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Pulmonary Pleura Metastasis from Low-Grade Appendiceal Mucinous Neoplasm with Appendiceal Neuroendocrine Tumor: A Case Report and Literature Review

Gao J[1], Fan R[2], Lu M[1*] and Liu J[3*]

[1]Department of Pathology, Xiamen Humanity Hospital Fujian Medical University, China

[2]Department of Pathology, Xiang'an Hospital of Xiamen University, School of Medicine, Xiamen University, China

[3]Department of Toxicology, School of Public Health, Jilin University, China

[*]Corresponding author:

Mingzhi Lu,
Department of Pathology, Xiamen Humanity Hospital, No.3777 Xianyue Street, 361000, Fujian, China. E-mail address: lumingzhixmha@163.com
Jinyu Liu, Department of Toxicology, School of Public Health, Jilin University, No. 1163 Xinmin Street, Changchun, 130021, Jilin, China.
E-mail address: jy_liu@jlu.edu.cn

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Jie Gao and Ruirui Fan share the Co-first author.

Keywords:

LAMN; Metastasis; Pathological diagnosis

1. Abstract

Low-grade Mucinous Appendiceal Neoplasm (LAMN) is a rare appendiceal tumor and has low malignant potential. LAMN and Appendiceal Neuroendocrine Tumor (ANET) are the most frequent benign and malignant appendiceal lesions. A collision between LAMN and Appendiceal neuroendocrine tumor (ANET)is an exceedingly rare condition. So far, evaluating the literature, just 10 cases of collision LAMN and ANET have been reported. Here, we report a case of pulmonary pleura metastasis from LAMN with ANET that combined examinations via morphology, immunohistochemistry, and medical history. Our work will help to improve the accuracy of the diagnosis of the tumour and avoid misdiagnosis and mistreatment.

2. Introduction

Epithelial neoplasia of the appendix is grouped into LAMN, HAMN, serrated polyp, adenoma, adenocarcinoma, mucinous adenocarcinoma, and poorly differentiated adenocarcinoma with signet ring cells [1]. LAMN accounts for 73% of mucinous epithelial neoplasms. LAMN is described by the supplanting of normal appendiceal mucosa with an undulating, or flat mucinous epithelial [2]. "Pushing invasion" through the appendiceal wall represents a typical pattern for LAMN [3]. LAMN has a benign morphologic appearance and aggressive biological potential. When LAMN reaches out through the muscularis propria regularly, it results in numerous mucus on the peritoneal membrane, named Pseudomyxoma Peritoneum (PMP) [4]. However, extraperitoneal metastasis of LAMN is extremely rare. Currently, fewer than 10 cases of lung metastasis from LAMN have been reported [5]. ANET involves around 30-80% of all appendiceal tumors and is the most frequent among a wide range of NET. The vast majority of ANET have an excellent prognosis, with a 5-year survival rate near 100% in the lower tumor stages [3]. A collision between LAMN and ANET is an exceedingly rare condition. They are the consequence of two particular neoplasms, without any transition zone, resulting from the proliferation of two different cellular lines. Reviewing the literature, only 10 other cases of collision LAMN and ANET are reported worldwide [6].

In our case, we describe the histopathological aspects of an unusual pulmonary pleura metastasis from LAMN with ANET. Accurate and standardized pathological diagnosis of LAMN is very important for the selection of appendiceal treatment.

3. Case Report

A 48-year-old Chinese woman was found to have an elevated CEA of about 6 ng/ml during a physical examination and was followed up regularly. The CEA increased gradually every 3-6 months and is now as high as 24.75 ng/ml. 2 months ago, PET-CT revealed a tumorous lesion in the right pleura, an appendiceal lesion, and a pelvic effusion. Open exploratory surgery revealed an enlarged appendix with a 3.5×2 ×2cm mass, a red lesion of approximately 3×1cm in extent on the plasma surface of the right mid-ascending colon, and

scattered mucus on both the omentum and the right suprahepatic diaphragm, with no significant abnormalities on the rest of the diaphragm. Right hemicolectomy, total hysterectomy, and double adnexectomy were performed. Histopathologic evaluation in LAMN showed the normal mucosa of the appendix was replaced with an undulating epithelial monolayer of uniform columnar cells with apical mucin and basal hyperchromatic nucleus. LAMN tumour cells are close to the sclerotic fibrous stroma rather than lamina propria with low-grade atypia (Figure-1A). In ANET, large and small nests were composed of polygonal cells with salt-and-pepper chromatin and cytoplasmic brightly eosinophilic basally-located granules. No mitosis was observed (Figure-1B). The results of immunohistochemistry of the Ki-67 showed a low percentage (1%) of ANET cells staining (Figure-1C). Immunohistochemistry showed ANET cells were positive for Syn and SSTR (Figure-1D). The morphology of tumour cells in the pulmonary pleura was consistent with LAMN(Figure-1E). Elastic fiber staining showed that the pulmonary pleura was intact, indicating that LAMN was transferred to the pulmonary pleura via the esophageal lacuna (Figure-1F). In conclusion, a clinical diagnosis of pulmonary pleura metastasis from LAMN with ANET was confirmed by expert consultation. The treatment of advanced tumour is also different from LAMN and ANET and affects the prognosis and survival of patients. Therefore, an accurate and standardized pathological diagnosis of this collision tumour is very important for the selection of the treatment.

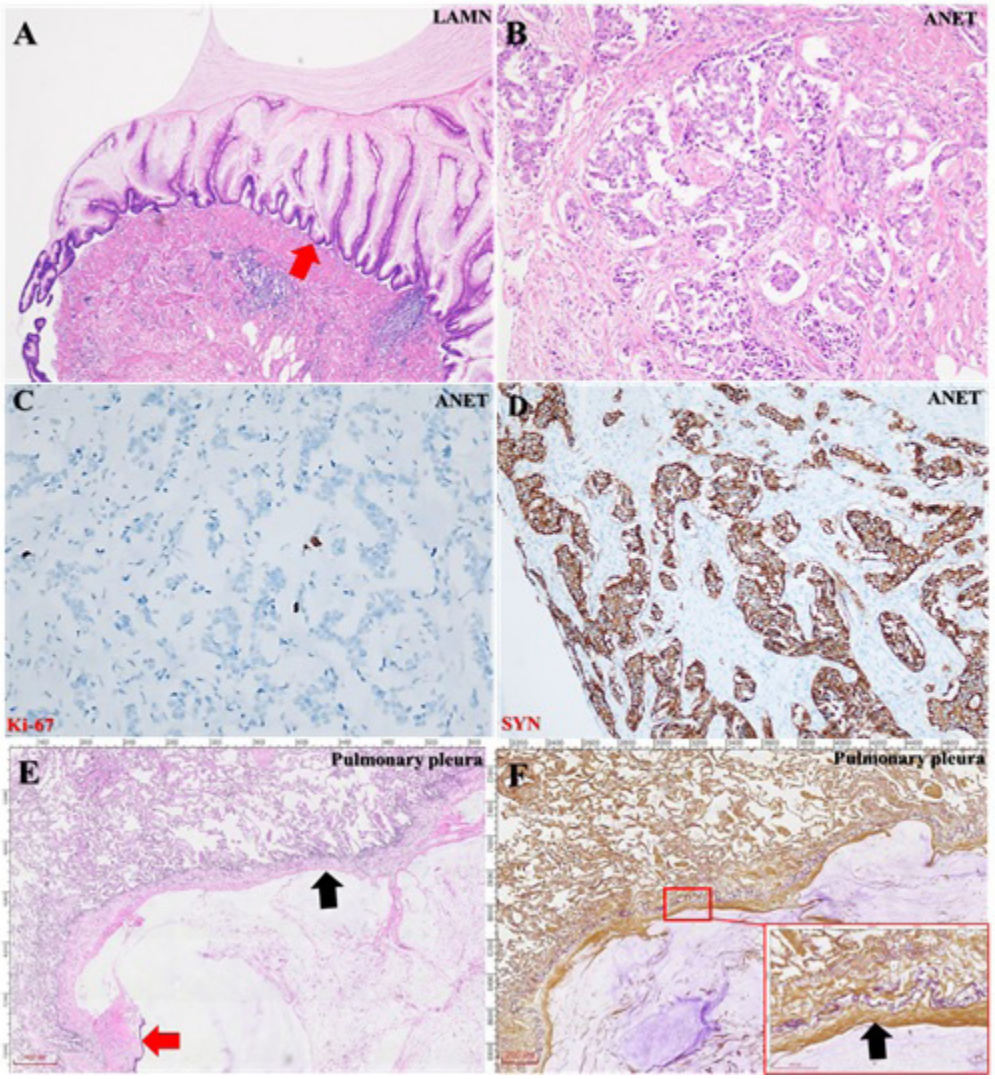

Figure 1: (A) HE staining in LAMN. The normal mucosa was replaced with an undulating epithelial monolayer of uniform columnar cells with apical mucin. **(B)** HE staining in ANET. Large and small nests were composed of polygonal cells with salt-and-pepper chromatin and eosinophilic granules. **(C)** Immunohistochemical staining in ANET for Ki67 is 1%. **(D)** ANET cells were positive for SYN. **(E)** Pulmonary pleura HE. The morphology of tumour cells in the pulmonary pleura was consistent with LAMN (red arrow). The pulmonary pleura was intact (black arrow). **(F)** Elastic fiber staining showed that the pulmonary pleura was intact (black arrow).

4. Discussion

LAMN has low-grade cytological characteristics, however, it is easy to develop into PMP with recurrence and metastasis, so regular follow-up is required. Attention should be paid to observing the position of the tumour and the margin. Early and accurate diagnosis is of great significance for the prognosis of patients. Therefore, pathologists especially need to make a distinction between LAMN and the following types of similar tumours. It is difficult to distinguish appendiceal mucinous tumours from ovarian mucinous tumours because of the close location and the nonspecific serum tumour markers, increasing the challenge of diagnosing LAMN [7]. Prominent features of LAMN are atrophy of submucosa and lamina propria at the lesion, but the mucosal muscular of serrated polyps/adenomas is intact [8]. HAMN has papillary and sieve structure, and the tumour cells showed considerable atypia, polymorphonuclear and pathological mitosis [8]. In immunohistochemistry, LAMN usually expresses CDX2, and SATB2. Without PAX8 expression, LAMN can be distinguished from ovarian mucinous tumours [9]. There are usually KRAS / GNAS co-mutations in LAMN, and ovarian mucinous tumours are usually accompanied by KRAS mutations [10].

There is no consensus regarding surgical treatment for LAMN. In this case, the patient underwent right hemicolectomy, omentum resection, cauterization of peritoneal lesions, total hysterectomy, bilateral adnexal resection, and HIPEC. Currently, follow-up is recommended to start yearly, including an abdominal computed tomography scan and determination of serum tumour markers. Furthermore, CA125 and CA19-9 are significant prognostic elements to predict patients' prognosis [11].

References

1. Xiao J, Li P, Liu W. Analysis of Clinical Characteristics of Low-Grade Appendiceal Mucinous Neoplasm (LAMN): A Retrospective Cohort Study of 51 LAMN Patients[J]. J Invest Surg. 2021; 34(7): 721-7.
2. Yanai Y, Saito T, Hayashi T, et al. Molecular and clinicopathological features of appendiceal mucinous neoplasms[J]. Virchows Arch. 2021; 478(3): 413-26.
3. Villa M, Sforza D, Siragusa L, et al. A Low-Grade Appendiceal Mucinous Neoplasia and Neuroendocrine Appendiceal Collision Tumor: A Case Report and Review of the Literature[J]. Am J Case Rep. 2021; 22: e927876.
4. Li X, Zhou J, Dong M, et al. Management and prognosis of low grade appendiceal mucinous neoplasms: a clinicopathologic analysis of 50 cases[J]. Eur J Surg Oncol. 2018; 44(10): 1640-5.
5. Xu L, Burke A. Pulmonary metastasis from a low grade mucinous appendiceal neoplasm[J]. Pathology. 2013; 45(2): 186-8.
6. Sugarbaker PH, Ben-Yaacov A, Hazzan D, et al. Synchronous primary neuroendocrine and mucinous epithelial tumors present in the same appendix. Case report of 2 patients[J]. Int J Surg Case Rep. 2020; (67): 76-79
7. Zhang W, Tan C, Xu M, et al. Appendiceal mucinous neoplasm mimics ovarian tumors: challenges for preoperative and intraoperative diagnosis and clinical implication[J]. Eur J Surg Oncol. 2019; 45(11): 2120-5.
8. Givalos N, Manolidis G, Kochylas N, et al. Unusual case of intraluminal cecal recurrence of a low grade appendiceal mucinous neoplasm (LAMN)—Case report and brief literature review[J]. Int J Surg Case Rep. 2018; 51:112-116.
9. Aldaoud N, Erashdi M, AlKhatib S, et al. The utility of PAX8 and SATB2 immunohistochemical stains in distinguishing ovarian mucinous neoplasms from colonic and appendiceal mucinous neoplasm[J]. BMC Res Notes. 2019; 12(1): 770.
10. Bastien MC, Blanchard F, Lamy A, et al. A case of Pseudomyxoma Peritonei of an unexpected origin[J]. Diagn Pathol. 2021; 16(1): 119.
11. Perivoliotis K, Christodoulidis G, Samara AA, et al. Low-Grade Appendiceal Mucinous Neoplasm (LAMN) Primarily Diagnosed as an Ovarian Mucinous Tumor[J]. Case Rep Surg. 2021; 2021: 5523736.