Case Report

Mucin-Secreting Gastric Adenocarcinoma with Rhabdoid Areas

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ABSTRACT

Rhabdoid tumor, first described in kidneys of infants and children, is an aggressive tumor that has been reported in several extrarenal locations. In this report, we describe the case of a 40-year-old patient with gastric adenocarcinoma composed of histologically well-differentiated glandular areas and focal rhabdoid zones. The rhabdoid component showed typical features such as abundant eosinophilic cytoplasm, eccentric nuclei, prominent nucleoli and intense focal positive immunohistochemical cytoplasmic reaction for vimentin. Recognition of the rhabdoid phenotype in gastrointestinal tract neoplasms is important because this feature is associated with poor prognosis and unresponsiveness to conventional therapy.

Key Words: Gastric adenocarcinoma, rhabdoid, immunohistochemistry, prognosis

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Rhabdoid tumors were first described as a variant of Wilms tumor carrying an unfavorable prognosis, but soon it became clear that this tumor was a distinct entity.[1] Rhabdoid tumors have now been reported in nearly all organs of the body, both renal and extrarenal including esophagus, stomach, small and large intestine,[2] central nervous system,[3] lung and mediastinum, skin and soft tissue. These tumors are more common in childhood. Further, composite tumors having areas typical of adenocarcinoma and focal Rhabdoid differentiation have also been described, the Rhabdoid differentiation being associated with a poorer prognosis.[4] We describe a rare case of gastric adenocarcinoma in a 40-year-old woman with focal rhabdoid differentiation.

CASE REPORT

A 40-year-old woman presented with complaints of lump in the abdomen since two months, which was rapidly increasing in size and was associated with decreased appetite and weight loss. On examination, the patient was found in poor general condition with severe pallor and pedal edema. On palpation, a 10 × 8 cm lump was noted in the epigastrium, which was firm, mobile and nontender. CT scan showed concentric thickening of the stomach wall in the region of the distal body and pylorus, along with multiple hypoechoic shadows in the liver parenchyma [Figure 1]. On upper GI endoscopy, large ulcer was seen in the distal part of the stomach, reaching up to the pylorus. The pylorus could not be negotiated by scope. A biopsy taken simultaneously from the pylorus was however noncontributory.

A provisional clinical diagnosis of carcinoma was made and a distal gastrectomy with gastrojejunostomy performed. A large circumferential growth arising from the pylorus, adherent to the pancreatic capsule with mesocolon was found. Multiple local gastric lymph nodes were also found enlarged. The liver showed multiple deposits.

We received a distal gastrectomy specimen measuring 12 × 7 × 4 cm with attached omentum measuring 14 × 12 × 3 cm. There was a circumferential ulceroproliferative growth in the region of the body and antrum with maximum tumor diameter of 9 cm [Figure 2]. The tumor was close to the distal margin. A tumor perforation was present in the posterior wall of stomach in antral region along the lesser curvature. Multiple lymph nodes were dissected from along the lesser curvature.

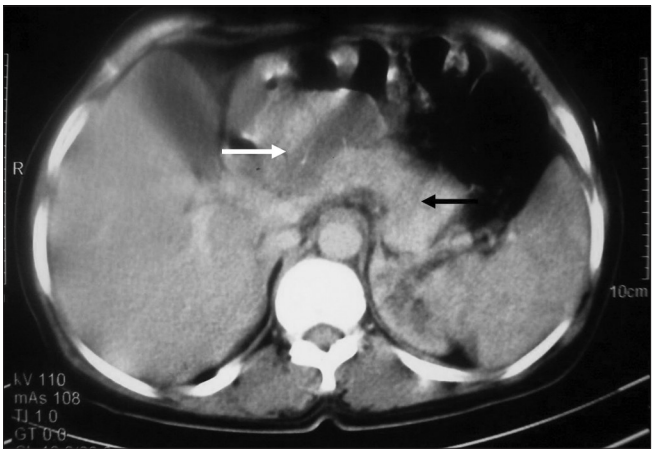

Figure 1: CT scan revealing growth in the stomach wall (white arrow) infiltrating into the pancreas (black arrow)

On microscopy, the tumor was found to be predominantly composed of large lakes of mucin with clusters as well as singly lying atypical cells with high N:C ratio, pleomorphic nuclei and prominent nucleoli. Many signet ring cells were also seen [Figure 3]. There was an adjacent area with tumor cells lying in solid sheets, with dyscohesive round-to-oval cells having moderate amount of eosinophilic cytoplasm, eccentric nuclei and prominent nucleoli [Figure 4]. Occasional cells showed hyaline (PAS positive) inclusions. There was evidence of lymphatic permeation with metastatic deposits in lymph nodes. Immunochemistry revealed cytokeratin and EMA positivity in both well-differentiated mucinous areas as well as solid areas and CEA negative in both. The solid area was also focally positive for vimentin and negative for smooth muscle antigen and ckit. On the basis of morphological and immunochemical findings, a diagnosis of mucinous adenocarcinoma (signet ring type) with foci of rhabdoid differentiation was made.

DISCUSSION

Malignant rhabdoid tumor was originally described in 1978 as a renal malignant tumor of infancy and childhood[1]. Tumors of similar clinicopathological features have been cited in a variety of extrarenal sites and are known to carry a poor prognosis. Also composite extrarenal rhabdoid tumors (CERTs), in which recognizable parent neoplasms are admixed with rhabdoid areas, have been recognized in several topographic sites including CNS,[5] subcutaneous tissue and GIT.[6] Though the pure rhabdoid tumors are more common in infancy and childhood, the composite tumors show a wide range of age distribution. However, this type of morphological combination remains rare and only a few cases of gastric adenocarcinoma with rhabdoid areas have been reported in the literature till date.[11-13] One case of gastrointestinal stromal tumor (GIST) with rhabdoid areas has also been reported.[14]

Most of the gastric adenocarcinomas with rhabdoid areas are poorly differentiated and show widespread metastases to the surrounding organs and liver at the time of diagnosis, similar to the findings in our case. This is probably due to a high rate of lymphatic and vascular permeation at the time of diagnosis. The tumor calls show positive immunoreactivity for both vimentin and cytokeratin in their cytoplasm. Utsunomiya *et al.* studied 239 poorly differentiated adenocarcinomas of the stomach with solid components after reviewing 3578 cases, for the expression of vimentin. Of these, 15 cases (0.4%) were vimentin positive and 12 (0.3%) were associated with varying amount of rhabdoid phenotype and prominent lymphatic permeation.[15] The authors suggested that the dyscohesive nature of the cells maybe responsible for the rapid metastases and vimentin may affect cell-to-cell interaction. They also found prognosis of vimentin positive solid tumors poorer than vimentin-negative poorly differentiated tumors

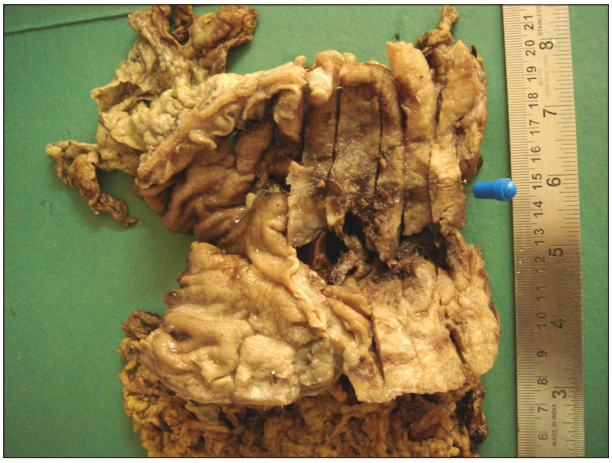

Figure 2: Gross appearance of the stomach showing a large ulcerative growth in the distal region, reaching almost up to the resected margin (marker)

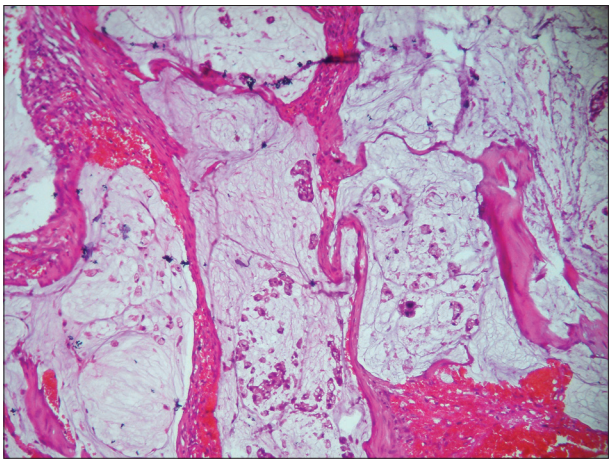

Figure 3: Section from the stomach showing well-differentiated adenocarcinoma with pools of mucin and floating small glands and signet ring cells. H and E, ×100

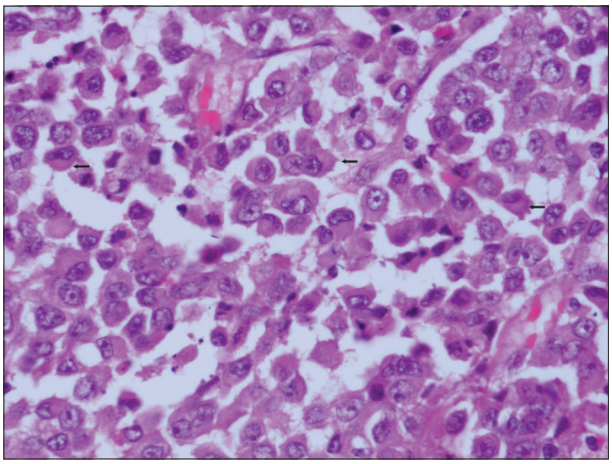

Figure 4: High-power view of solid area showing dyscohesive cells with intracytoplasmic eosinophilic inclusions (arrows), eccentric nuclei and prominent nucleoli. H and E, ×400

with solid areas. Coexpression of vimentin and cytokeratin along with the rhabdoid phenotype is thus considered to be a marker of poorer prognosis. In our case also, the tumor was widespread at the time of diagnosis and the patient died within four months despite treatment, thus confirming an aggressive biological behavior of such tumors. Recognition of the rhabdoid phenotype in gastrointestinal tract neoplasms is important because this feature is associated with poor prognosis and unresponsiveness to conventional therapy.

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