Histologically, it is easy to make the distinction between glomus tumors and hemangiomas or angiosarcomas. Hepatocellular carcinoma and hepatoblastoma are ruled out due to the low levels of alpha-fetoprotein. The negativity of desmin in conjunction with the histologic distribution rules out a vascular leiomioma. Glomus tumors from any part of the body can metastasize to the liver. Nonetheless, in our patient, no other primary tumor was found.

Despite having completely resected the tumor, and due to its uncertain malignant potential, the patient is currently in appropriate clinical follow-up with periodical studies.

We conclude that glomus tumors should be considered in the differential diagnosis of liver tumors with positive immunohistochemistry for SMA, which has been shown to be a useful diagnostic method.

**Authorship**

- Study design: FNF and JAJR;
- Data collection: ECA and NSV;
- Analysis and interpretation of the results: FNF, ECA and NSV;
- Article composition: ECA and NSV;
- Critical review and approval of the final version: FNF and JAJR.

**REFERENCES**


Enrique Calcerrada Alises,* Natalia Sarabia Valverde, Francisco Navarro Freire, José Antonio Jiménez Ríos

*Corresponding author.
E-mail address: enrique_alises@hotmail.com
(E. Calcerrada Alises).

2017;5(4) 232–241

© 2016 AEC. Published by Elsevier España, S.L.U. All rights reserved.

**Intrahepatic Masson Tumor (Intravascular Papillary Endothelial Hyperplasia)**

Tumor de Masson intrahepático (hiperplasia endotelial papilar intravascular)

Intravascular papillary endothelial hyperplasia (IPEH), or Masson’s tumor, is a rare benign intravascular lesion. It is a reactive process in the context of venous stasis, in which there is a proliferation of endothelial papillary structures that are organized around thrombi.1-3

* Please cite this article as: Ramallo Solis I, Tinoco González J, Senent Boza A, Bernal Bellido C, Gómez Bravo MA. Tumor de Masson intrahepático (hiperplasia endotelial papilar intravascular). Cir Esp. 2017;95:235–237.
It was first described in 1923 by Masson as an endothelial papillary hyperplasia in the lumen of hemorrhoidal veins, and was considered a vascular neoplasm, termed “vegetant intravascular hemangioendothelioma”. In 1932, Henschen observed that it is a reactive process, not an endothelial neoplasm, therefore renaming it *L'endovasculite proliférante thrombopoiétique*. In 1976, Clearkin and Enzinger suggested that thrombosis precedes endothelial proliferation, and the thrombotic material constitutes a matrix for its development. After several studies, it was verified that the lesion was a vascular proliferation secondary to venous stasis. In 1990 and based on immunohistochemical studies, Albrecht and Kahn described a similar progression between endothelial hyperplasia and thrombosis. Both are positive for ferritin, histiocytic markers and, eventually, vimentin in the early stages, while at the end of the process they are only positive for factor VIII antigen.

Masson's tumors mainly affect the head and neck vessels, fingers, trunk and skin veins, while abdominal involvement is exceptional. Only one case of hepatic involvement has been reported in the literature.

We present the case of a 39-year-old male, with no medical history of interest, who was asymptomatic. During routine examination, hepatomegaly was detected upon abdominal palpation; ultrasound demonstrated a 10 cm lesion suggestive of hemangioma.

The study was completed with an abdominal CT scan, MRI and tumor marker study. The CT scan showed a lesion measuring 10×9.5×7 cm occupying segments VI and VII, which was hypodense and had lobed edges. Contrast uptake by the peripheral nodes and centripetal filling, without complete filling of the lesion, were suggestive of hemangioma (Fig. 1). MRI detected a lesion the same size as CT, with lobed edges and central areas of necrosis; peripheral uptake and centripetal filling were compatible with hemangioma. No pathological tumor marker alterations were observed (Fig. 2).

During follow-up, the patient presented abdominal discomfort secondary to the lesion size, although there were no apparent changes to the lesion. After assessing the case in committee, surgery was indicated, and an atypical segmentectomy was conducted of segments VI and VII. The postoperative period transpired without incident; the patient progressed favorably and was discharged on the 5th postoperative day. During outpatient follow-up, there were no incidents in the first year after surgery.

The pathology report identified a 170 g fragment measuring 12×9 cm that, after serial sections, was identified as a poorly defined, sponge-like, purplish subcapsular lesion measuring 10×7 cm, inside of which a 4×3 cm whitish, well-defined fleshy area was found. Microscopically, a benign vascular process was identified with endothelial proliferation in the small and medium-sized vessel lumens, compatible with Masson's tumor.

Intravascular papillary endothelial hyperplasia is a rare benign condition requiring differential diagnosis, primarily with hemangiosarcoma. Initially, it was considered a malignant disease, and Masson described it as a neoplasm secondary to the degeneration of a venous thrombosis. However, after several studies, it was demonstrated that Masson tumors consist of a proliferation of endothelial cells that are organized around thrombi, thereby becoming established as a reactive process in the context of venous stasis.

Histopathologically, 3 forms have been described: the “pure” form, which is most frequent, appears de novo in the dilated vascular spaces in patients without comorbidities and without vascular abnormalities; the “mixed” form, which appears in vessels with abnormalities such as arteriovenous malformations, hemangiomas, pyogenic granulomatoses and chronic diseases such as paroxysmal nocturnal hemoglobinuria; and, finally, the “extravascular” form, which is associated with trauma-related hematomas.

The development of intravascular papillary endothelial hyperplasia takes place in several stages. In the early stages, there is endothelial cell growth within the thrombus. These cells begin to proliferate and secrete collagenases, which partially and irregularly digest the thrombus, developing papillary structures. Finally, these papillary structures combine to form anastomosed vascular structures.

Fig. 1 – Triple-phase CT showing the lesion in segments VI and VII measuring 10×9.5×7 cm with contrast uptake in the peripheral nodes and centripetal filling, suggestive of hemangioma.

Fig. 2 – MRI: lesion with lobed edges and central areas of 19×9.5×7 cm, located in segment VI and VII.
The most frequent locations are the vessels of the head and neck, fingers, trunk and cutaneous veins. Intra-abdominal lesions are rare, and even rarer are those in the liver.

Epidemiologically, a slightly higher incidence has been observed in women than in men, with a ratio of 1.2:1, with no predilection for age. Cases have been reported in patients ranging from 7 months to 81 years of age.

Intravascular papillary endothelial hyperplasia is an uncommon disease that can simulate other diseases. It is therefore necessary to carry out a detailed differential diagnosis, and the definitive diagnosis is often reached by the pathology study.

Treatment of IPEH depends on its location and, considering that it is a benign disease, the symptoms that it causes. In general, surgical resection is the treatment of choice for IPEH as it provides a very good prognosis and very low recurrence rate (mainly in the mixed and extravascular forms). Surgery is considered curative in the pure forms when the margins of the surgical specimen are free, with low reported rates of recurrence in the mixed and extravascular forms.

### Funding

The authors have received no public or private funding for the completion of this article.

### References


Irene Ramallo Solís, José Tinoco González, Ana Senent Boza, Carmen Bernal Bellido, Miguel Ángel Gómez Bravo

Unidad de Cirugía Hepatobiliopancreática y Trasplante Hepático, Hospital Universitario Virgen del Rocío, Sevilla, Spain

*Corresponding author.*

E-mail address: irene.ramallo@gmail.com (I. Ramallo Solís).

2173-5077/ © 2016 AEC. Published by Elsevier España, S.L.U. All rights reserved.

---

Spontaneous Pulmonary Hernia: Presentation of Four Cases and Literature Review☆☆

Hernia pulmonar espontánea: presentación de 4 casos y revisión de la literatura

Pulmonary hernias are characterized by a protrusion of the lung through a defect of the chest wall. Herniations of the lung can be either acquired or spontaneous. Predisposing factors have been described, such as obesity, chronic obstructive pulmonary disease (COPD) or the chronic use of corticosteroids. We present the cases of 4 patients with spontaneous lung hernias treated at our hospital in 2015. The first case is that of a 60-year-old patient who was overweight and had a history of COPD. In February, he reported the appearance of a hematoma on the right thoracic wall, and a CT scan diagnosed a right pulmonary herniation between the 8th and 9th ribs. By thoracotomy, primary closure was performed with non-absorbable sutures. In April, he was diagnosed with pleural effusion compatible with empyema.

---


☆☆ The data published in this article have been previously presented as an oral communication at the Spanish National Thoracic Surgery Conference, (SECT), May 5, 2016.