

Atypical giant hepatic hemangioma. A case report

Hemangioma hepático atípico gigante. Presentación de caso

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Summary

Giant hepatic hemangiomas are lesions included in the atypical hemangiomas category. In most cases they have a benign behavior, predominantly present in women with ages ranging from 20 to 60, and are commonly located in the right liver lobe with a size larger than 4 cm. The diagnosis is usually made incidentally because they generally don´t produce any symptoms; however, when they are large they tend to manifest persistent abdominal symptoms. In images, among the most common findings in ultrasonography are: hyperechoic lesions with peripheral feeding vessels and posterior acoustic reinforcement; on CT the most frequent findings are heterogeneous lesions with a markedly hypoattenuating central area that has nodular peripheral enhancement in the arterial phase after intravenous contrast administration; some atypical findings include hemorrhage, thrombus, calcifications, capsules or pseudocapsules. Although most of these lesions only require observation, some patients require treatment, either surgical (enucleation, resection) or interventional radiology procedures such as angiographic selective embolization.

Key words (MeSH)

Hemangioma
Tomography
Interventional radiology
Liver

Palabras clave (DeCS)

Hemangioma
Tomografía
Radiología intervencionista
Hígado

Resumen

Los hemangiomas hepáticos gigantes son lesiones contempladas dentro del grupo de hemangiomas atípicos. En su mayoría, son benignos, predominan en mujeres de 20 a 60 años de edad, tienen un tamaño mayor a 4 cm y comúnmente están localizados en el lóbulo hepático derecho. Su diagnóstico suele ser incidental debido a que no causan síntomas; sin embargo, cuando son de gran tamaño tienden a manifestar sintomatología abdominal persistente. En imágenes, entre los hallazgos más comunes en ultrasonografía se encuentran: lesiones hiperecoicas, vasos de alimentación periféricos y refuerzo acústico posterior. En tomografía axial computarizada (TAC), los hallazgos más frecuentes son la visualización de lesiones heterogéneas con un área central marcadamente hipoatenuante y realce nodular periférico en fase arterial tras la introducción de medio de contraste intravenoso; los hallazgos atípicos son hemorragias, trombosis, calcificaciones, cápsulas o pseudocápsulas. Aunque la mayoría de estas lesiones solo requieren observación, algunos pacientes requieren tratamiento, ya sea quirúrgico (enucleación, resección) o medidas intervencionistas, como la embolización selectiva por angiografía.

Introduction

Hemangiomas are the most common benign hepatic vascular lesions among liver tumors. They have an incidence of 73% and a prevalence of 1% to 20% in the general population; the female to male ratio varies between 2:1 and 5:1 (1). Giant hemangiomas of the liver are notably less frequent, representing up to 10% of all hepatic hemangiomas (2); these are defined as larger lesions when they are 4 cm in diameter with atypical findings in the images; although other authors consider them as larger lesions when they are 6 to 10 cm in diameter, which given their large size can cause hepatomegaly, mass effect, cough and abdominal discomfort in up to 27% of the cases (2, 3). Their origin is congenital mesenchymal, consisting of blood-filled cavities fed by the hepatic arterial circulation, lined with a wall of endothelial cells, generally of peripheral location (4).

The differentiation of atypical giant hemangiomas from other hypervascular tumors can be challenging because not all of them show a characteristic pattern; moreover, some hypervascular tumors can simulate a peripheral nodular enhancement similar to that of hemangiomas (5), making their diagnosis a challenge for radiologists.

Different imaging methods can be used to determine this pathology, such as ultrasonography, computed tomography (CT) and magnetic resonance imaging (MRI); likewise, CT angiography plays an important role, given its high preoperative value.

Although hemangiomas usually do not require treatment, only follow-up, there is a small group of them that can become malignant, even complicated

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with hemorrhage or thrombosis; in addition, if they are large, they can cause symptoms such as abdominal pain, and sometimes need to be treated by surgical procedures (6).

Clinical case presentation

A 40-year-old female patient with no relevant medical history consulted the emergency department due to clinical symptoms of abdominal pain in the right hypochondrium, of 15 days of evolution, intensified in the last 4 hours. A total abdominal ultrasound was performed (Figures 1a, 1b) which showed a large lesion of 10 cm in length in its largest diameter, in the left hepatic lobe, highly vascularized (Figure 1b). The extension paraclinical tests with hemogram, bilirubin, renal function tests (BUN and creatinine), liver function and electrolytes (sodium, potassium and chlorine) showed no evidence of alterations. Subsequently, an abdominal CT scan was performed in 3 phases (Figures 2 and 3) for a better characterization of the lesion, which showed peripheral nodular filling, centripetal in arterial and portal phase, with progressive nodular relace and absence of central filling in persistent venous phase, indicative of giant atypical hepatic hemangioma. Given the patient's symptomatology, subselective hepatic transarterial embolization was performed using a microcatheter with coaxial passage in the main artery (left hepatic artery) through which embolizing material (microparticles) was administered (Figure 4). The procedure was performed without complications. The patient is currently in good health and prognosis is favorable..

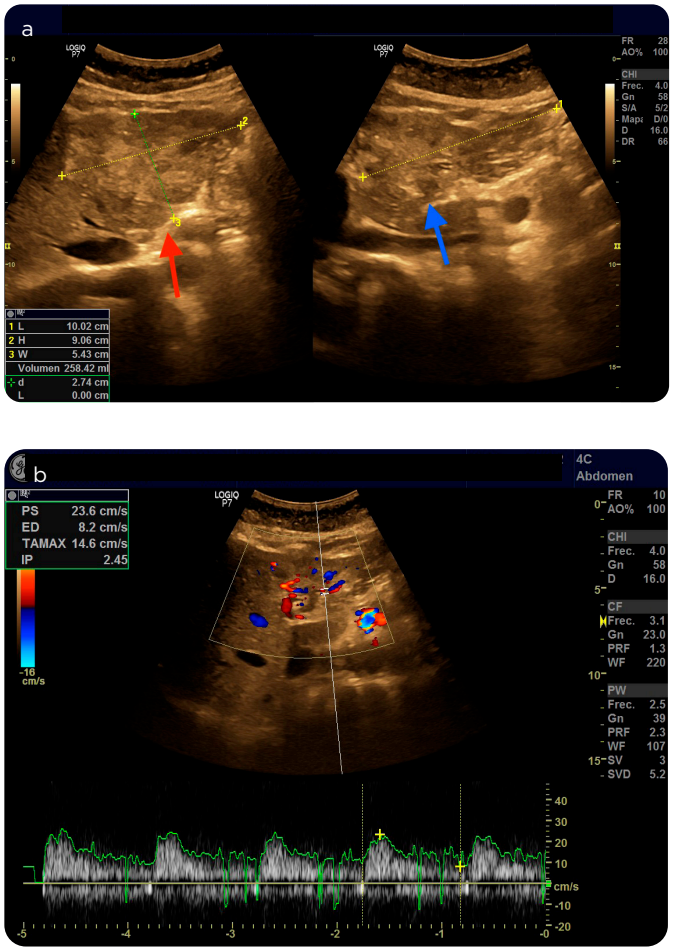


Figure 1. a) Axial and longitudinal sections of total abdomen ultrasonography in B-mode. Heterogeneous predominantly hyperechoic image with lobulated borders, (red arrow in axial section, blue arrow in longitudinal section), in the left hepatic lobe that mainly occupies segments II and III, with a maximum diameter of 10 cm. b) Color Doppler scan and spectral analysis evidences peripheral feeding vessels with low velocities.

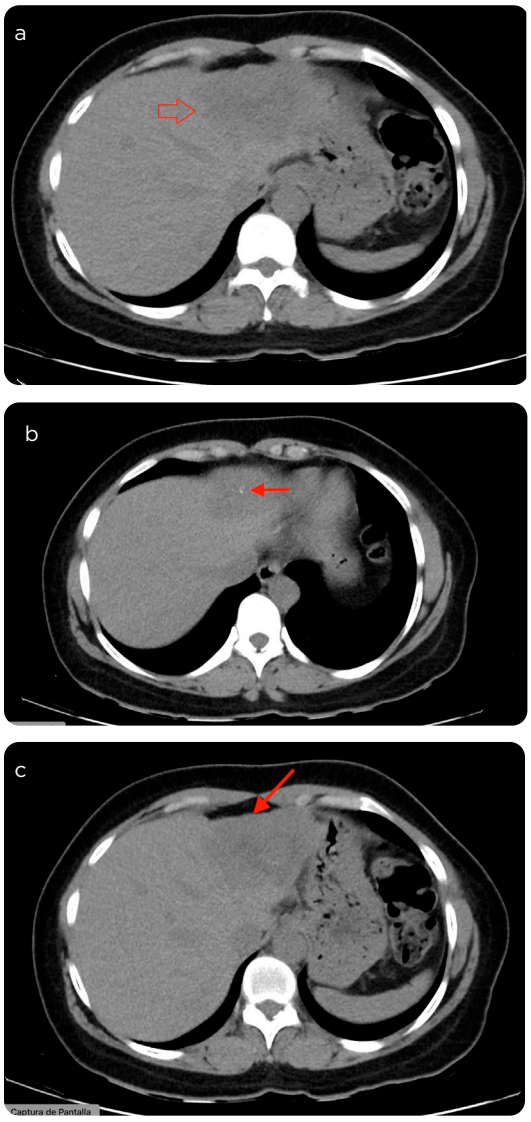


Figure 2. a-c) Tomography in axial axis window of the abdomen in single phase. Image (red arrow in a) lobulated hypoattenuating with respect to the hepatic parenchyma in the left hepatic lobe (segments I-II) with punctate calcifications (red arrow in b), with lobulation of the hepatic contour and hepatic retraction (red arrow in c).

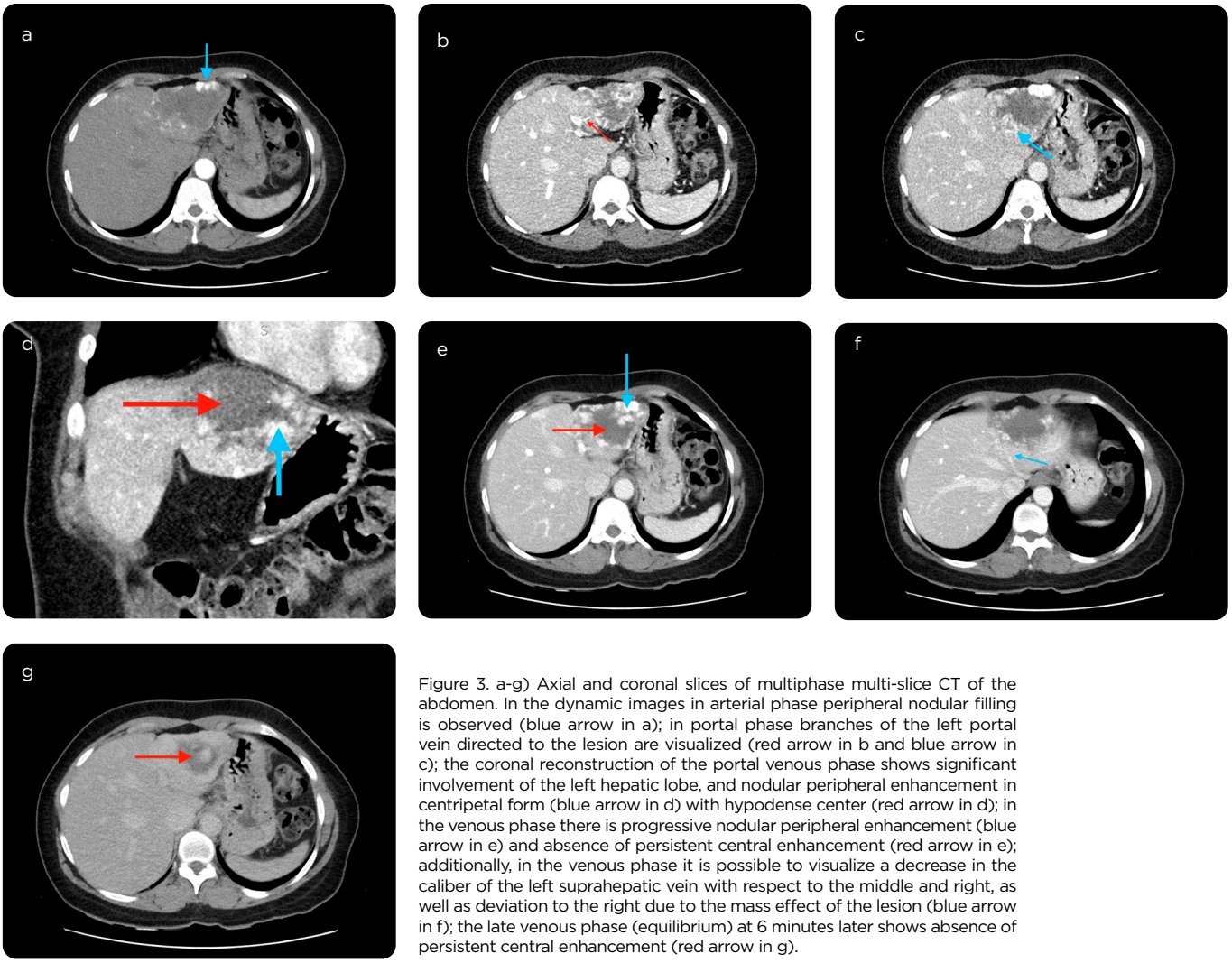


Figure 3. a-g) Axial and coronal slices of multiphase multi-slice CT of the abdomen. In the dynamic images in arterial phase peripheral nodular filling is observed (blue arrow in a); in portal phase branches of the left portal vein directed to the lesion are visualized (red arrow in b and blue arrow in c); the coronal reconstruction of the portal venous phase shows significant involvement of the left hepatic lobe, and nodular peripheral enhancement in centripetal form (blue arrow in d) with hypodense center (red arrow in d); in the venous phase there is progressive nodular peripheral enhancement (blue arrow in e) and absence of persistent central enhancement (red arrow in e); additionally, in the venous phase it is possible to visualize a decrease in the caliber of the left suprahepatic vein with respect to the middle and right, as well as deviation to the right due to the mass effect of the lesion (blue arrow in f); the late venous phase (equilibrium) at 6 minutes later shows absence of persistent central enhancement (red arrow in g).

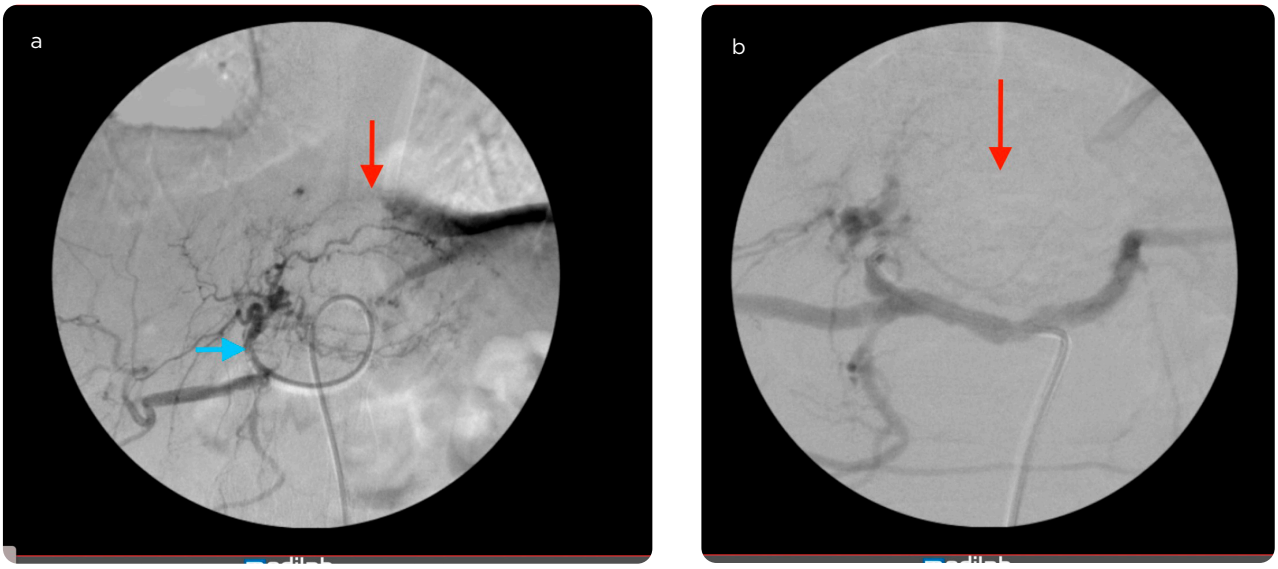


Figure 4. a and b) Selective angiography of the left hepatic artery (blue arrow in a) shows evidence of a large, vascularized lesion in the left hepatic lobe (red arrow in a); digital subtraction angiography images after subselective hepatic transarterial embolization showed a significant decrease in arterial flow, without visualization of intralesional capillaries (red arrow in b).

Discussion

Liver lesions are a diagnostic challenge, especially hemangiomas, when they present atypical characteristics; in ultrasound the frequent pattern are hypoechoic lesions with peripheral echogenic halo and important color Doppler flow, its less frequent pattern are heterogeneous lesions that can reach a large size of up to 12 cm. In CT they can be seen as hypodense and heterogeneous images with progressive enhancement from the periphery to the central region. Up to 20% of hemangiomas have atypical characteristics (giant size, sudden filling, calcifications, sclerosis, cystic-necrotic degeneration, pedunculated, with liquid-liquid level inside) (1).

MRI can be particularly accurate to diagnose a hepatic hemangioma. Even without contrast, hemangiomas have a pathognomonic appearance on MRI in most cases. On MRI, these tumors are characterized as homogeneous, well-demarcated lesions with very high signal intensity on T2-weighted images with markedly hyperintense cleft-like area and some hypointense internal septa (1).

This case was characterized by a large lesion with lobulated borders, hypodense with respect to the hepatic parenchyma, with portal and systemic feeding vessels, nodular peripheral enhancement without central enhancement, and some calcifications in its interior; mainly, what allowed the diagnostic orientation was the behavior of the lesion in the dynamic tomography images after the administration of contrast medium, the absence of underlying liver disease and the size of the lesion.

Among the main differential diagnoses of giant atypical hemangiomas are malignant lesions, such as hypervascular liver metastases. The pattern of nodular enhancement in contrast studies, characteristic of hemangiomas, has a sensitivity of 88% and a specificity ranging from 84 to 100% to differentiate hepatic hemangioma from hypervascular metastases, which present an early ring enhancement from the periphery to the center, and the classic peripheral hypodense halo due to compression of the hepatic parenchyma present in malignant lesions; on the other hand, there is hepatocellular carcinoma, which shows important enhancement in arterial phase with its classic central washout and persistence of capsular enhancement in portal venous phases; angiosarcomas must also be considered in these, which are usually multiple and with nodular enhancement (7).

Another important, but less frequent diagnosis that must be taken into account is focal nodular hyperplasia that appears in CT as a hypodense or isodense lesion in portal phase, characterized by its central hypervascular scar, which in certain occasions is associated with atypical hepatic hemangioma given their vascular nature (3).

Among the most important complications of atypical hepatic hemangiomas are rupture and hemorrhage, which are observed after trauma or biopsy, respectively.

The hemangioma is a soft tumor and usually does not compress adjacent structures. However, thrombosis or internal hemorrhage can transform the lesion into a solid mass and compress biliary and vascular structures, such as the inferior vena cava and portal vein, causing perfusion abnormalities and fatal complications (3).

Hepatic hemangiomas are generally asymptomatic and therefore only require observation, although some may require surgical management. Their main indications are: abdominal pain, mass effect and Kasabach-Merritt syndrome (2). There are multiple treatments under study, including surgical techniques (enucleation, resection), for some hepatic hemangiomas; however, the treatment of multiple and/or giant hepatic hemangiomas continues to be an important clinical challenge, for which, in certain cases, the first option is not surgical management, but coadjuvant interventional management (8).

Interventional procedures with selective angiographic embolization should be performed only in cases of symptomatic hemangiomas, hemangiomas that grow progressively or hemangiomas with a high risk of bleeding (8).

Embolization should be as selective as possible in the branches that feed the lesion to reduce intraoperative bleeding and tumor growth, with embolizing material according to the radiologist's experience (gelfoam, coils, polyvinyl alcohol and isobutyl cyanoacrylate). First, peripheral embolization of vessels within the lesion should be performed with gelfoam or polyvinyl alcohol, followed by embolization of the main arteries with coils (8).

In this case, interventional management with supraselective transarterial embolization using microparticles was decided, since the symptomatology of persistent abdominal pain and compression of vascular structure (suprahepatic vein) were clear indications for the procedure. Among the most important contraindications are coagulation disorders, concomitant treatment with antiaggregants and/or anticoagulants and allergy to iodinated contrast medium; none of them in this case.

Control images showed a significant decrease in the arterial flow of the lesion, without visualization of the previously identified capillaries. The most common complications of this intervention are pain, pyrexia, nausea and leukocytosis, which last a few days (8); however, in this case there were no complications.

Conclusion

In summary, giant atypical hemangiomas are lesions larger than 4 cm, with radiological characteristics that must be taken into account for an accurate diagnosis (progressive peripheral nodular enhancement that in some cases may not present central filling, and necrotic cystic changes). Additionally, it is important to identify the potentially fatal complications that may occur due to their size, such as compression of adjacent structures. Interventional radiology has a boom in management through selective embolization of the lesion, which avoids the complications of a major surgical procedure and effectively reduces intralesional vascularization, with a consequent decrease in lesion size and improvement of symptoms. However, an individualized evaluation of the patient will always be necessary to choose the most appropriate treatment in each case.

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