hypertrophied. The liver was mobilized and a right hemihepatectomy could be performed by classic anatomical resection. To secure a safe division of the right hepatic bile duct, a small bougie was inserted into the left hepatic duct. The operation lasted 10 hours and required 18 units of blood. The hemangioma weighed 7.5 kg and measured $24 \times 14 \times 14$ cm after fixation. Microscopy showed a cavernous hemangioma intermingled with areas of fibrosis and organized hematomas. The postoperative course was without major complications. The patient has been followed postoperatively at her local hospital once a year.

Discussion

Giant hepatic hemangioma is histologically a benign disease, but literature [7] and the case presented here show that very large hemangiomas may cripple the patient. Macroscopically they are often well-circumscribed, reddish, hypervascular, compressible lesions. Histologically the tumors reveal large blood-filled spaces lined by endothelial cells and separated by thin fibrous septa [5]. In general, the blood circulation within these tumor vessels is slow [3]. Most hemangiomas are probably of congenital rather than of neoplastic origin [3, 5] and there are no reports of malignant transformation. They grow by ectasia rather than by hypertrophy or hyperplasia. Hemangiomas in the liver can be solitary or multiple [8]. The tumors are sharply demarcated from the surrounding liver tissue and are rarely calcified. They vary in size, those larger than 4 cm in diameter being referred to as giant hemangiomas. Hemangiomas in the liver can be combined with hemangiomas in other organs. Hepatic hemangiomas are usually located peripherally or are subcapsular, mostly in the posterior segments of the right liver lobe [3].

Hepatic hemangiomas are the most common hepatic tumor. Hemangiomas have a female predominance in all age groups, but are most frequently found in the third, fourth, and fifth decades of life [9, 10]. In more than 50% of cases, hemangiomas are multiple [3]. Hemangiomas have unknown etiology, although studies have suggested a possible relationship with the intake of steroidal hormones such as estrogen [11].

The two largest hemangiomas ever published weighed about 18 kilograms. One was found incidentally at autopsy in 1918 and another was successfully resected in China and reported anonymously in the Chinese Medical Journal in 1979 [12, 13]. Several studies have reported that the
The majority of hepatic hemangiomas remain stable in size and that complications are relatively rare [14, 15]. Farges et al. [16] reported in their series of 163 patients with a mean follow up of 92 months that only nine hemangiomas increased in size while seven decreased.

As with malignant tumors, hepatic hemangiomas are usually clinically silent until they reach a considerable size [5]. Pain is related to the size of the hepatic hemangioma [10, 16, 17]. One study concluded that hemangiomas in the right liver lobe tended to be larger than those in the left liver lobe, which could indicate that tumors in the right liver lobe grow until they produce discomfort or symptoms. Discomforting symptoms such as pain, nausea, vomiting, and weight loss can be caused by the tumor’s compression of adjacent organs [5]. In a series of 87 patients with hepatic hemangioma, 54% of the patients’ gastrointestinal symptoms had causes other than the hepatic hemangioma [16]. The exact mechanism of hepatic-induced pain is not fully understood. It is known that hemorrhage, infarction, and thrombosis can cause pain [5], possibly due to distension of Glisson’s liver capsule.

The risk of life-threatening complications such as spontaneous rupture is low. Yamamoto et al. [18] showed in 1991 that only 28 cases of spontaneous rupture of a liver hemangioma had been published, which must be considered a low number taking into account the high prevalence of hepatic hemangiomas. Hepatic hemangiomas are therefore rarely the direct cause of intra-abdominal hemorrhage. However, even though it is a rare complication, hemorrhage should always be taken into consideration, as the mortality rate at spontaneous rupture is almost 60% [19]. In case of rupture, the treatment is acute surgery or, alternatively, vascular embolization if surgery cannot be performed [5]. Elective liver resection should be saved for patients with unacceptable pain, compression of adjacent organs, Kasabach-Merritt syndrome, or suspicion of malignancy [10].

Abdominal US examinations are performed more often today than earlier and, with the increasing use of imaging modalities such as CT and MRI, more hepatic nodules and tumors are being identified incidentally [5]. Diagnostic modalities consist of Doppler US, spiral CT, MRI, technetium-99m red blood cell scintigraphy, and angiography, which all demonstrate characteristic vascular findings with high sensitivity and specificity. Hepatic hemangiomas normally have characteristic radiologic features that make it possible to distinguish
them from other hepatic nodules. On US, the most common feature is a homogeneous hyperechoic liver nodule without any Doppler signal [3]. Depending on the size, the appearance of hepatic hemangiomas in a US scan can be variable, which limits its use. An inconclusive US examination is an indication for performing CT [2].

On CT, hepatic hemangiomas are often shown as hypodense well-defined lesions with early peripheral enhancement and slowly centripetal filling after administration of an iodine contrast medium [3]. This pattern is seen in about 50% of hepatic hemangiomas. The typical characteristic sign of hemangiomas has a positive predictive value of greater than 80% [20]. Leslie et al. report that in differentiating between hepatic hemangiomas and hypervascular metastases, contrast-enhanced CT scans have a sensitivity of 88% and a specificity of 84–100% [21].

SPECT (Tc-99m-labeled red blood cells) has a sensitivity of 97% and a specificity of 83% in suspecting hepatic hemangiomas larger than 1 cm [22]. When the specific location is in doubt, it can be useful to use a combination of SPECT and CT. If hepatic hemangiomas are small they have an atypical appearance and enhancement pattern on CT, which is why it may be necessary to perform MRI [23].

MRI is currently the method of choice in the diagnostics of hepatic hemangiomas [24, 25] and is believed to be superior to CT [26]. On an MRI scan with a combination of T2-weighted images and serial gadolinium-enhanced gradient echo imaging, hepatic hemangiomas are homogeneous, clearly demarcated, and have a very high signal intensity [3, 24]. The features considered pathognomonic on MRI are well-defined lobular margins, T2-weighted hyperintensity, and progressive nodular centripetal enhancement [24]. It is possible to suggest criteria that indicate benign tumors: size less than 4 cm, discrepancy in radiologic diagnosis, and stability in size over time [5, 27].

Several studies indicate a high risk of bleeding when percutaneous needle biopsy is performed for diagnostic reasons and recommend an operative procedure when the diagnosis is undetermined [16, 28]. The present authors disagree that an operative procedure is recommended with an undetermined diagnosis; they perform percutaneous US-guided needle biopsy routinely without any complications.

There is still debate regarding the treatment of liver hemangiomas. The majority of hepatic hemangiomas are asymptomatic and can be managed with observation alone [2]. Management options range from observation with regular US examinations to a variety of radiological and surgical interventions. Arterial ligation, radiotherapy, and corticosteroid and interferon therapy have been implemented without any convincing success [2]. So far, surgical resection is the only curative treatment and is indicated in patients in whom the surgical risk is assessed as acceptable and the symptoms unacceptable [8]. Estrogen therapy may induce growth of hemangiomas [11]. Herman et al. [10] emphasized, as other authors [16, 29], that tumor size is not a definite indication for surgery as long as pain and discomfort are tolerably managed with analgesics. Operative indications can be changed so that persons with an occupational risk of hepatic injury, such as boxers and football players, may be candidates for surgery earlier [30].

If surgery is indicated, different techniques can be used, including enucleation, hepatic resection, and laparoscopic techniques. The first enucleation was performed in 1988 by Alper et al. [31]. The obvious advantages of enucleation are less operative blood loss and a reduced risk of biliary leakage [32]. One report state that enucleation is the surgical procedure of choice [5, 33]. Some authors advocate enucleation [31, 32, 34] and others liver resection [35, 36]. The choice of procedure should also be based on tumor location and the number of hemangiomas. If the hemangioma is located deep within the liver parenchyma or in posterior hepatic segments, a formal anatomical resection should be the preferred operative procedure [2]. In specialized centers, liver resection mortality has declined to rates lower than 3% [10]. Laparoscopic surgery should be reserved for minor hepatic resections and wedge resections [37].

Conclusions

Indications for surgery in hepatic hemangiomas are severe unacceptable symptoms, growth of the tumor, inability to exclude malignancy, and persons with a high risk of hepatic injury and complications. The size of the hemangioma should not be the only indication for surgery. When the diagnosis is uncertain despite thorough radiological evaluation and biochemical analysis, percutaneous biopsy, and at the latest open laparotomy with biopsy and/or resection, are recommended.
References


Address for correspondence:
Flemming Burcharth
Department of Surgical Gastroenterology
Herlev Hospital
University of Copenhagen
DK-2730 Herlev, Denmark
Tel.: +45 404 647 07
E-mail: flbu@heh.regionh.dk

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