

# Gastric Carcinoma Demonstrating Rhabdoid Features

## Rabdoid Özellikler Gösteren Mide Karsinomu

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### ABSTRACT

Rhabdoid tumor, first described as an aggressive kidney tumor in infants and children and also observed less commonly in extra-renal locations, is seen very exceptionally in the gastrointestinal system. Gastrointestinal tumors with rhabdoid features are also extremely rare. A 59-year-old man with a long history of nausea, vomiting and weakness had been diagnosed as "diffuse-type gastric carcinoma" by the evaluation of endoscopic biopsy specimen at an other hospital. The total gastrectomy specimen showed an ulcerated mass measuring 7x4x4 cm located at the lesser curvature. The tumor penetrated the entire gastric wall and invaded the perigastric fat. The histopathological and immunohistochemical evaluation revealed a gastric carcinoma with rhabdoid features. The tumor was presented to emphasize the importance of clinical, morphological and immunohistochemical differential diagnosis from diffuse type gastric carcinoma as it has a very poor prognosis and is rarely seen in the gastrointestinal system.

**Key Words:** Stomach, Rhabdoid tumor, Gastric cancer

### ÖZ

İlk kez infant ve çocukların böbreklerinde agresif bir tümör olarak tanımlanan, daha az sayıda ekstra-renal yerleşimlerde de saptanan rabdoid tümör, gastrointestinal sistemde çok nadir görülmektedir. Rabdoid özellikler gösteren gastrointestinal tümörler de oldukça nadirdir. 59 yaşında erkek hasta, uzun süren bulantı, kusma, halsizlik yakınması üzerine başvurduğu dış merkezde, yapılan endoskopik biyopsi sonucu "diffüz tipte mide karsinomu" tanısı almıştır. Merkezimizde değerlendirilen total gastrektomi materyalinde küçük kurvatur yerleşimli, 7x4x4 cm ölçülerde, yüzeyi ülser, ortası kirlili sarı renkte, mide duvarını tam kat tutarak perigastrik yağ dokuya çıkmış tümöral lezyon görülmüştür. Tümörden alınan örneklerin histopatolojik ve immünohistokimyasal incelemesinde tümör "Rabdoid özellikler gösteren mide karsinomu" tanısı almıştır. Klinik, morfolojik ve immünohistokimyasal olarak diffüz tipte mide karsinomundan ayrılması gereken bu tümör gastrointestinal yerleşimde çok nadir görülmesi ve klinik olarak son derece kötü prognoz göstermesi nedeniyle önemini vurgulamak amacıyla sunulmuştur.

**Anahtar Sözcükler:** Mide, Rabdoid tümör, Mide kanseri

### INTRODUCTION

Rhabdoid tumor was first defined by Beckwith in 1978 as the rhabdomyosarcomatoid subtype of Wilms' tumor in the kidneys of infants and children (1,2). The tumor has also been reported in extra-renal locations and is seen rarely in the gastrointestinal system (2). Rhabdoid tumor makes up 0.1-0.2% of all gastric carcinomas (3). We present a case of gastric adenocarcinoma with rhabdoid features having a malignant and aggressive clinical course which could be confused with diffuse type gastric carcinoma.

### CASE REPORT

A 59-year-old male had presented to an external center for gastric pain, nausea, vomiting and weakness for approximately 45 days. His family history was insignificant and medical history revealed Type 2 diabetes mellitus

and benign prostate hyperplasia. Systemic findings were normal while routine laboratory tests showed low total protein, calcium and phosphorus levels. The endoscopic biopsy at the same center had revealed "diffuse type gastric carcinoma" and he had been referred to our hospital. Total gastrectomy was performed by the General Surgery Department and the evaluation of the total gastrectomy material at the Pathology Laboratory revealed a tumoral lesion 7x4x4 cm in size with an ulcerated surface, a dirty yellow center and peripheral hemorrhage (Figure 1). The tumor invaded the full thickness of the gastric wall and spread to the perigastric fat. Multiple histological sections from the tumor showed scattered single cells that had lost cohesiveness with a diffuse infiltrative growth pattern in a poor stoma and ulcerated surface (Figure 2). There was widespread necrosis and the tumor consisted of round or

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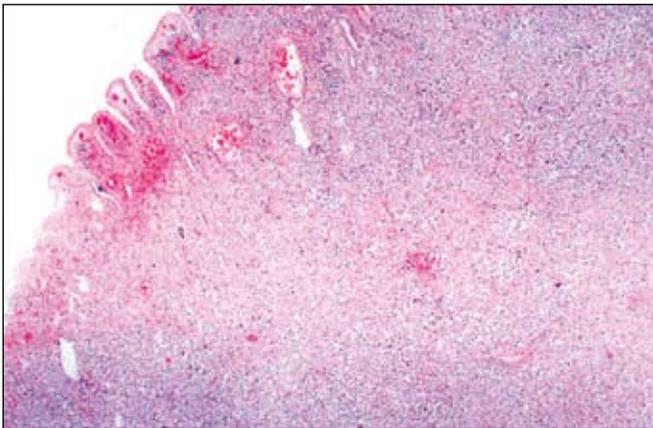
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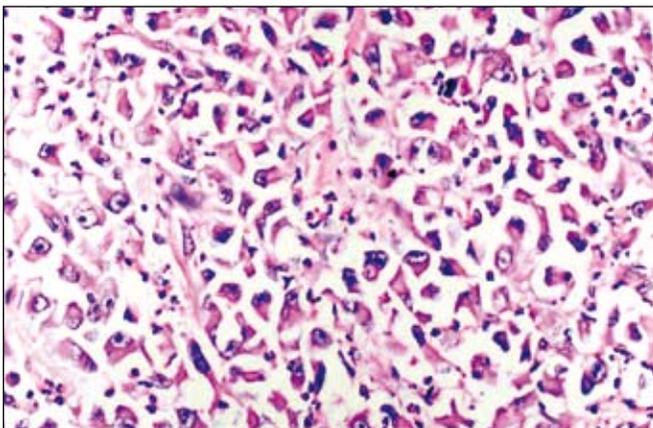
polygonal, large, vesiculated atypical cells with eccentric nuclei and prominent nucleoli in many areas. Tumor cells were seen to have a plasmacytoid appearance due



**Figure 1:** Dirty-gray tumoral lesion on the lesser gastric curvature with an ulcerated surface.

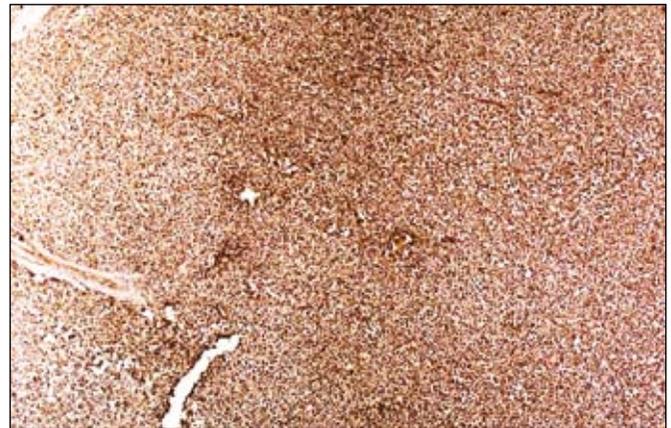


**Figure 2:** Tumor rich in cells and poor in stroma with superficial ulceration and diffuse infiltrative pattern (H&E, x40).

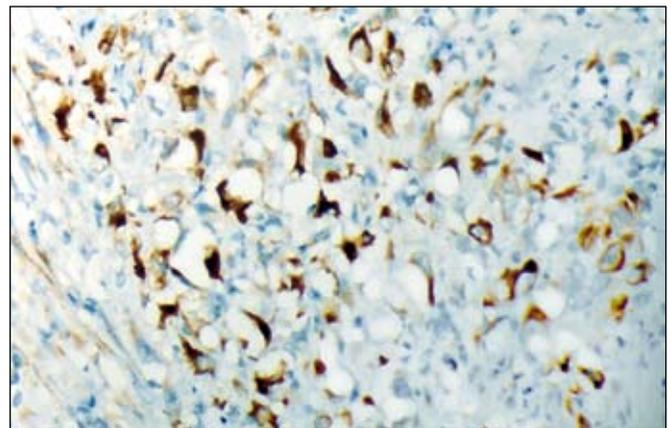


**Figure 3:** Round-polygonal tumor cells with large and eccentric nuclei, prominent nucleoli and eosinophilic inclusions in a large cytoplasm (H&E, x400).

to homogenous eosinophilic paranuclear inclusions that pushed the nuclei aside (Figure 3). There were a few cells similar to signet cells with PAS positivity in the tumor with no malignant tumoral glands. Immunohistochemical analysis showed that the tumor cells were diffusely positive for epithelial membrane antigen, vimentin and neuron-specific enolase. There was focal positivity for pancytokeratin, cytokeratin-7 and cytokeratin AE1/AE3 in individual tumor cells (Figure 4,5). Cells showing cytokeratin positivity had the same morphological appearance as those with vimentin positivity. S100, HMB45, CD117, LCA, CD138, CD-20, CD-3, actin and desmin were negative. A diagnosis of “gastric carcinoma with rhabdoid features” was made with these histopathological and immunohistochemical findings. We found tumoral infiltration showing the same characteristics in 14 of the 30 dissected lymph nodes. There was no tumor in the gastric surgical margins while the systemic screening tests showed no distant organ metastasis. Radiotherapy was



**Figure 4:** Diffuse and strong vimentin positivity in tumor cells (x40).



**Figure 5:** Pancytokeratin positivity of the rhabdoid cells (x400).

planned following evaluation at the Oncology Council but the patient got worse after 2 cycles of radiotherapy and died 6 months after the surgery.

### DISCUSSION

The term “malignant rhabdoid tumor” has been used to define a heterogenous group of neoplasms that consist of cells with “rhabdoid” cytological features. Rhabdoid tumor can develop de novo from nonneoplastic cells or following the transformation of another tumor. These tumors may therefore include non-rhabdoid tumor components and these are named “composite malignant rhabdoid tumor” (4). Extra-renal locations such as soft tissue, orbit, brain, paravertebral region, heart, breast, skin, nasopharynx, vulva, liver, pancreas, uterus, bladder, prostate, tongue, esophagus, stomach, colon, small intestine, thymus and salivary gland have been reported (1). The rhabdoid morphology consists of non-cohesive, polygonal, large cells with an eccentrically placed, large and vesiculated nucleus, a prominent nucleolus and large cytoplasm. These cells contain eosinophilic, hyaline globular inclusions at paranuclear locations that have pushed the nucleus aside in their cytoplasm. These inclusions develop as a result of the band- and coil-shaped accumulation of intermediate filaments and show both vimentin and cytokeratin immunoreactivity. The determination of cytokeratin and vimentin positivity in cells with the rhabdoid morphology is therefore important in the differential diagnosis, as in our case. Positivity of these immune markers makes it impossible to fully differentiate the epithelial or mesenchymal origin of the tumor. However, there is no myogenic differentiation either on electron microscopy or immunohistochemically in the tumor cells (2).

The presence of cells with rhabdoid morphology should bring this diagnosis to mind. The presence of a large number of rhabdoid tumor cells forming sheets was striking in our case. However, we did not come across any cellular ratio necessary to make a diagnosis of rhabdoid tumor in the literature and we therefore did not check any ratios. The differential diagnosis in our case consisted of GIST, sarcoma, sarcomatoid carcinoma, malignant melanoma, lymphoma and myeloma with the above-mentioned histological findings. However, these other diagnoses were not supported morphologically or immunohistochemically. It is important to recognize this rare gastric tumor with its morphological and immunohistochemical features because of its aggressive clinical course and unfavorable prognosis as in our case (5-9).

Renal rhabdoid tumors are seen in infants and children while extra-renal rhabdoid tumors are seen in patients of

advanced age. Schofield et al. have found mutations and deletions of chromosome 22q11-12 in most renal rhabdoid tumors and have postulated that the relevant gene plays a role in the development of renal rhabdoid tumors as a tumor suppressor gene (10). Douglass et al. have found chromosome 22 monosomy in extra-renal malignant rhabdoid tumors of the central nervous system. However, similar cytogenetic features were not found in other extra-renal locations (11). Inactivation or mutation of suppressor genes therefore still needs to be elucidated regarding the development of extra-renal rhabdoid tumors. Ota et al. have stated that rhabdoid tumors differentiate from primitive pluripotent cells, leading to the phenotypic heterogeneity (12). It is therefore debated whether extra-renal rhabdoid tumors are a subtype of poorly differentiated tumors or a separate entity. Besides the homogenous phenotype and the lack of ultrastructural features that enable a definite diagnosis, the heterogeneity of cytogenetic abnormalities indicate that these tumors are not a specific entity.

Less than 20 cases located in the gastrointestinal system have been reported until 2008 (2,5-7). Most are located in the stomach and in males with advanced age. The tumor is usually large at the time of diagnosis and the patients die with distant organ metastases within one year.

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