



Right Hepatectomy for Giant Hepatic Hemangioma With Progressive Growth in a Case of Relative Hyperestrogenism[☆]

Hepatectomía derecha por hemangioma hepático gigante de crecimiento progresivo en un contexto de hiperestrogenismo relativo

Hemangiomas are the most frequent benign liver tumors, with an incidence of up to 20% in the general population. They are mostly single lesions that are smaller than 1 cm, asymptomatic and stable over time, although occasionally they may grow and cause the symptoms of space-occupying lesions. When they surpass 4 cm in size, they are known as “giant hemangiomas”. Although the causes of their growth are unknown, it has been suggested that estrogen may play an important role.

We present the case of a 38-year-old male, with a history of partial androgen insensitivity syndrome that was diagnosed because of an ambiguous phenotype during childhood, hypospadias and cryptorchidism. The patient was studied due to recurring hepatic colic, at which time he was diagnosed with a hepatic hemangioma measuring 13 cm in diameter in the right liver lobe. As the patient was asymptomatic, periodic follow-up studies were scheduled. Over the course of 4 years, the lesion was observed to grow in size to 21 cm, and another 6-cm hemangioma appeared in the caudate lobe, at which time he was sent to our unit. Because the patient was asymptomatic, required major surgery and had a history of partial androgen insensitivity syndrome, we decided to initiate treatment with tamoxifen. Although initially the growth was slower (for 3 years), a slight increase in the size of the lesion was detected. Given the volume of the lesion and its proximity to the suprahepatic veins (only the left suprahepatic vein was permeable 0–5 cm from the lesion) and compression of the vena cava (Fig. 1), we decided to perform surgical exeresis and considered the possibility of radical hepatectomy. After preparation of the patient for a possible bypass, in the end a right hepatectomy was performed with exeresis of the caudate lobe, without requiring bypass.

The surgical specimen weighed 3800 g (Fig. 2), and that of the hepatic remnant was 1.8% of the patient’s body weight. The pathology study revealed a cavernous hemangioma measuring 22 cm and another measuring 5 cm in the caudate lobe. The postoperative period was uneventful, and the patient was discharged on the 10th day post-op.

Hepatic hemangiomas are benign lesions whose pathology has not been fully determined. Originating in the mesodermal layer, they are considered congenital vascular malformations that grow through ectasia and not by hyperplasia. Their incidence is greater between the third and fifth decades of life, and they are more common in

women. Generally, they are single lesions (although they can present in both lobes [10%]) that are most frequently found in the right lobe.^{1–3} Their growth is slow and has been associated with hormonal factors such as exposure to high levels of estrogen and progesterone, such as in multiple births, pregnancy and oral contraception. This theory, however, is based on case reports, and estrogen receptors have not been demonstrated in all hemangiomas.⁴

In general, they are asymptomatic lesions and, therefore, the diagnosis is usually incidental. Nonetheless, patients may sometimes present symptoms that are directly related with the size of the lesion and compression of the adjacent structures. These symptoms can vary from abdominal pain, compression of local structures, thrombosis and lesion infarction to hemoperitoneum due to tumor rupture, which is exceptional.^{1,2}

Experimental studies have demonstrated that selective estrogen receptor modulators, like tamoxifen, could control the growth rate of hemangiomas; however, as stated before, not all hemangiomas present hormone receptors, so this therapy is not 100% effective. The case we present is unique because of the hormonal situation of hyperestrogenism secondary to partial androgen insensitivity syndrome, for which treatment with tamoxifen was indicated. During its use, less growth was observed, but the proximity to the suprahepatic veins guided our decision to operate.⁵

Although surgery is still the only curative treatment for hemangiomas, exeresis is only justified in symptomatic patients, with established complications, rapid growth or uncertain diagnosis (since biopsies are contraindicated due to the risk of hemorrhage). In asymptomatic patients, periodical radiological follow-up is the best option.^{2,6}

Since the first resection of a hepatic hemangioma by Hermann Pfannenstiel in 1898, the surgical technique has evolved. Currently, enucleation is the preferred technique as it offers better preservation of the hepatic parenchyma and fewer complications. Other techniques described are segmental resection, ligation of the hepatic artery and liver transplantation in cases of Kasabach-Merritt syndrome (intravascular disseminated coagulation).⁷

In the case that we present, right hepatectomy was performed as the anatomical resection seemed safer and the part of the parenchyma to be resected was minimal, given the atrophy of the right liver and hypertrophy of the left. Furthermore, due to the proximity of the hemangioma to the cava and the need for prolonged clamping, cannulation was

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Fig. 1 – Abdominal CT scan showing the size of the mass occupying practically the entire right liver as well as the small lesion of the caudate lobe.



Fig. 2 – Surgical specimen–right hepatectomy.

planned and performed for a possible venovenous bypass and vascular exclusion. In the end, the exclusion was not necessary.⁸

There are reports of postoperative morbidity rates between 10%–27% and mortality rates of 0%–2%,² which demonstrate the safety of the technique. However, as these lesions are benign, we believe that surgery should only be used in select cases after having carefully evaluated the surgical benefits and any possible postoperative complications.

Authors' Contribution

All the authors have made a contribution to this article, have read and approved of the manuscript, and have complied with the requirements for authorship.

Conflicts of Interests

The authors have no conflicts of interests.

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