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Malignant rhabdoid tumour in the stomach

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ABSTRACT

In this case report, a rare tumoral morphology in the gastrointestinal system is introduced. Malignant rhabdoid features are rarely observed in the gastrointestinal system but this discrimination is important due to the tumor's poor prognosis, aggressive behavior and early metastasis.

Key Words: Gastric neoplasms, adenocarcinoma, rhabdoid tumor

INTRODUCTION

Malignant rhabdoid tumor is one of the most aggressive and lethal tumors in pediatric oncology (1). It is observed most often in the head, neck, urogenital tracts and skeletal system. However, they have also been described in the skin, brain, mediastinum, liver and other peritoneal structures especially in the adult age group, albeit very rarely. Rhabdoid tumor accounts for approximately 0.1-0.2% of gastric cancers (2). This article presents a patient with gastric carcinoma showing rhabdoid characteristics that may be confused with diffuse type gastric carcinoma.

CASE PRESENTATION

The patient was informed that she has a rare disease; her written consent and her husband's oral consent were received for publication purposes given that she was hospitalized at a teaching and research hospital.

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The seventy-year-old female patient was referred to our clinic with a diagnosis of gastric carcinoma, following evaluation of her gastric pain that had been present for three months. Her medical history included diabetes mellitus for 10 years and previous cholecystectomy. Her laboratory studies showed: leucocyte: 8500/µL, Hgb: 13.2 g/dL, glucose: 109 mg/dL, with renal and hepatic function tests and electrolyte values in the normal ranges. The CEA and CA-19-9 values were also in the normal ranges.

On endoscopy, the esophagus lumen and mucosa were identified to be normal; a stalked polipoid mass, 2 cm in size, was observed in the cardia and biopsies were obtained from this site. The pylorus was centralized, bulbus and the second portion of duodenum were normal. The endoscopic preliminary diagnosis was cardia tumor. The biopsy results revealed a signet ring cell carcinoma. It was also stated that lymphoma could not be ruled out in the biopsy specimen.

No masses and infiltrations were observed in the pulmonary parenchyma in the thoracoabdominal CT scan. The liver was normal, the gallbladder was not observed (previous cholecystectomy). The common bile duct measured 15 mm at the level of porta hepatis. A heterogeneous mass originating at the level of gastric cardia, stretching along the lesser curvature and showing a prominent polipoid extension towards the lumen with a lobulated, contoured extension to the gastrohepatic ligament was observed.

The patient underwent total gastrectomy, Roux-Y esophagojejunostomy and D-2 lymph dissection with the diagnosis of gastric carcinoma. During surgery, a 5 cm lesion with heterogeneous and nodular surface, originating 4 mm below the gastric cardia, extending along the lesser curvature was observed, the tumor invaded left lobe medial segment. No metastasis were identified in the liver and other visceral organs. On the 6th post-operative day, the anastomosis was controlled via oral contrast media and she was started on oral diet; the patient had no additional problems and was discharged on post-operative day 9.

The pathology result of the patient was reported as gastric carcinoma showing rhabdoid characteristics (malignant rhabdoid tumor). It was identified that the tumor involved all gastric layers and there

was vascular invasion. In the immunohistochemical study, diffuse, strong staining with vimentin was observed and metastasis was identified in one of the dissected lymphatic nodules; the other lymph nodules were stated to be reactive. The patient was considered as T3N1M0 in post-operative staging. The patient was accepted as gastric carcinoma variant, and weekly fluorouracil-folinic acid (FUFA) regimen was initiated considering her performance status, as well. In the 14th week of treatment, a liver metastasis was identified on abdominal tomography. The patient was prescribed 6 courses of docetaxel-cisplatin-fluorouracil (DCF) regimen due to progression. As progression in the metastatic lesions of the liver was identified 2 months after the end of treatment, the patient was started on a single agent capecitabine therapy. The patient was prescribed three courses and her response evaluation studies are awaited in her last control visit.

DISCUSSION

Malignant rhabdoid tumors are one of the most aggressive tumors of the pediatric age. They are very rarely observed in non-renal locations such as the respiratory and gastrointestinal system. Even though it is a well-defined entity in kidneys and central nervous system in the pediatric age, it is seen rather rarely in soft tissues (3-5). Since it does not have specific symptoms, the disease is diagnosed at advanced stages (6). The disease is rarely observed in the stomach. All reported cases are old, and are characterized by either gastrointestinal bleeding or large, ulcerated masses (7). Our patient did not have any bleeding complaints, however, she had a large mass that had invaded the liver. The most frequently observed positive immunohistochemical marker is vimentin. The other mesenchymal and epithelial markers have also been reported as positive at varying amounts. However, the tumor cells are almost always negative for desmin and S 100. In our patient, strong and diffuse staining only with vimentin was observed.

Gastrointestinal malignant rhabdoid tumors are slightly differentiated carcinomas localized in the esophagus, stomach, small intestine, colon, pancreas and hepatobiliary tract. In most patients, remote metastasis develops shortly after diagnosis. The metastatic invasion pattern is multiple and in the form of transmural tumor implants in the duodenum and small intestine (6).

Rhabdoid morphology is composed of large polygonal cells with eccentric location and vesicular nuclei. These cells have eosinophilic inclusions that displace the nucleus. The identification of cytokeratin and vimentin positivity is crucial for differential diagnosis, as was the case with our patient. In this way, the epithelial or mesenchymal origin of the tumor can be identified.

For differential diagnosis, histologic findings should be kept in mind and GIST, sarcoma, malignant melanoma and lymphoma should be considered (8, 9). Due to the aggressive clinical behavior and poor prognosis of the disease, its differential diagnosis should be made.

Ueyama et al. (10) identified only 4 patients among 5437 gastric carcinoma cases who had this morphology. These tumors have a very poor prognosis in a way similar to primary renal rhabdomyosarcoma, and they have a very aggressive clinical course (11). It is stated that these 4 rhabdoid tumor patients died within 6 months due to the disease. In contrast to our patient, these patients had distant organ and lymph node metastasis identified during laparotomy.

CONCLUSION

Based on a retrospective literature review, it was observed that gastric carcinoma patients with such morphology survive less than one year (12). In several studies, it was especially emphasized that the observation of rhabdoid characteristics in tumor cells was associated with poor prognosis (13). Therefore, we believe that the identification of rhabdoid phenotype in the stomach is of paramount importance. In fact, the prognosis of these patients is very poor, with even worse response to chemotherapy and radiotherapy.

Informed Consent: Verbal informed consent was obtained from patient who participated in this study.

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