Case Report

Primary Glomus Tumor of the Liver

Sakineh Amoueian MD[1], Naser Tayyebi Meibodi MD•[1], Hasan Tavoosi MD[2], Vahid-Reza Ekramifard MD[2], Armin Attaranzadeh MD[3], Mehdi Montazer MD[1]

Abstract

Primary glomus tumors are extremely rare in the liver. To this date, only two cases of primary glomus tumors of the liver have been reported in the literature. This report is the third case of this specified tumor. The patient was a 50 year-old Iranian woman who presented with a history of epigastric pain and nausea. Abdominal computed tomography (CT) scan showed a huge hepatic mass that replaced the entire left lobe. Surgical biopsy showed that the specimen consisted of a vascular network confined by the proliferation of epithelioid cells with round-to-oval nuclei and eosinophilic cytoplasms. The cells expressed diffuse reactivity for vimentin, smooth muscle actin and CD34, whereas no reactivity was observed with carcinoembryonic antigen, synaptophysin, chromogranin, cytokeratin, and human melanoma black 45. We conclude that glomus tumors must be considered in the differential diagnosis of hepatic masses, with immunohistochemical staining for smooth muscle actin being a useful method to prove the diagnosis.

Keywords: glomus tumor, immunohistochemistry, liver

Introduction

Glomus tumors are uncommon benign vascular neoplasms arising from Sucquet-Hoyer arteriovenous anastomoses that act as thermo-regulatory organs in the skin. Classically, they are found under the fingernails, yet their existence has also been reported in the respiratory tract, mediastinum and lungs, gastrointestinal tract, and genitourinary system.[1] Almost all glomus tumors of the digestive system occur in the stomach and those originating from the hepatobiliary tract are extremely rare.[2] To the best of our knowledge, only two primary glomus tumors of the liver have been previously described.[3–4] Here, we report the third case.

Case Report

The patient was a 50-year-old Baloch woman living in Zabol, a small town in Iran, on the border with both Afghanistan and Pakistan. She presented in August 2009 with a three month history of vague epigastric pain and nausea. The only noteworthy finding on physical examination was an enlarged left liver lobe. Laboratory tests showed a mild increase in alkaline phosphatase (293 U/L) and bilirubin (total=1.8 mg/dL, conjugated=0.29 mg/dL). All other tests (renal and liver function tests, hematolologic and coagulation tests, carcinoembryonic antigen and alfa-fetoprotein) were within normal limits. Medical and family histories were unremarkable. Both abdominal ultrasonography and computed tomography (CT) scan demonstrated multiple fused cystic liver cysts measuring 12 cm in greatest diameter that replaced almost the entire left lobe of the liver (Figure 1). Diagnostic investigations did not reveal any other suspicious lesion in other areas of the body. The patient underwent diagnostic surgical biopsy (Figure 2).

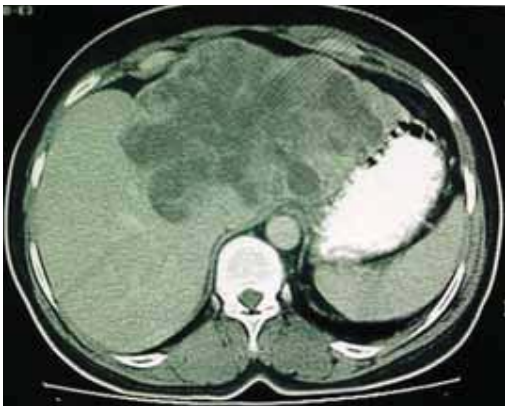

Figure 1. Abdominal CT scan showing a cystic mass that replaced the entire left lobe of the liver.

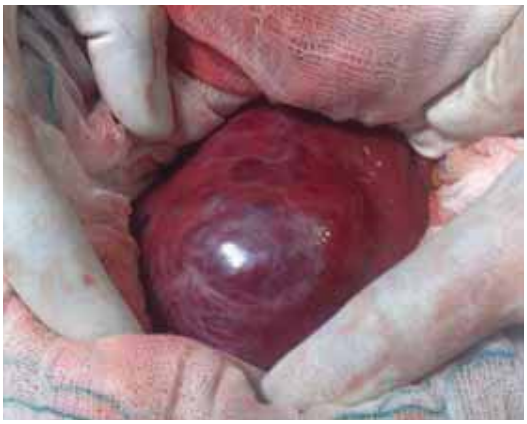

Figure 2. Multiple subcapsular cystic lesions of the liver.

Microscopically, the specimen consisted of a vascular network with normal endothelium, surrounded by the proliferation of monomorphic epithelioid cells with round-to-oval nuclei and eosinophilic cytoplasms (Figure 3). There was no atypia or mitotic activity. An immunohistochemistry (IHC) panel was performed in order to reach a primary diagnosis. The specimen expressed strong

Authors' affiliations: [1]Department of Pathology, Imam Reza Hospital, Mashhad University of Medical Sciences, Mashhad, Iran, [2]Department of Surgery, Imam Reza Hospital, Mashhad University of Medical Sciences, Mashhad, Iran, [3]Department of Pathology, Shiraz University of Medical Sciences, Shiraz, Iran.
•Corresponding author and reprints: Naser Tayyebi Meibodi MD, Department of Pathology, Imam Reza Hospital, Mashhad, Iran.
Tel: +98-511-802-2234, Fax: +98-511-859-1057, E-mail: amouians@mums.ac.ir
Accepted for publication: 9 November 2010

diffuse reactivity for vimentin, smooth muscle actin (SMA) and CD34, whereas reactivities to carcinoembryonic antigen (CEA), synaptophysin, chromogranin, cytokeratin (CK), and human melanoma black 45 (HMB45) were completely negative (Figure 4). Regarding the IHC study results, the lesion was diagnosed as a glomus tumor.

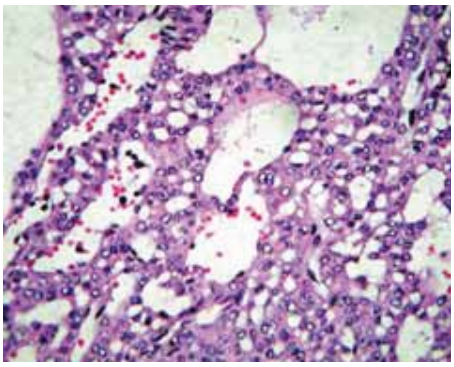

Figure 3. Microscopic investigation, demonstrating a vascular network surrounded with a proliferation of epitheliod cells (H&E, right, 400x).

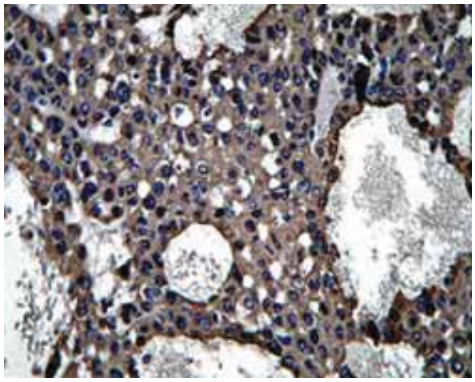

Figure 4. Immunohistochemical study for smooth muscle actin, showing strong reactivity. Similarly, the specimen was reactive for vimentin and CD34 (400x).

The patient was a candidate for therapeutic embolization prior to a surgical resection, but no suitable vessel was found on angiography. Thus, surgical lobectomy was recommended as the only therapeutic alternative. However, the patient refused to undergo a resection surgery due to its potential hazards. After six months of follow-up, the patient's symptoms neither alleviated nor progressed. Thereafter, she underwent surgical removal in another medical center in February 2010 and the mass was excised. The final diagnosis was also a glomus tumor. Afterwards, the patient reported no further discomfort.

Discussion

We reported a primary glomus tumor of the liver. To date, only two other such tumors have been reported: the first by Gassel et al. and the second by Jaiswal et al.[3,4] Glomus tumors usually occur in young adults and generally there is no sex predominance, except in digital lesions which affect females more prevalently.[1] To date, all the reported primary liver glomus tumors have been found in ages over fifty with a male-to-female ratio of 2:1. Nevertheless, it is too early to generalize sex and age distributions in the epidemiology of liver glomus tumors.

Both multicentric and symplastic, malignant glomus tumors have been described, none of which were reasonable in the current case. Multicentric tumors generally occur in children showing an autosomal dominant inheritance; and those with a symplastic, malignant nature usually present with a sarcomatous component, cell atypia or high mitotic activity.[1] Malignant glomus tumors elsewhere in the body may metastasize to the liver; nonetheless, no proof for another primary tumor was found in our case.[5,6] Additionally, benign glomus tumors may seldom undergo malignant transformation and behave aggressively; thus, complete removal is essential in all cases.[7,8]

Most vascular tumors of the liver include hemangioma, epithelioid hemangioendothelioma and angiosarcoma.[9] Histologically, it is somehow easy to distinguish a hemangioma or angiosarcoma from a glomus tumor, but differentiating a hemangioendothelioma is more difficult due to morphological similarities. Hemangioendothelomas express CD34 and vimentin, yet they lack the reactivity for SMA, which strongly supports the diagnosis of a glomus tumor.[10]

Two other excluded differential diagnoses were paragangliomas and perivascular epithelioid cell tumors (PEComas). Particularly, non-reactivity for neuroendocrine immunostains, such as synaptophysin and chromogranin, ruled out a paraganglioma, whereas lack of HMB45 discarded a PEComa.[10,11]

In summary, primary liver glomus tumor is a new entity in hepatopathology and must be considered in the differential diagnosis of hepatic masses. Immunostaining for SMA is necessary to prove the diagnosis and complete resection is strongly advised.

Acknowledgement

The authors thank Dr Ghaffarzadegan for his cooperation in performing the immunohistochemical stainings.

References

1. Calonje E, Fletcher CDM. Vascular tumors. In: Fletcher CDM, ed. *Diagnostic Histopathology of Disease*. 3rd ed. Philadelphia: Churchill Livingstone; 2007: 70 – 72.
2. Miettinen M, Paal E, Lasota J, Sobin LH. Gastrointestinal glomus tumors: a clinicopathologic, immunohistochemical, and molecular genetic study of 32 cases. *Am J Surg Pathol.* 2002; **26:** 301 – 311.
3. Gassel HJ, Klein I, Timmermann W, Kenn W, Gassel AM, Thiede A. Presentation of an unusual benign liver tumor: primary hepatic glomangioma. *Scand J Gastroenterol.* 2002; **37:** 1237 – 1240.
4. Jaiswal VR, Champine JG, Sharma S, Molberg KH. Primary glomangioma of the liver: a case report and review of the literature. *Arch Pathol Lab Med.* 2004; **128:** 46 – 49.
5. Brewis C, Bottrill ID, Wharton SB, Moffat DA. Metastases from glomus jugulare tumours. *J Laryngol Otol.* 2000; **114:** 17 – 23.
6. Brathwaite CD, Poppiti RJ Jr. Malignant glomus tumor. A case report of widespread metastases in a patient with multiple glomus body hamartomas. *Am J Surg Pathol.* 1996; **20:** 233 – 238.
7. Hegyi L, Cormack GC, Grant JW. Histochemical investigation into the molecular mechanisms of malignant transformation in a benign glomus tumor. *J Clin Pathol.* 1998; **51:** 872 – 873.
8. Kayal JD, Hampton RW, Sheehan DJ, Washington CV. Malignant glomus tumor: a case report and review of the literature. *Dermatol Surg.* 2001; **27:** 837 – 840.
9. Bioulac-Sage P, Laumonier H, Laurent C, Blanc JF, Balabaud C. Benign and malignant vascular tumors of the liver in adults. *Semin Liver Dis.* 2008; **28:** 302 – 314.
10. Hammar SP. Immunohistology of lung and pleural neoplasms. In: Dabbs D, ed. *Diagnostic Immunohistochemistry.* 2nd ed. Philadelphia: Chirchil Livingstone; 2006: 343, 351.
11. Hunt JL, Barnes L. Immunohistology of head and neck neoplasms. In: Dabbs D, ed. *Diagnostic Immunohistochemistry.* 2nd ed. Philadelphia: Chirchil Livingstone; 2006: 251.