INTRODUCTION

Hemangiomas are the most frequent benign hepatic tumors, being found in about 7% of cases in necropsy studies. They are more prevalent in women (2.5 times more often than men) in the age group between 30 and 50 years. The liver is their main localization; the right lobe is involved in 60% of hemangiomas and the left in 25%. Sometimes more than one tumor can develop. The liver most often develops two or three tumors, and multiple hemangiomas can occur in the same organ.

LIVER HEMANGIOMAS: ULTRASOUND AND CLINICAL FEATURES*

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Abstract

Hemangiomas are the most common benign tumors of the liver, occurring in all age groups, and more frequently in adults. The vast majority of hemangiomas are small, asymptomatic, and are incidentally discovered. Larger lesions may eventually produce symptoms. The sonographic aspect of these tumors varies, the lesions being typically small, well defined and hyperechoic. In this study the authors review clinical and sonographic features of hemangiomas, highlighting the clinical significance of such features to be taken into consideration in the treatment of affected patients.

Keywords: Hemangioma; Liver; Ultrasound.

INTRODUCTION

Hemangiomas are the most frequent benign hepatic tumors, being found in about 7% of cases in necropsy studies. In the literature, a study reports an even higher occurrence — 25% of the cases analyzed. In the present paper, the authors evaluate clinical and ultrasonographic aspects of these tumors, highlighting the significance of combining such aspects to achieve a correct diagnosis and an adequate treatment of patients.

DISCUSSION

Typically, hemangiomas are single, with less than 5 cm in diameter (Figure 1), but two or more lesions (Figure 2) may be found in 10% to 30% of patients. They occur in all age ranges, more frequently in adults in the third, fourth and fifth decades of life. Women would be more frequently affected than men. In the Mayo Clinic series, 66% of the cases were women. Hemangiomas should be more appropriately studied since, usually, they do not require specific treatment. Although some authors report an even distribution in both hepatic lobes, others consider that hemangiomas would tend to localize more superficially.
Figure 3. Predominantly hyperechogenic giant hemangioma presenting heterogeneous echogenicity, with anechoic central anechoic areas.

Figure 4. Hyperechogenic giant hemangioma.

Figure 5. Hemangioma with an anechoic area representing cystic degeneration.

and in the posterior segment of the right hepatic lobe (6).

These lesions, also, may be called “giant” (Figure 3), although there is not a consensus regarding the minimum diameter of a giant hemangioma (Figure 4). Several authors consider as giant the lesions measuring more than 4 cm, 6 cm and 8 cm (7), yet others say that giant hemangiomas would be those > 10 cm (3).

In some cases, hemangiomas present with peculiar characteristics. Some lesions may present cystic degeneration (Figure 5), many times centrally located, varying in extent, and, in some situations present like a lesion containing a significant cystic component (cystic hemangioma) (8). Areas of necrosis also may be observed, as well as central fibrotic areas. These aspects — cystic alterations, necrosis and fibrotic alterations — are more frequently found in larger lesions (usually those > 4 cm) (9).

Another unusual occurrence is the fast growth of these lesions, which usually does not suggest neoplastic transformation, but rather ectasia of the preexistent vessels (10), associated with necrotic and hemorrhagic phenomena. Also, a rare possibility of infection in hemangiomas has been reported (11). Some lesions may present themselves pedunculated (6).

Usually, hemangiomas are asymptomatic, with less than 50% of patients presenting clinical manifestations (12), frequently associated with non-specific abdominal symptoms, like pain in the epigastrium and right hypochondrium, besides sensation of weight in the upper abdomen. Didactically, one can say that < 4 cm hemangiomas are rarely symptomatic. It is in cases of larger lesions and on physical examination that we can observe clinical manifestations. In larger lesions, different degrees of hepatomegaly may be found. Generally, when there are specific symptoms, chronic or acute abdominal pain is observed. Although the origin of such pain has not completely cleared up yet, it is suggested that it is caused by the growth of the lesion, resulting in distention of the Glisson’s capsule. More rarely, these lesions may associate with obstructive jaundice and alterations of hepatic enzymes (13), besides gastric obstruction, torsion (in cases of pedunculated lesions) and spontaneous rupture (14).

According some authors, the incidence of hemangiomas would be higher in multiparous women (15); also, the lesions could increase in size during the gestation (15,16). Other sporadic reports mention the possibility of an increase in these lesions in women receiving exogenous estrogen (17). These relations should be understood as occasional occurrences; however they give the opportunity of discussing a possible relation between female hormones and these tumors (7).

Other manifestation reported in the literature is the association of cavernous hemangiomas with thrombocytopenia and hypofibrinogenemia, due to probable consumption coagulopathy and platelets trapping — the called Kasabach-Merrit syndrome. This syndrome has been originally described as an association between thrombocytopenia, afibrinogenemia (hypofibrinogenemia) and skin and spleen hemangiomas in children. However, the term has currently been utilized to designate cases of consumption coagulopathy and platelets trapping associated with hepatic hemangiomas. Usually, this syndrome is found in
children, and is rare in adults\(^{(1)}\). Also, in children, congestive heart failure may occur as a result of arteriovenous fistulas in the hemangiomas, especially in the large ones\(^{(18)}\).

Regarding the hepatic hemangiomas evolution, Trastek \textit{et al.}\(^{(19)}\) have analyzed 36 patients with hepatic hemangiomas during a period of up to 15 years (mean = five years and a half). These authors have not observed any case with bleeding, increase in clinical discomfort or death of patients. In four patients, the lesions increased, and in three, decreased. In the remaining patients, the lesions did not present any alteration in size.

Occasionally, hemangiomas may change their echogenicity with a change in decubitus (Figure 6) or with a Valsalva maneuver. Another important aspect to be taken into consideration is that hemangiomas may present inflammatory alterations, fibrosis and thrombosis, resulting in higher rigidity of some lesions. Cases with long standing thromboses may suffer calcification\(^{(6)}\).

Hemangiomas, also, may be associated with other hepatic lesions like cysts, hepatocellular adenomas (Figure 7) and focal nodular hyperplasia, as well as Rendu-Osler-Weber syndrome\(^{(6)}\). In some studies, hemangiomas have been found in up to 25% of cases of focal nodular hyperplasia\(^{(20,21)}\). Another association reported, although rare, is that between hepatic hemangiomas and hemangiosarcomas\(^{(10,22)}\).

At ultrasound, frequently these lesion present as hyperechogenic (Figure 8) and well defined, homogeneous images, especially those with < 3–4 cm (Figure 9)\(^{(23–25)}\). The presence of a small, central, hypoechogenic area also may be observed. The hyperechogenicity probably results from multiple interfaces between vascular spaces. Additionally, a posterior acoustic enhancement may be demonstrated in some lesions\(^{(26)}\). It is important to note that, in steatotic livers, hemangiomas may frequently present hypoechogenic (Figure 10).

More heterogeneous aspects may be identified, especially in larger lesions. Sometimes, now more, now less echogenic areas representing fibrosis or ectatic vascular spaces. Some authors have observed these more heterogeneous aspects, especially in lesions with > 8 cm\(^{(7)}\).

There are reports on other findings like presence of central, hypoechogenic areas dominating the aspect of lesions in a variable extent (Figure 11); others present predominantly hypoechogenic with a possibly hyperechogenic peripheral margin (Figure 12). Such hyperechogenic peripheral margin found in these hemangiomas, is a quite useful sign for differentiating these lesions from other hypoechogenic masses in the
liver\textsuperscript{(24)}. When hemangiomas have this more heterogeneous aspect, the differentiation from other focal hepatic lesions of the liver becomes difficult.

The lesion may present with lobulated margins\textsuperscript{(18)}. According to some authors, calcifications would be unusual\textsuperscript{(27)}, however other authors report their presence in up to 20\% of cases\textsuperscript{(28,29)}. It is important to note that, usually, hemangiomas do not present any hypoechogenic halo around the lesion\textsuperscript{(30)}, which also helps to differentiate them from other focal hepatic lesions.

Ultrasound contrast enhancement (microbubbles) also may demonstrate the vascular behavior of these lesions. Prolonged and moderate periods of contrast agent (microbubbles) retention inside the lesions have been reported\textsuperscript{(31)}.

Doppler ultrasound present variable results in cases of hemangiomas. In many cases, no intralesional sign is detected (Figure 13), because of the low velocity of the blood flow in this lesions. In other cases, sparse venous signs are detected inside hemangiomas (Figure 14), presenting a hypointense and continuous spectral pattern (Figure 15)\textsuperscript{(32)}.

As previously mentioned, the greatest majority of these lesions do not demand specific treatment, with the possible exception of those patients with bulging lesions and chronic, debilitating symptoms. Cases of hemorrhage resulting from the rupture of these tumors, especially in neonates, are reported in the literature. However, such cases are extremely rare, particularly in adults\textsuperscript{(5)}.

Figure 9. Multiple hepatic hemangioma. Hyperechogenic images.

Figure 10. Hypoechogenic hepatic hemangiomas in steatotic livers.

Figure 11. Hepatic hemangioma with central hypoechogenicity dominating the aspect of the lesion.

Figure 12. Hepatic hemangiomas presenting central hypoechogenic area and hyperechogenic periphery.
Hemangiomas frequently do not cause compression of adjacent vessels (portal and hepatic venous branches and hepatic arterial branches), adapting themselves to these structures, without causing significant compression. However, it is important to note that larger lesions, as well as those presenting fibrohemorrhagic alterations, sometimes cause compressive disorders in the surrounding vascular structures.

Still, regarding vascular tumors in children, we must describe the infantile hemangioma and pediatric hemangiomas, since they represent the most frequent benign vascular tumors in this age range. Some aspects of hemangiomas deserve special considerations. A higher tendency to rupture in neonates and infants than in adults has been reported. These tumors may produce high output heart failure because of arteriovenous fistulas, a phenomenon that has not been observed in adults.

Microangiopathic hemolytic anemia, thrombocytopenia and hypofibrinogenemia are rare manifestations, occurring more frequently in the pediatric group than in adults. The association of thrombocytopenia and hypofibrinogenemia is called Kasabach-Merrit syndrome. In the child, these lesions are associated with skin hemangiomas in up to 50% of patients. Many cases of death in children occur because of associated heart failure. When observed in infants and neonates, these hemangiomas are different from those found in adults and are rarely identified in children. Hemangiomas in neonates and infants are mesenchymal masses, presenting active endothelial growth. Some authors call them hemangioendotheliomas. These tumors may reach large dimensions.

Usually, children affected by hemangiomas or hemangioendotheliomas are less than six years old, and most of them are diagnosed before six months of age. Clinically, infantile hemangiomas and hemangioendotheliomas are characterized by a set of three entities: hepatomegaly, congestive heart failure and skin hemangiomas. Kasabach-Merrit syndrome may occur both in cases of hemangioendotheliomas and hemangiomas affecting infants. Frequently, an abdominal mass is observed at physical examination. High output heart failure attributed to arteriovenous shunts (anastomosis) in the liver, is an usual cause of death. There are studies demonstrating that the natural history of these tumors may course to spontaneous regression. As previously mentioned, according to some authors there is a possibility of rupture determining the patient death. Although usually hemangioendotheliomas are histologically benign lesions, malignant degeneration of these tumors has been described.

Ultrasound demonstrates the presence of a single or multiple lesion with variable echogenicity. Its aspect is solid, however, permeated cystic (anechoic) areas may be observed. Calcifications also may be present. The diagnosis of these benign vascular tumors of the liver in children may be corroborated by the evidence of alterations in hepatic arteries and veins, as well as in the celiac trunk. The celiac trunk and the common hepatic artery may be dilated, while the caliber of the aorta is decreased below the origin of the celiac trunk. Additionally, the caliber of hepatic veins draining the lesion may be increasing. The above described changes in the vessels caliber are not identified as features of pediatric malignant hepatic tumors and are quite suggestive of benign vascular tumors (hemangiomas and infantile hemangioendotheliomas).

**CONCLUSION**

The authors conclude that hemangiomas are relatively frequent benign tumors whose usual aspect is that of well circumscribed, hyperechogenic lesions. However, there are variations in sonographic presentations, and one should be always alert to these possibilities which should be taken into consideration in the differential diag-
nosis of hepatic masses, besides guiding supplementary evaluations for a conclusive diagnosis.

REFERENCES