



Primary gastric rhabdomyosarcoma. A Case report

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Abstract: Background: Rhabdomyosarcoma (RMS) is a malignant tumor of striated muscle of mesenchymal origin regarded as the most recurrent soft tissue sarcoma in children and adolescents. We describe in this paper an unusual case of primary alveolar rhabdomyosarcoma of the stomach and review the literature. **Case presentation :** A 15-year-old girl presented with chronic gastric pain and vomiting evolving for 3 months. Computerized abdominal tomography detected enlarged celiac and lumboaortic lymph nodes. Gastroscopy revealed a friable cardiac mass. Biopsies obtained from both the gastric tumor and the abdominal lymph nodes showed clear histopathological features of an alveolar rhabdomyosarcoma. **Conclusion :** Primary alveolar rhabdomyosarcoma of the stomach is extremely rare. Better awareness of this entity is necessary for early diagnosis and treatment.

Keywords: Gastric tumor, alveolar rhabdomyosarcoma, Clinicopathological features.

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Case Report

INTRODUCTION

Mesenchymal gastric sarcomas are rare, representing < 1 % of gastric tumors. While rhabdomyosarcoma is common in children accounting for up to 50% of soft tissue sarcomas, it usually presents in the head, neck or genitourinary organs, with gastric origin rarely reported (Francis, J., & Young, P. 2017).

We present in this paper an unusual case of a 15 year old patient with primary gastric alveolar rhabdomyosarcoma along with its clinical, macroscopic and histopathological features. We also briefly review the literature.

CASE REPORT

The patient was a 15-year-old girl with no remarkable medical history, admitted to the gastroenterology department of Mohamed the VI University Hospital of Marrakesh for chronic epigastric pain, nausea, and vomiting evolving for 3 months in a context of a profound deterioration of the general state including asthenia and weight loss estimated at 10 kg / 3 months.

General examination didn't reveal any abnormalities except for tenderness in the epigastric region. No abdominal nor extra-abdominal mass was palpated. There was no lymphadenopathy.

Routine laboratory parameters were found to be normal, except for a severe hypokaliemia due to vomiting which was successfully treated.

Endoscopic examination had shown esophageal candidiasis with an inflammatory stricture of the lower esophagus, impenetrable with the endoscope, the anatomic-pathological study of which was without particularities.

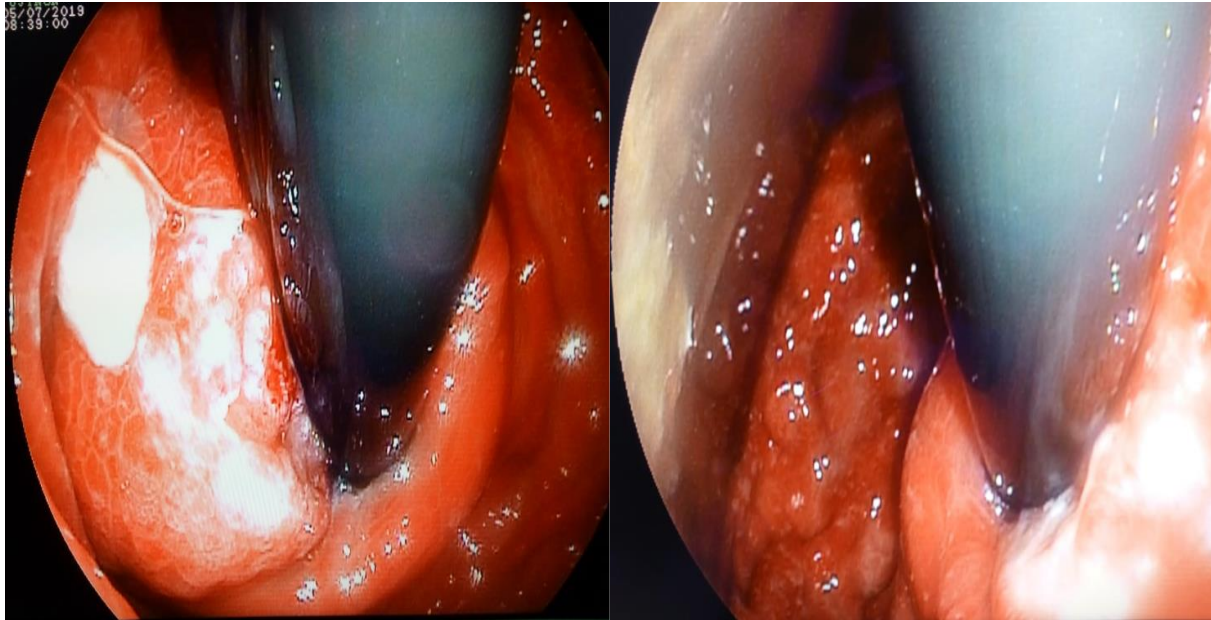
Computerized abdominal tomography detected enlarged celiac and lumboaortic lymph nodes, measuring for the largest 59 × 69 mm, hypodense, slightly enhanced by the product of contrast, adjacent to the retroperitoneal vessels and disseminated along the small gastric curvature. A CT-guided lymph node biopsy was then performed. Microscopic examination showed features of an alveolar rhabdomyosarcoma.

Gastroscopy, repeated after 14 days of intra-veinous anti-fungal drug, revealed a friable

cardial mass. The surface was markedly necrotic and bled easily on touch. The mucosa around the mass was congestive and edematous. The gross appearance was suggestive of gastric cancer. Multiples biopsies of the tumoral mass were obtained and sent for pathological examination. The

histopathology report indicated a gastric localization of an alveolar rhabdomyosarcoma.

The patient was then transferred to the oncology department to start systemic chemotherapy but died a week later probably from tumor progression.



DISCUSSION:

Rhabdomyosarcoma is a malignant tumor that arises from immature cells that are destined to form striated skeletal muscle. It is the most common soft tissue sarcoma in children and adolescents, accounting for approximately 5% of all pediatric cancers and about half of all soft tissue sarcomas. The annual incidence in children and adolescents is 4.3 cases per million children (Paulino, A. C., & Okcu, M. F. 2008).

Due to its origin in a totipotent cell, RMS not only occur in skeletal muscle but also in other locations, such as the head, neck, genitourinary tract and bile ducts (Eguía-Aguilar, P. *et al.*, 2016). Primary gastric location has been very rarely reported in the literature.

In this paper, we presented the case of a patient with primary gastric RMS. To the best of our knowledge, this is the first case of a gastric alveolar RMS seen in this part of the world. According to the available sources, less than 16 cases of gastric RMS were reported or referenced in the literature so far (Wang, Y. *et al.*, 2019 ; & Palermo, M. *et al.*, 2012).

Gastric RMS 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 (Palermo, M. *et al.*, 2012). The tendency for gastric RMS to metastasize to lymph node and the lung is in keeping with previous observations of RMS arising from other sites (Palermo, M. *et al.*, 2012). There is no indication that the gastric RMS in our case or in other case reports represented a metastasis from some other site. Indeed, metastases to stomach by RMS are uncommon.

The initial diagnosis is often difficult to establish, especially on biopsy material. In several instances, the correct diagnosis was established only at autopsy (Fox, K. R. *et al.*, 1990).

The World Health Organization (WHO) classification includes four types of RMS : embryonal RMS, alveolar RMS, spindle cell RMS, and pleomorphic RMS. The embryonal and alveolar variants are the more frequent histological types. Of these, alveolar RMS is the one with the worse prognosis (Rudzinski, E. R. *et al.*, 2015).

The alveolar subtype accounts for 31% of all cases of RMS [4]. Alveolar rhabdomyosarcomas (ARMS) usually appear in adolescence; and are typically located in the extremities with high capacity to metastasize (Paulino, A. C., & Okcu, M. F. 2008 ; & Wang, Y. *et al.*, 2019). Their histology is characterized by a septum of fibrous connective

tissue with neoplastic cells attached, similar to the alveolar spaces observed in the lung, where some of the cells detach and occupy the space. It is composed of cells uniformly polygonal with high grade round or oval hyperchromatic nucleus (Eguía-Aguilar, P. *et al.*, 2016).

Embryonal rhabdomyosarcoma accounts for 60% of cases in children and occurs mostly in the genitourinary or head and neck regions. Histologic examination shows a mixed population of small round tumor cells with hyperchromatic nuclei and large polygonal-shaped tumor cells with abundant eosinophilic cytoplasm (Wang, Y. *et al.*, 2019).

Spindle cell rhabdomyosarcoma is a subtype of embryonal rhabdomyosarcoma that accounts for 3% of all cases and occurs mostly in the paratesticular region, while Pleomorphic rhabdomyosarcoma being the least common of the four subtypes mostly occurring in the 30–50 years age-group (Wang, Y. *et al.*, 2019).

In our case, the multiple biopsies obtained from both the gastric tumor and the metastatic abdominal lymph nodes showed clear histopathological features of an alveolar rhabdomyosarcoma, which, to the best of our knowledge, has never been reported in the literature before.

There is very little information on the results of chemotherapy or other drugs in gastric rhabdomyosarcomas. Therefore, it seems that surgical treatment of these tumors is the best option (Palermo, M. *et al.*, 2012).

The type of chemotherapy that patients receive with RMS depends on the risk factors they present. Low or intermediate risk patients receive vincristine, dactinomycin, and cyclophosphamid. In high-risk patients, the combination of ifosfamide-etoposide or ifosfamide and doxorubicin is used. These patients usually have a poor prognosis (Eguía-Aguilar, P. *et al.*, 2016). Unfortunately our patient died before having the chance to start her chemotherapy probably due to tumor progression.

CONCLUSION:

To conclude, Rhabdomyosarcoma of the stomach is a rare neoplasm distinct from other sarcomas. We reported in this paper an extremely rare case of primary gastric ARMS. Better awareness of its features and the differential diagnoses will help in early diagnosis and treatment.

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