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Case Report

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Laparoscopic Resection of Malignant Perivascular Epithelioid Cell Neoplasm (PEComa) of the Gastrocolic Ligament: Case Report

Youness BAKALI MD[1], Sarah BENAMMI MD[1*], Sabrine DERQUAOUI MD[2], Mouna ALAOUI MD PhD[1], Farid SABBAH MD PhD[1], Fouad Zouidia MD PhD[2], Abdelmalek HRORA MD PhD[1], Mohamed RAISS MD PhD[1]

[1]Surgical Department "C" Ibn Sina University Hospital, Morocco
[2]Histology Department Ibn Sina University Hospital, Morocco

***Corresponding author:** Dr Sarah BENAMMI ; Surgical Departement C, Ibn Sina University Hospital, University Mohamed V of Rabat, 10000, Rabat, Morocco. Tel: +212661776062 ; E-mail: sara-benammi@hotmail.fr

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Introduction

Perivascular epithelioid cell neoplasms are rare mesenchymal tumors that can affect any part of the human body. The term PEComas nowadays represents a family of subgroups including multiple histological entities, and multiple possible anatomic sites [1,2]. Gastrointestinal tract (GI) is a common site of origin and counts for 20% to 25% [1]. However primary omental PEComa is a rare localisation with only few reported cases [3]. Clinical symptoms are not specific; and biologic behavior of this entity is still unclear [1]. PEComas are classified following number of criterias into benign, uncertain malignant potential and malignant categories. Though despite classification of the entity, standardized treatment has not been yet established. Therefore, owing to the rarity of this pathology, clinical report in this matter would be beneficiary for better knowledge of this condition. To our knowledge, we report a rare case of coelioscopic resection of a PEComa rising in the greater omentum diagnosed postoperatively.

Keywords: PEComa; Perivascular epithelioid cell neoplasms; Greater omentum PEComa; Laparoscopy

Case Presentation

A 65-year-old man with a history of hypertensive cardiomyopathy presented with mild abdominal discomfort evolving since 3 months. No additional sign or weight loss were assessed. Physical examination was standard. Tests revealed negative tumor markers ACE and CA19-9. A CT-Scan showed an exoluminal heterogenous gastric mass in the antro-pyloric region, with defined borders favoring a GIST diagnosis (Figure.1).

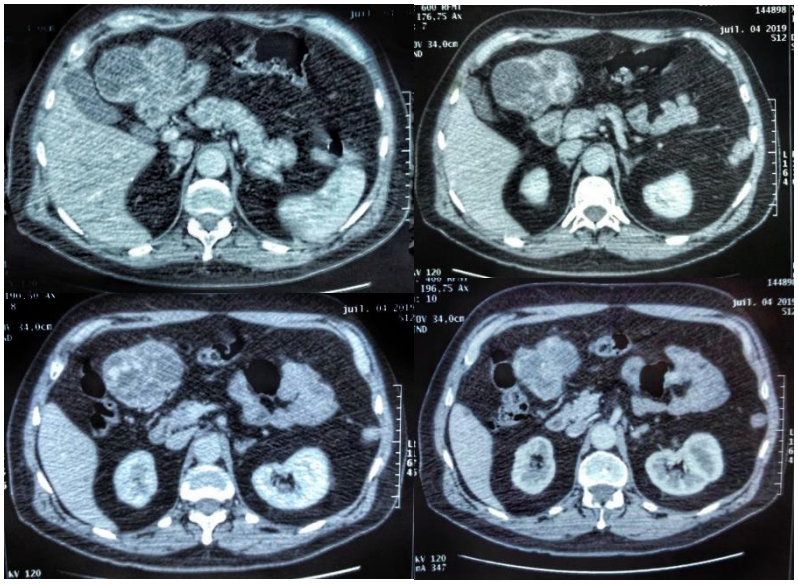

Figure 1: Abdominal CT-Scan showed an exoluminal heterogenous gastric mass in the antro-pyloric region, with defined borders favoring a GIST diagnosis.

Upper EGD exploration showed extrinsic compression in the antropyloric region. No mucosal abnormalities were observed and no biopsy was taken. Finally, the patient underwent upper gastric ultrasonography comforting the suspected diagnosis of GIST tumor of the antrum measuring 76mm. Decision was made favoring coeliospic resection. Abdominal cavity exploration confirmed absence of metastasis. After complete exposure from the umbilical ligament, the mass presented as a round multi-lobulated dark-red to light-brown tinged solid mass measuring 5cmx7cm approximately, midway between the antro-pyloric part of the stomach and the greater omuntum (Figure.2).

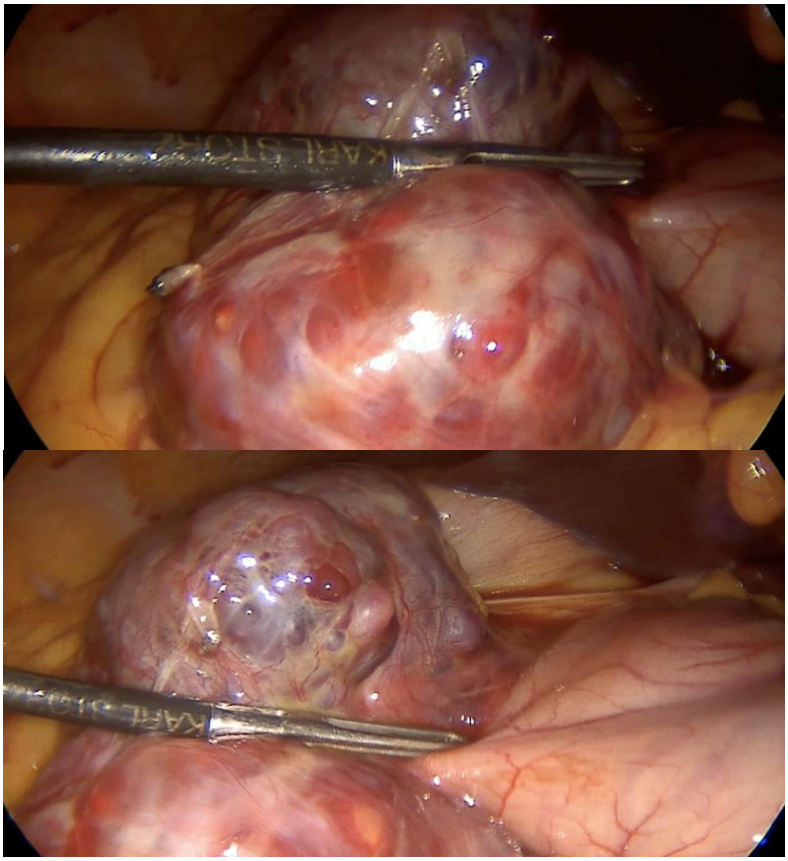

Figure 2: Round multi-lobulated dark-red to light-brown tinged solid mass measuring 5cmx7cm approximately, midway between the antro-pyloric part of the stomach and the greater omuntum.

Neither inflammatory adhesions nor extensive adhesion with the stomach where present. Therefore radical resection was possible. Immunohistochemical staining of the mass confirmed a malignant PEComa. On macroscopical examination postoperatively, the tumor measured 7.5 × 6.5 cm; it was a gray nodular and well circumscribed mass with necrotic areas. Hematoxylin-eosin stained sections revealed a tumor consisting of epithelioid ovoid cells arranged in nests or large fascicles. In some areas, the tumor was dominated by spindle cells. The cytoplasm was clear, abundant and eosinophilic. Nuclei were round showing mild pleomorphy with small nucleoli. Mitoses were estimated to 3/50 high power fields. The nests were separated by abundant capillary vessels. On immunohistochemistry, neoplastic cells stained positive for Melan-A, SMA, and H-caldesmone. They were negative for S-100 protein, cytokeratin (AE1/AE3), CD34, CD117 and Dog-1. Based on the pathological findings, the mesenteric mass was diagnosed as malignant perivascular epithelioid cell tumor. Post-operative evolution was uneventful. The patient was discharged on day 2 and remains symptom free 12 months post-operatively on follow-up. No adjuvant therapy was conducted.

Discussion

Regarding our clinical case, we report a malignant PEComa of the great omentum resected with laparoscopy surgery. Perivascular Epithelioid Cell (PEComa) tumours are mesenchymal tumours defined by the proliferation of perivascular epithelioid cells expressing melanocytic and muscle markers. Histological and immunohistochemical features are essentially identical in those tumours. The spectrum of pecoma includes: Epithelioid angiomyolipoma (AML); clear cell sugar tumour of the lungs (CCST); lymphangioleiomyomatosis (LAM); hepatic falciform ligament clear cell myomelanocytic tumors (CCMMT); and soft-tissue clear cell myomelanocytic tumors [4]. On histology, neoplastic cells have clear or eosinophilic cytoplasm and a small round, central nuclei. Nucleoli are inconspicuous. Sometimes, marked nuclear atypia are present focally [5]. Pecoma's consist of bands, nests, or swirls of either epithelioid or spindle cells [4] wich are arranged around blood vessels. Commonly peripheral cells are spindle shaped whereas periluminal cells are epithelioid [5]. Due to differences in prognosis, clinical behavior and histologic features, pecoma have been classified into benign, uncertain malignant potential and malignant categories. If the tumor shows nuclear

pleomorphism or multinucleated giant cells only, or with a size >5 cm, it is classified as pecoma of uncertain malignant potential. Malignant subtype was defined by Folpe et al. as tumors with two or more of the high risk indicators including: 1-tumor size >5 cm, 2-infiltrative growth pattern with high nuclear grade and cellularity, 4-mitotic count ≥1/50 HPF, 5-necrosis, and 6-vascular invasion [6]. The tumour in our case showed infiltrative growth, vascular invasion, nuclear atypia, and high mitotic activity (2/10 HPF) and was thus considered to have malignant potential. Definitive diagnosis requires immunostaining, the hallmark of pecoma is the co expression of SMA and melanocytic markers (HMB 45 and MELAN A) [6]. Neoplastic cells are negative for Cytokeratin and CD34. Some cases showed positivity of S-100 protein, CD56 and CD99 [6,4]. The differential diagnosis of PEComa is broad and depends on the anatomic site, it requires careful histological and immunohistochemical analysis and in some cases, genetic analysis. In fact, pecoma have to be distinguished from carcinomas, smooth muscle tumours, melanoma and adipocytes neoplasms [5,7].

First, this report describes the non-specificity of bot clinical presentation and radiological examination in addition to the few published similar cases [3]. Secondly, regarding the imprecise clinical, biological and radiological picture of this entity, management of differential diagnosis and selection of therapeutic strategy is challenging. The mainstay treatement of PEComas is essentialy surgical resection [2]. Following surgery it is not described to conduct adjuvant treatment [8]. Though choosing adequate surgical approach is key considering the unpredictability of both its diagnosis and its histological nature and prognosis. Hence our case illustrates the advantages of carrying a laparoscopic surgery approach due to the multiple conveniences it has to offer; laparoscopic surgery is less invasive, it permits the ability to explore globally the abdominal cavity for metastasis or associated lesions, and finally it is more aesthetic for the patient's postoperative scars. In addition, laparoscopic surgery is at its advantages and carries a technical ease for wedge resection of the mass considering the PEComa as a ligament tumour, therefore as a mobile mass. Finally, there is hope regarding possible medical treatement, with ongoing clinical studies to explore novel therapies in the activity of immune check point inhibitors [9].

Conclusion

Owing to the various possible locations, morphological diversity, non-specific clinical manifestation and unpredictable biological behaviour, various differential diagnosis has to be considered, therefore rending their diagnosis difficult and even more the clinical management. Given the difficulty of preoperative diagnosis, it is therefore imperative to carry out a radical surgical excision, in which case laparoscopy proved its advantages.

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