Primary gastric rhabdomyosarcoma. Case report

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Summary
Rhabdomyosarcomas are rare and malignant tumors. There have been reported two histological types of gastric rhabdomyosarcomas, the pleomorphic and embryonal cell types. We report the case of a 53-year-old male with endoscopic diagnosis of a Bormann type III ulcer which revealed a gastric primary rhabdomyosarcoma. Ultrasound showed two liver lesions, two hepatic pedicle lymph nodes and a huge primary gastric tumor. CT scan revealed a primary gastric tumor. The patient was submitted to a distal gastrectomy with a Billroth II reconstruction and a resection of the distal liver metastases at segment IV. The patient was discharged uneventfully on the eighth postoperative day. The gold standard for a final diagnoses is the immuno-histochemical staining of the endoscopic biopsy. There is very little information on the results of chemotherapy and the surgical treatment is the best choice.

Key words. Gastric tumors, rhabdomyosarcoma, oncology, surgery, treatment, diagnosis.

Rhabdomyosarcoma primario gástrico. Presentación de un caso

Resumen
Los rabdomiosarcomas son tumores malignos raros. Hay reportados dos tipos histológicos de rabdomiosarcoma gástrico: pleomórficos y de células embrionales. Presentamos el caso de un paciente de sexo masculino de 53 años de edad al cual se le realizó una endoscopia encontrándose una úlcera tipo III de Bormann que resultó ser un rabdomiosarcoma gástrico. Los estudios de ultrasonido informaron dos imágenes compatibles con metástasis hepáticas. El enfermo fue sometido a una gastrectomía distal con reconstrucción a lo Billroth II y a la resección de las metástasis hepáticas. La evolución postoperatoria fue satisfactoria y se otorgó el egreso hospitalario al octavo día. El patrón de oro para el diagnóstico final es la inmunohistoquímica y la biopsia endoscópica. El tratamiento final es la cirugía. Rabdomiosarcomas son rares y malignos tumores. En el tracto gastrointestinal se presenta su localización en el estómago con mucha menor frecuencia. La primera publicación en la literatura referida a rhabdomyosarcoma gástrico fue reportada por Fox et al en 1990.1 Presentamos el caso de un 53-year-old man with a primary gastric rhabdomyosarcoma, together with its clinical, surgical, macroscopic and histopathological features.

Palabras claves. Tumores gástricos, rhabdomyosarcoma, oncología, cirugía, tratamiento, diagnóstico.

Case report

We report the case of a 53 year-old Caucasian male admitted to the Hospital Nacional Professor Alejandro Posadas, due to an upper digestive bleeding with mucocutaneous pallor, dyspnea and weight loss. An upper gastric video endoscopy was performed, identifying a Bormann Type III ulcer in the gastric roof and in the greater gastric curvature. A biopsy was taken and the pathology report indicated: gastric rhabdomyosarcoma, AM2 + 30%, MIF4 + 20%, Actin SP + 70%, EMA + 70%, VIM + 90%, Desmin + 90%.

The patient was referred to the Department of Surgical Gastroenterology and was scheduled for surgery, after completing the preoperative work-up. A double contrast gastroduodenal radiography revealed a large tumor in the antrum (Figure 1). An ultrasonography was performed and revealed two
hypoechoic solid lesions in the liver, compatible with metastases, and two lymph nodes in the hepatic pedicle. In the epigastrium a large heterogeneous, hypoechoic lesion was identified. A CT scan was performed showing a primary heterogeneous lesion on the gastric wall (Figure 2). A laparotomy was performed. A large tumor in the greater curvature of the abdomen and liver metastases in the left liver lobe were identified (Figures 3 and 4). The patient underwent a distal gastrectomy with a Billroth II reconstruction, a cholecystectomy and a resection of the distal liver metastases in segment IV.

The patient was taken to the intensive care unit (ICU) for immediate postoperative recovery. Enteral nutrition was initiated on the third post-operative day. After the nasogastric tube was disconnected on the fourth post-operative day, an oral diet was started. The patient was discharged uneventfully on the eighth postoperative day.

The pathology report indicated: gastric pleomorphic spindle cell rhabdomyosarcoma, with liver metastases. Tumor size 15 x 12 x 8 cm (Figure 5). Involvement of perivisceral adipose tissue. Capsular metastases and two positive lymph nodes of the greater gastric curvature. Proximal and distal resection margins not involved. Residual gastric mucosa with lymphocytic chronic gastritis associated to *Helicobacter pylori bacillus*. Gallbladder: chronic cholecystitis, cholesterosis. Immunohistochemistry report: Vimentin + 90%, Actine SP + 80%, Desmin + 70%, CD 34 -, S-100 -, C-Kit -, MyoD1 + 30%, Myf4 + 30% (Figures 6 and 7).
At present the patient is undergoing chemotherapy at the Oncology Department of the Hospital Prof A Posadas, with normal tumor markers and clinical parameters.

Discussion

Rhabdomyosarcomas of the stomach are a rare neoplasm, different from other sarcomas of the stomach with less than 15 cases reported or referenced in the world literature.

Gastric rhabdomyosarcomas are aggressive tumors (median survival: 2.5 months) in children and adults. They generally present with signs and symptoms due to metastatic disease to the lung or cervical lymph nodes and are often difficult to be diagnosed. Inaccurate diagnosis and treatment options are the cause of radiation therapy given to the metastases and not to the primary tumor. In contrast, gastric sarcomas in general have a longer median survival, tend to present with symptoms in the gastrointestinal tract and typically are accurately diagnosed during the life of the patient.1,2

Gastric rhabdomyosarcomas tend to metastasize to the lymph nodes and lung. In the literature, there is no indication that gastric rhabdomyosarcomas metastasize to other organs from some other site. Indeed, metastases to the stomach arising from a rhabdomyosarcoma are rare.1,3 In our patient we identified non-adenocarcinoma liver metastases, which are much more uncommon.

Fox et al reported two histological types of gastric rhabdomyosarcomas, the pleomorphic and embryonal cell types. Embryonal rhabdomyosarcomas are formed by small poorly differentiated cells intermingled with occasional round or elongated rhabdomyoblasts. These tumors exhibit highly cellular areas together with loose, less cellular myxoid areas. The degree of differentiation of embryonal rhabdomyosarcomas may vary from area to area and from patient to patient, but generally they tend to mimic embryonal and fetal skeletal muscle development. Pleomorphic rhabdomyosarcomas have features similar to embryonal rhabdomyosarcomas but also have an additional prominent cellular component composed of small to large, irregularly shaped, loosely arranged, scattered round to pleomorphic cells, some of which have a deeply eosinophilic cytoplasm.1

*Helicobacter pylori* infection decreases intralumi-
Gastric rhabdomyosarcoma

Mariano Palermo y col

Gastric rhabdomyosarcoma, which in turn decreases the antioxidant mechanisms. Also in the long-term, due to the atrophy and metaplasia, beta-carotene levels decrease and gastric cancer risk increases. Our patient was also *Helicobacter pylori* positive, thus increasing the risk factors for gastric primary cancer.

Most of the tumors such as lymphomas, carcinoid tumors, carcinomasarcomas, leiomyosarcomas, and rhabdomyosarcomas were reported to be associated with adenocarcinoma of the stomach. The sarcoma component in the case published by Kazuki in 2003, occupied less than 10% of the entire tumoral area and consisted of spindle cells some of which presented rhabdomyosarcoma differentiation. Our case showed a predominant sarcoma (rhabdomyosarcoma) component, being a rare presentation in gastric primary tumors. Histologically, the most common carcinoma component in the gastric carcinoma is the tubular adenocarcinoma, followed by papillary adenocarcinoma.

There is very little information on the results of chemotherapy or other drugs in gastric rhabdomyosarcomas. Therefore, it is our opinion that the surgical treatment of these tumors is the best option.

In conclusion, these primary gastric tumors are rare tumors with high malignant potential, in general of uncertain etiology. At present, no clinical tests are available for early diagnosis (ultrasound, CT, gastrography with barium). The gold standard for the definitive diagnosis is the immuno-histochemical staining of the endoscopic biopsy.

References