



Available online at  
**ScienceDirect**  
[www.sciencedirect.com](http://www.sciencedirect.com)

Elsevier Masson France  
**EM|consulte**  
[www.em-consulte.com/en](http://www.em-consulte.com/en)



## IMAGE OF THE MONTH

# Multicystic schwannoma of the porta hepatis

Najat Mourra<sup>a,\*</sup>, Nikias Colignon<sup>b</sup>, Ghazi Zaatari<sup>c</sup>

<sup>a</sup> Department of Pathology, Hôpital Saint-Antoine, AP-HP, Paris, France

<sup>b</sup> Radiology Department, Hôpital Saint-Antoine, APHP, Paris, France

<sup>c</sup> Pathology Department, American University, Beyrouth, Lebanon

### KEYWORDS

Porta Hepatis Tumor;  
Schwannoma;  
Hepatic tumors

A previously healthy 67-year-old woman complained of epigastric fullness for several months which has recently increased. Laboratory analyses were normal, as well as upper and lower gastrointestinal endoscopy. Abdominal Magnetic Resonance Imaging (MRI) and Computed Tomography (CT) revealed a 6 cm cystic lesion, involving hepatic segment I. As the metastatic work-up was negative, the patient was referred to our hospital for surgery.

Intraoperatively, the lesion was extrahepatic, arising from the porta hepatis, and connected to the peri-hepatic artery tissue. After careful dissection, the lesion was removed “*en-bloc*” and sent for histopathological examination.

Review of MRI at our institution showed a well-defined retrohepatic tumor, composed of a solid component slightly hyperintense on T2-weighted images and cystic areas hyperintense on T2-weighted images. Axial gadolinium-enhanced T1-weighted fat suppressed MRI showed significant enhancement of the solid component in the late phase. Enhanced CT scan in a coronal oblique reconstruction showed a well-

defined tumor in the porta hepatis, with solid and cystic components. This mass was clearly extra hepatic, with an extrinsic compression and displacement of the portal vein (Fig. 1A).

The postoperative recovery was uneventful and the patient is still alive and well year later.

The Fig. 1B shows a gross section of this globoid mass which appears grey at the periphery, with wide bright yellow zones and cystic degenerations in the center. The Fig. 2A shows a high power view hematoxylin-eosin stained sections of the mass, and Fig. 2B an immunohistochemical stain of it.

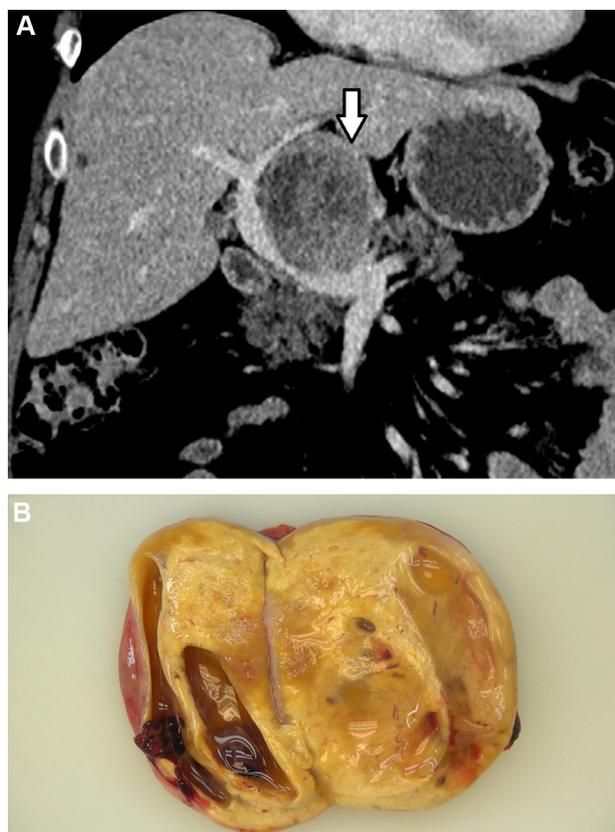
Microscopic examination demonstrated an encapsulated proliferation of spindle-shaped tumor cells, surrounded by a patchy lymphocytic infiltrate, and a normal nerve (Wright). These cells have (Fig. 2A) abundant, weavy faintly eosinophilic cytoplasm without discernible margins, and monomorphic spindle nuclei without atypia or mitosis. The blood vessels are thick-walled and hyalinized (Top). The tumor cells were strongly positive for S-100 protein on immunohistochemistry (Fig. 2B) and negative for melanocytic (HMB45), muscular (desmin, and caldesmon), and gastrointestinal stromal tumor markers (c-Kit, DOG1).

\* Corresponding author.

E-mail address: [najat.mourra@aphp.fr](mailto:najat.mourra@aphp.fr) (N. Mourra).

<https://doi.org/10.1016/j.clinre.2019.11.004>

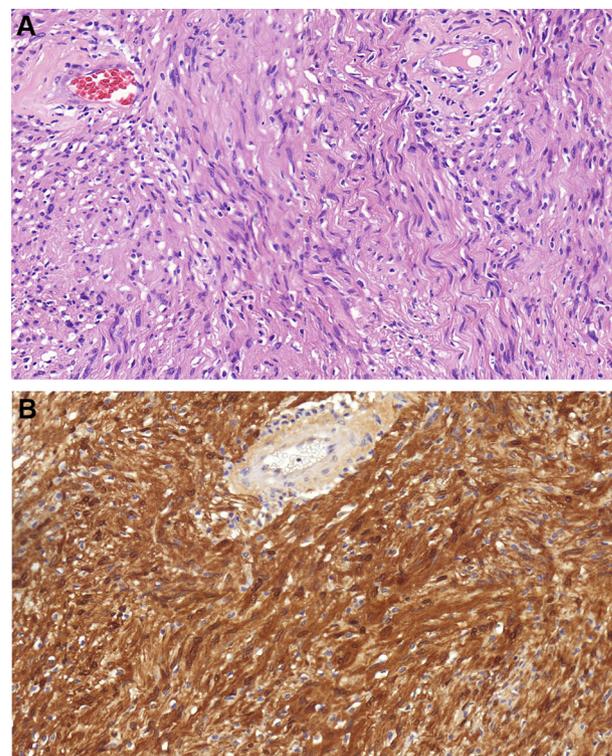
2210-7401/© 2019 Elsevier Masson SAS. All rights reserved.



**Figure 1** A. Enhanced CT scan in a coronal oblique reconstruction showed a well-defined tumor in the porta hepatis, with solid and cystic components. This mass was clearly extra hepatic, with an extrinsic compression and displacement of the portal vein. B. Gross section of this globoid mass which appears grey at the periphery, with wide bright yellow zones and cystic degenerations in the center.

Schwannomas are benign tumors arising from the sheaths of peripheral nerves; they rarely appear in the abdominal cavity. Schwannomas of the porta hepatis is very rare with only a few cases reported in the literature. In a review of 15 patients [1], the mean age at time of diagnosis was 45 years (range 29–74) and the male-female ratio was 6:9. Only 40% of patients were symptomatic with abdominal distension or discomfort as the most common symptom, followed by abdominal pain and jaundice [1,2]. As this case illustrates, the origin and the diagnosis of the tumor was not in the preoperative differential diagnosis of: primary hepatic tumor, Klatskin's tumor and, giant lymph node hyperplasia [1,2]. In an anecdotal report, the diagnosis was made on fine needle biopsy; this diagnostic procedure is not usually resorted to because of concern on finding a safe access to the tumor, and performing a biopsy on a cystic lesion is not recommended [1,3].

The ultrasound imaging often shows isoechoic or hypoechoic solid masses with well-defined limits. Generally, a CT scan shows a well-defined hypodense heterogenous mass with peripheral enhancement and the MRI masses with hypointensity on T1-weighted images and heterogenous



**Figure 2** A. High power view of tumor cells having abundant, weavy faintly eosinophilic cytoplasm without discernible margins, and monomorphic spindle nuclei without atypia or mitosis. B. Tumor cells strongly positive for S-100 protein on immunohistochemistry.

hyperintensity [2]. Surgery is the treatment of choice and is usually curative [3].

In conclusion, the preoperative diagnosis of porta hepatis schwannoma is very difficult due to its rarity, the complex anatomy of this region, and the non specific imaging or clinical manifestations. Therefore, schwannoma should be included in the differential diagnosis of porta hepatis cystic tumors. If feasible, fine-needle biopsy can be helpful in narrowing down the preoperative differential diagnosis and outlining the surgical treatment.

## Contribution

Najat Mourra: concept and design of this paper.

Nikias Colignon: acquisition of data.

Najat Mourra, and Ghazi Zaatari: writing the manuscript.

Final approval of the manuscript: all authors.

## Funding

None.

## Disclosure of interest

The authors declare that they have no competing interest.

## References

- [1] Yoshida A, Yamao K, Takenaka M, Nakai A, Omoto S, Kamata K, et al. Neurilemmoma mimicking a multilocular cystic lesion of the liver. *Intern Med* 2018;57:3377–80.
- [2] Yin SY, Zhai ZL, Ren KW, Yang YC, Wan DL, Liu XY, et al. Porta hepatic schwannoma: case report and a 30-year review of the literature yielding 15 cases. *World J Surg Oncol* 2016;14:103–9.
- [3] Lopes CV, Teixeira UF, Waechter FL, Sampaio JA, Hartmann AA. Porta hepatis schwannoma diagnosed by endoscopic ultrasound-guided fine-needle biopsy. *Endoscopy* 2015;47:E257–8.