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A case report and review of the literature

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Gastric medullary carcinoma. A case report and review of the literature

Medullary gastric carcinoma (MGC) is a rare gastric neoplasm characterized by histological appearance of dense lymphocytic infiltration of the stroma. The prevalence of MGC among all gastric neoplasms is about 1%. Although it is classified within the poorly differentiated carcinomas, the biological activity of MGC is similar to that of the well differentiated carcinomas. Hereby, we present a case of a 64-year-old male patient who underwent upper gastrointestinal (GI) endoscopy for abdominal pain over the last 2 months. The pathological findings of the GI endoscopy specimen revealed adenocarcinoma of the stomach. Subsequently, the patient underwent total gastrectomy and D2 lymph node dissection. Pathological evaluation of the excised specimens samples were compatible with MGC (gastric carcinoma with lymphoid stroma) which was staged as T2N0M0. This paper purposed to describe the clinical and pathological findings of MGC.

KEY WORDS: Gastrectomy, Gastric carcinoma, Medullary

Introduction

Gastric cancer is reported to remain one of the leading causes of cancer-related deaths worldwide, even with some rare subtypes such as medullary gastric carcinoma (MGC) [1,2]. MGC, characterized by histological appearance of dense lymphocytic infiltration of the stroma has a favorable prognosis despite its classification among poorly differentiated gastric neoplasms [2]. MGC, which is also named as lymphoepithelioma-like gastric carcinoma constitutes about 1% of all gastric neoplasms. It is more frequent after the 6th decade of life in males [3].
Medullary gastric carcinomas are morphologically small tumors localized superficially on mid and proximal regions of the stomach [4]. Although one would easily expect a poor prognosis owing to the classification of this carcinoma among poorly differentiated gastric neoplasms and its proximal localization, previous data indicate that MGC exhibits favorable clinicopathological features similar to that of the well differentiated tumors, probably due to the presence of lymphoid stroma [5]. Lymph node and peritoneal metastasis is less frequent in MGC compared to typical scirrhous adenocarcinomas. While mortality occurs due to peritoneal metastasis in scirrhous adenocarcinomas, subjects with MGC often die secondary to hematogenous metastasis which is similar to that of the subjects with well differentiated tumors [6]. Postoperative survival of the patients with MGC is similar to those with well differentiated adenocarcinomas and significantly longer than subjects with scirrhous adenocarcinomas [7].
This paper purposed to describe a case of MGC which is an extremely rare type gastric neoplasm and to review the literature regarding MGC.

Case Report

A 64-year-old female patient with no previous medical history presented with abdominal pain over the last two

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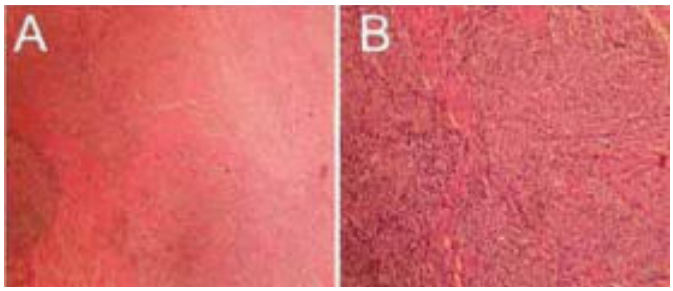

Fig. 1: Microscopic demonstration of dense lymphocytic infiltration of the stroma.

months. Physical examination and blood test including CA 19-9 and CEA were normal. The patient underwent an upper gastrointestinal (GI) endoscopy, which demonstrated an ulcerative lesion on the cardia of the stomach with the surface covered by fibrin. Pathological findings of the endoscopy specimen revealed a diffuse type signet-ring cell adenocarcinoma of the stomach. Abdominal computed tomography showed no abnormal findings. Subsequently, the patient underwent total gastrectomy and D2 lymph node dissection.

Macroscopically, the tumor was 4x3 cm in size and had irregular edges. Pathological evaluation of the excised specimens samples were compatible with MGC (gastric carcinoma with lymphoid stroma) which was staged as T2N0M0 (Fig. 1). The tumor was limited to muscularis propria and there were no signs of subserosal, perineural, and vascular invasion (pT2). The total 44 lymph nodes were reactive (N0).

Discussion

Gastric carcinoma is reported to be the third leading cause of death due to cancer [8]. However, medullary gastric carcinoma, in other words lymphoepithelioma-like gastric carcinoma is a rare gastric adenocarcinoma which is usually diagnosed in male individuals over 60 years of age. It is characterized microscopically by the attachment of neoplastic epithelial cells that show intensive lymphoid proliferation. T and B lymphocytes, multinucleated giant macrophages, and dendritic and plasma cells are often involved in this stromal lymphoid infiltration [9]. MGC is histomorphologically diagnosed by the presence of diffuse lymphoid proliferation within the neoplastic epithelial cells. Lypmphoid metastasis is rare in MGC. Nakamura et al have reported that more than 80% of the lymphoepithelioma-like gastric carcinomas are associated with EBV [10].

Although it is classified within the group of poorly-differentiated gastric neoplasms, the biological behaviour and survival rate of MGC is similar to that of the well-differentiated gastric adenocarcinoma [7-11]. Male gender, invasion depth, stage and the presence of lymphoid stroma has been shown to be related to prognosis [12]. The 5-years survival rate of GMC and non-medullary adenocarcinamoas are reported to be %83 and %46, respectively [11]. Compared to adenocarcinoma, MGC develops in a smaller area and in upper regions of the stomach such as corpus and cardia. Moreover, it is less invasive. In this case, the tumor was located in cardia of the stomach. Histopathological evaluation showed that MGC was limited to muscularis propria and there were no signs of subserosal, perineural, vascular invasion and lymph node invasion.

In conclusion, MGC is rare gastric carcinoma, which is identical to well-differentiated gastric adenocarcinomas in terms of biological behaviour and survival rate. Death usually results from hematogenous metastasis. Early diagnosis and surgical treatment is therefore crucial to improve survival. In this paper, we presented an early stage MGC that was treated by total gastrectomy.

Riassunto

Carcinoma gastrico midollare (MGC) è una rara neoplasia gastrica caratterizzata dalla comparsa istologica di una densa infiltrazione linfocitica dello stroma. La prevalenza di MGC tra tutte le neoplasie gastriche è di circa l'1%. Sebbene sia classificato tra i carcinomi scarsamente differenziati, l'attività biologica di MGC è simile a quella dei carcinomi ben differenziati. Con la presente, presentiamo un caso di un paziente maschio di 64 anni che è stato sottoposto a endoscopia del tratto gastrointestinale superiore (GI) per dolore addominale negli ultimi 2 mesi. I risultati patologici del campione di endoscopia gastrointestinale hanno rivelato adenocarcinoma dello stomaco. Successivamente, il paziente è stato sottoposto a gastrectomia totale e dissezione dei linfonodi D2. La valutazione patologica dei campioni di campioni asportati era compatibile con MGC (carcinoma gastrico con stroma linfoide) che è stato classificato come T2N0M0. Questo documento si proponeva di descrivere i risultati clinici e patologici di MGC.

References

1. Corso G, Seruca R, Roviello F: *Gastric cancer carcinogenesis and tumor progression*. Ann Ital Chir, 2012; 83(3):172-76.

2. Falzarano S, Mourmouras V, Mastrogiulio M, La CM, Vindigni C: *Undifferentiated gastric carcinoma with lymphoid stroma (lymphoepithelioma-like carcinoma/medullary carcinoma).* Pathologica, 2009; 101(1):15-7.

3. Pavithra Herath CH, Chetty R: *Epstein-Barr virus-associated lymphoepithelioma-like gastric carcinoma.* Archives of pathology & laboratory medicine, 2008; 132(4):706-9.

4. Aslan F, Yükrük FA, Buğdaycı Başal F, Durnalı A: *Gastric medullary carcinoma: A rare case report.* Case reports in oncological medicine, 2016; 2016:2875471.

5. Adachi Y, Mori M, Maehara Y, Sugimachi K: *Poorly differentiated medullary carcinoma of the stomach.* Cancer, 1992; 70(6):1462-66.

6. Grogg KL, Lohse CM, Pankratz VS, Halling KC, Smyrk TC: *Lymphocyte-rich gastric cancer: Associations with Epstein-Barr virus, microsatellite instability, histology, and survival.* Modern pathology, 2003; 16(7):641-51.

7. Otsuji E, Kuriu Y, Ichikawa D, Ochiai T, Okamoto K, Hagiwara A, et al: *Clinicopathologic and prognostic characterization of poorly differentiated medullary-type gastric adenocarcinoma.* World journal of surgery, 2004; 28(9):862-65.

8. Garofalo A, Carboni F, Federici O, Giofré M, Balla A, Valle M, et al.: *European single-center experience on 356 operated patients for gastric cancer*. Ann Ital Chir, 2020; 91:137-43.

9. Chetty R: *Gastrointestinal cancers accompanied by a dense lymphoid component: An overview with special reference to gastric and colonic medullary and lymphoepithelioma-like carcinomas.* Journal of clinical pathology, 2012; 65(12):1062-65.

10. Nakamura S, Ueki T, Yao T, Ueyama T, Tsuneyoshi M: *Epstein Barr virus in gastric carcinoma with lymphoid stroma. Special reference to its detection by the polymerase chain reaction and in situ hybridization in 99 tumors, including a morphologic analysis.* Cancer, 1994; 73(9):2239-49.

11. Huang KH, Wang RF, Yang MH, Wu CW, Fang WL, Li AFY, et al.: *Advanced gastric cancer patients with lymphoid stroma have better survival than those without.* Journal of surgical oncology, 2013; 107(5):523-28.

12. Dogan M, Savas B, Utkan G, Bayar S, Ensari A, Icli F: *A rare gastric neoplasm: Gastric medullary carcinoma.* Medical Oncology, 2011; 28(4):945-7.