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Case Report

Rare malignant glomus tumor of the stomach with liver metastases

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ABSTRACT

We report a case of a 72-year-old male admitted in our Unit with anemia and a 10 cm liver neoplasm. Computed tomography scan showed 2 lesions respectively in the II, III and, VIII segment of the liver. Surgical resection of the larger liver mass was performed and the tumor appeared as a solid-cystic mass and a diagnosis of malignant mesenchymal tumor not otherwise specified, was made. One month later a Computed tomography scan detected a dishomogeneous gastric mass, 6 cm in diameter, in the greater curvature, confirmed by esophagogastroduodenoscopy. The pathological diagnosis from endoscopic biopsy revealed a mesenchymal tumor requiring surgical removal for accurate diagnosis. The patient underwent relaparotomy and gastric resection and the pathological findings gave a diagnosis of a rare malignant glomic tumor of the stomach confirmed by the revision of previously performed hepatic resection classified as secondary lesion.

Introduction

Glomus tumors (GTs) are rare mesenchymal neoplasms made of cells resembling those of normal glomus body, classified as perivascular (pericytic) tumor. GT represents less than 2% of all soft tissue tumors and the majority of them are benign occurring in the distal extremities, particularly the hands and feet. The first report of primary gastric GT was made in 1948 by De Busscher [1] and in 1951 by Kay et al [2]. Gastric GTs are extremely rare and usually benign with only few cases reported being malignant. We report a case of a malignant glomus GT with liver metastases.

Case report

Institutional Review Board approval was obtained and informed consent from the patient was acquired. A 72-year-old male was admitted due to liver neoplasms with cystic-solid appearance, observed on ultrasound exam, measuring 10 × 8

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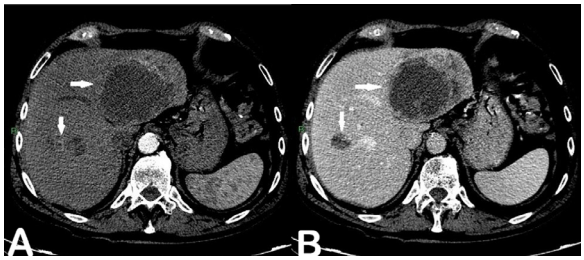

Fig. 1 – CECT (A: arterial phase and B: portal phase) images show 2 focal lesions (arrows) located in the II, III and VIII liver segments characterized by peripheral contrast enhancement and hypodense central area of colliquation.

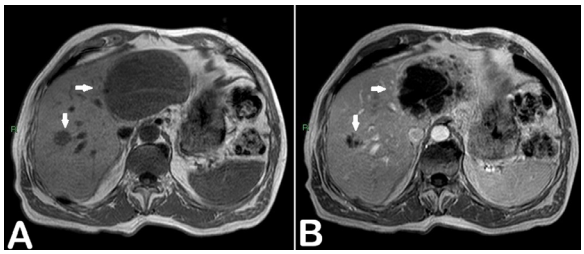

Fig. 2 – Pretreatment (A) axial T1-W TSE and (B) axial T1-W TSE post-gadolinium injection. MR images depict 2 lesions (arrows) located in the II, III and VIII segments of the liver. After administration of the contrast medium both lesions show disomogeneous enhancement inside and on their edge.

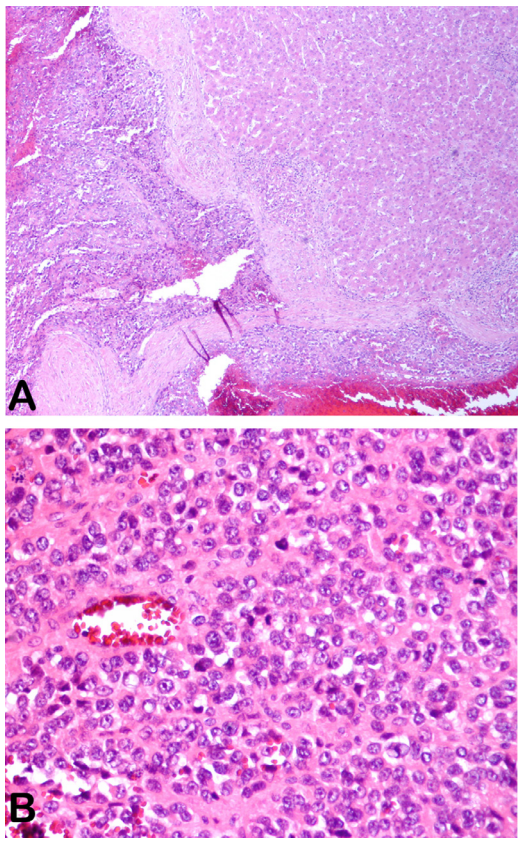

Fig. 3 – Microscopic aspect of liver (A, 40×) mass showing at high magnification (B, 200×) an anaplastic tumor characterized by epithelioid atypical cells with large round to oval nuclei and evident nucleoli. Mitotic activity and is readily observed. Perivascular organization of neoplastic cells is only haphazardly appreciated.

cm and 2 × 2 cm and located in the II, III and, VIII segment, respectively. He was asymptomatic, with moderate renal failure, GFR level 32.26 mL/min and anemia, hemoglobin levels 9.1 g/dL. Blood analysis was negative for infection including the echinococcus test, and common oncological markers were negative.

Contrast Enhanced Computed tomography (CECT) scan (helical scan; 0.6 seconds rotation time; pitch 0.9; 120 kV; 250 mA; image thickness of 2.50 mm) confirmed the 2 lesions that appeared hypodense at baseline precontrast with the larger presenting a partially calcified intralesional septa. After the intravenous administration of contrast medium (Iopromide 300 mgI/mL; 120 mL; 2.5mL/s) the lesions showed mild peripheral contrast enhancement (Fig. 1). As diagnostic interpretation was unclear, a magnetic resonance was performed with axial and coronal images using T1-weighted (W) turbo spin-echo, T2-W turbo spin-echo sequences integrated with fat suppression, DUAL sequences, and diffusion weighted imaging. The 2 lesions appeared hypointense on T1-W sequence with a slight hyperintensity on T2-W sequence and characterized by peripheral enhancement after administration of contrast agent (Gadoteric Acid 0.5 mmol/mL; 15 mL; 2.0 mL/s). In particular, the larger lesion also showed enhancement of the intratumoral septa (Fig. 2). It was decided to excise the larger liver mass. Tumor appeared as a solid-cystic mass 14 × 7 × 4 cm with yellow cystic walls and large hemorrhagic areas, and thickening at the resection edge. The patient was discharged after 5 days in good general condition.

Pathological findings revealed a nodular mass characterized by epithelioid and spindle pleomorphic cells with necrotic and hemorrhagic areas. The immunohistochemical study demonstrated only diffuse vimentin positive cells. A focal and a faint positive stain were observed for alpha-smooth muscle actin and synaptophysin. Proliferative index evaluated by ki-67 was 25%. Morphological and immunophenotype were nonspecific, thus a diagnosis of malignant mesenchymal tumor not otherwise specified, was made (Fig. 3).

After 30 days, CECT scan detected a dishomogeneous gastric mass 6 cm in diameter (Fig. 4), which was confirmed by esophagogastroduodenoscopy. The mass was located in the greater curvature, not seen in the previous radiological studies. Endoscopic biopsies were performed with the patient experiencing sudden onset hematemesis, requiring blood transfusions. The pathological diagnosis was a mesenchymal tumor requiring surgical removal for accurate diagnosis. The patient underwent relaparotomy and gastric resection of the mass. In addition, intraoperative thermo-ablation Radio Frequency Ablation (RFA) of the mass was performed in the VIII segment. No postprocedural complications were observed.

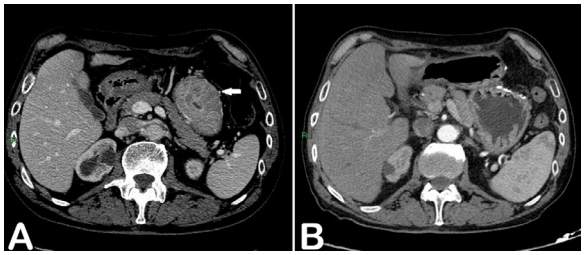

Fig. 4 – CT images before (A) and after (B) gastric resection. (A) dishomogeneous contrast-enhancement gastric area (arrow) located in the greater curvature. (B) results of gastric lesion resection.

Gross findings of the partial gastroresection showed the presence of an ulcerated round mucous membrane of 1.4×1 cm in diameter with hemorrhagic bottom overlying a nodular and hemorrhagic mass of 6×4.5 cm in diameter. Pathological findings of the gastric specimen showed a multinodular tumor characterized by ovoidal cells with round nuclei and slightly eosinophilic cytoplasm surrounding capillaries in a nested arrangement with sparse areas of perivascular hyalinosis. More pleomorphic areas with necrosis, nuclear atypia, frequently mitotic activities (14/10 high power fields (HPF)) and focal spindle cell changes were observed. The immunohistochemical profile showed: α-smooth muscle actin +, h-caldesmon +, desmin −, c-kit −, DOG-1 −, S-100 −, CD34, positivity in the vessels, CK-pan −, synaptophysin +. According to morphological and immunohistological findings, a diagnosis of Malignant Glomic tumor was made according to the 2013 WHO classification for both the gastric and previously performed hepatic resection (Fig. 5).

One month following the gastric resection, the patient underwent an abdominal CECT. At baseline precontrast acquisition, a mildly hyperdense lesion without contrast enhancement on the VIII liver segment, related to the outcomes of the RFA treatment, was seen (Fig. 6). No further lesions were found in the liver or the other abdominal organs. MRI

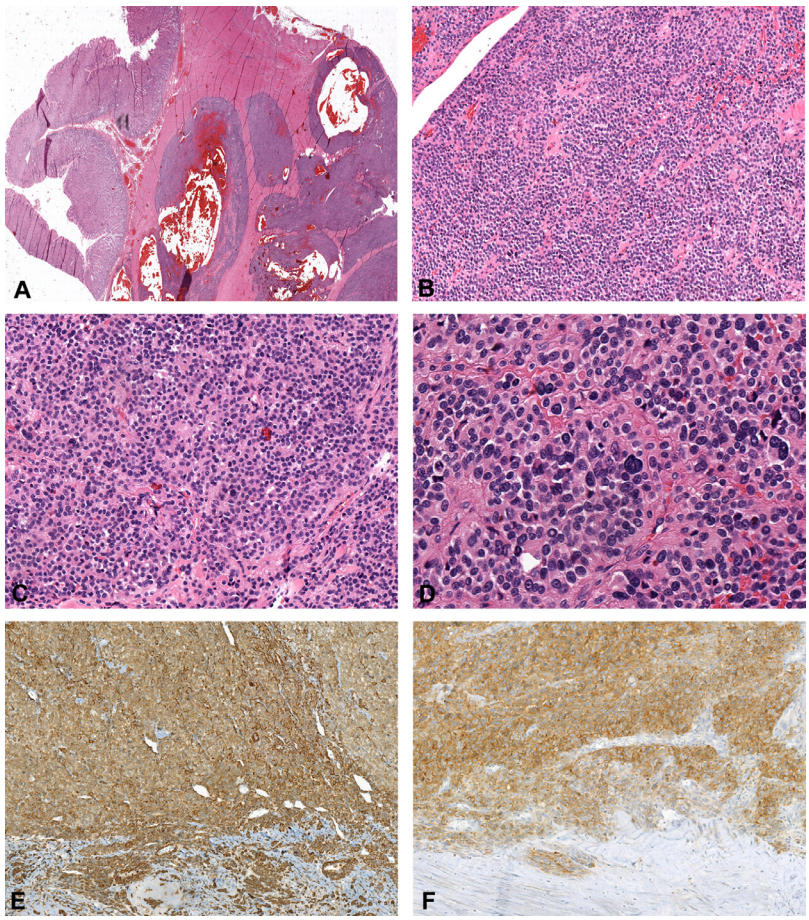

Fig. 5 – Representative histological pictures showing multinodular gastric tumor located between muscle fibers and characterized by prominent ectasic large vessels alternated with fibrous tissue (A). At higher magnification (B) and (C), monomorphic round cells with clear cytoplasm and prominent perivascular distribution have been seen. In other areas, (D) more pleomorphic cells with large nuclei, prominent nucleoli, and occasional multinucleated cells have been appreciated. Immunohistochemical study showing neoplastic cells diffuse positive for alpha-smooth muscle actin (E) and synaptophysin (F). (A) and (B) hematoxyil and eosin stain; original magnification: (A) 10×; (B), (E), and (F) 100×; (C) 200×; (D) 400×.

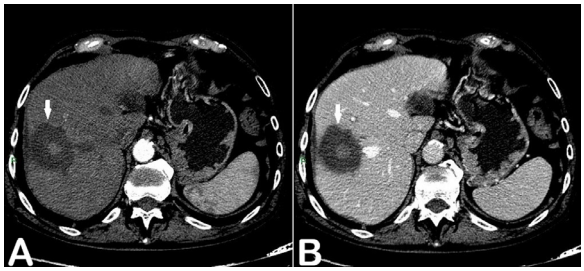

Fig.6 – CT after left hepatectomy and RFA of the lesion on the VIII liver segment. CT images (A) arterial phase and (B) portal phase, show a wide area of ablation (arrows) with a central nucleus of higher density surrounded by hypodensity. CECT do not depict evident contrast enhancement of the whole area. The CT aspect is due to different phase of tissue necrosis.

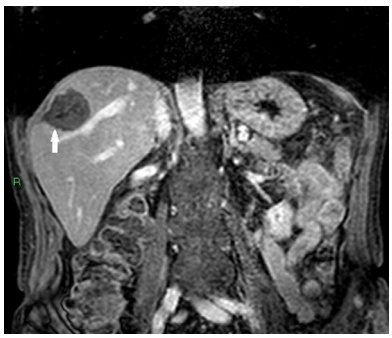

Fig. 7 – Coronal THRIVE postgadolinium MR image obtained after 3 months from RFA confirms the complete ablation of the lesion on the VIII liver segment (arrow).

evaluation, with the same sequences as the pretreatment, was used for the last control post 3 months, confirming the optimal outcomes of RFA, showing a focal area characterized by hypointensity in all postcontrast images (Fig. 7).

Discussion

GTs are extremely rare tumors and their gastric localization is limited to several cases described in literature [3]. Moreover, gastric localization of GT is usually benign, with histologically malignant GTs and clinically malignant GTs being extremely rare, with only few cases being found in the available literature [4–9].

Gastric GTs usually present as asymptomatic. Clinical manifestation may include specific symptoms, including epigastric pain, nausea or vomiting, ulcerous syndrome, and rarely upper gastrointestinal bleeding may be the leading clinical manifestation [10–12]. Radiological findings show enhancement on the arterial phase, as GTs are hypervascular tumors, with sharp demarcation corroborating our case findings. The radiological exams proved unuseful in the preoperative differential diagnosis [13]. In a retrospective study of 10 patients with gastric GT, HU et al [14] found that most GTs are mistaken for GIST or carcinoid tumors before surgery. Thus, they suggest considering GTs as a differential diagnosis when there is a gastric subepithelial mass, located in the gastric antrum, with strong arterial contrast enhancement. MRI findings, Liu KL et al [15] reported the same characteristics we had found in our patient, although MRI was unhelpful in differential diagnosis. GTs lack endoscopic characteristics, the usual appearance is a solitary mass, with an intramural localization located in the gastric antrum.

Miettinen et al [16] reported the analysis of 32 cases of gastrointestinal GT, 31 located in the stomach and 1 in the cecum. All patients were surgically treated, 15 patients (47%) had surgical wedge excision, sub-total gastrectomy was performed in 4 patients (12.5%), and the remaining patients (40%) were treated with antrectomy or hemigastrecomy. In this case series, only 1 (3%) behaved as malignant tumor and developed liver metastases, the patient died 50 months postoperatively. This patient had the largest tumor size, which measured 6.5 cm, while the average tumor size being 2 cm, however, histological features did not predict malignancy.

A review of Chinese literature reported 57 cases by Fang et al [17]. Most common localization found in the majority of patients was the gastric antrum (93%): other sites included gastric corpus, 3 patients, and the corpus-antrum junction in only 1 patient. All these patients were treated surgically, a wedge resection was performed in 36 patients (63%), subtotal gastrectomy in 18 patients (31.5%), and only 3 (5.2%) were treated on demand, depending on tumor localization. In a Korean clinicpathologic analysis of ten cases, Kang et al [18] reported a predominantly higher incidence in male patients, 8 out of 10, than in women, 2 out of 10. Gastric antrum was the most common localization, 7 out of 10. All patients (100%) underwent wedge surgical resection.

GTs are rare with preoperative highly challenging. GTs lack typical clinical, endoscopic, and radiological characteristics, with correct diagnosis attained solely by histological examination.

According to the 2013 WHO classification, the diagnosis of malignant GT includes: 1) marked nuclear atypia and any level of mitotic activity; 2) atypical mitotic figures. These features were all present in this case study. Presently, dimension (>2 cm) and deeper location are classified as “uncertain malignant potential” in the absence of other malignant characteristics despite the WHO classification.

A negative resection margin is considered the treatment of choice for this type of tumor. Regarding malignant cases, like this one, accurate treatment choice is limited as case reports in literature are lacking.

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