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A novel case of inflammatory pseudotumor of the spleen with concurrent invasive lepidic pulmonary adenocarcinoma: a case report

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Abstract

Background Inflammatory pseudotumor of the spleen and lepidic adenocarcinoma of the lung are uncommon slow-growing malignancies that have not been previously reported to occur concurrently.

Case presentation We present the case of a 63-year-old Caucasian man who presented with a splenic inflammatory pseudotumor-like dendritic cell sarcoma and was found to have a concomitant invasive lepidic adenocarcinoma of the lung. The patient underwent laparoscopic splenectomy to address the splenic mass. Three months later, he underwent video-assisted thoracoscopic surgery, wedge resection, superior segmentectomy, and mediastinal lymph node dissection to manage the lung lesion. Final pathology revealed pT1c, N0, M0, stage IA3 lepidic adenocarcinoma. The patient received his post-splenectomy vaccinations and will repeat a computed tomography of the chest 6 months postoperatively for pulmonary surveillance.

Conclusions This report highlights the indication for surgical intervention in the management of splenic masses, as well as the importance of early operations for low-grade splenic lesions. The simultaneous occurrence of inflammatory pseudotumor of the spleen and lepidic adenocarcinoma of the lung sheds light on the need for comprehensive evaluation and multidisciplinary treatment strategies for patients with rare concurrent malignancies. This case report may also be corroborated by future similar reports that may unfold a discovery of a genetic association or syndromic disorder. This case underscores the critical role of surgical intervention and thorough evaluation in patients with rare concurrent malignancies, such as splenic inflammatory pseudotumor and lepidic adenocarcinoma. Future cases may reveal potential genetic or syndromic links, further guiding treatment and surveillance strategies.

Keywords Lepidic adenocarcinoma (LPA), Inflammatory pseudotumor (IPT), Segmentectomy, Video-assisted thoracoscopic surgery (VATS), Splenectomy, Pulmonary function testing (PFT)

Background

Inflammatory pseudotumor (IPT) of the spleen is an uncommon disorder, as is lepidic adenocarcinoma (LPA), which makes up a small subgroup of invasive pulmonary adenocarcinoma. To date, and to the best of our knowledge, there are no documented cases of concomitant IPT of the spleen and invasive lepidic lung adenocarcinoma. IPT of the spleen is a rare, slow-growing neoplasm composed of proliferating cells known as follicular dendritic cells. This tumor is typically asymptomatic in its initial stages and may only be discovered incidentally during routine imaging or physical examination. As the tumor grows, it may cause pain or discomfort in the upper left abdomen and can lead to anemia or other blood disorders [1]. The tumor is strongly associated with

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Epstein–Barr virus (EBV) and lacks other common mutations such as *BRAF*, *NRAS*, *KRAS*, and *IDH2* genes, and rearrangements of *ALK* and *ROS1* genes [2]. Computed tomography (CT) and ultrasound (US) are effective imaging techniques for identifying IPT [3].

Lepidic adenocarcinoma of the lung is a subtype of lung adenocarcinoma characterized by a growth pattern in which cancer cells proliferate along the alveolar walls without invading the surrounding lung tissue [4]. The 5-year survival rate of this subtype ranges from 82% to 100%, depending on the extent of invasion [5]. Invasive LPA of the lung may require more aggressive surgical or systemic therapy depending on the extent of invasion and molecular characteristics. CT, position emission tomography- computed tomography (PET-CT), and magnetic resonance imaging (MRI) are imaging techniques used for diagnosis and staging and, with molecular biomarkers such as *EGFR* mutations and *ALK* rearrangements, may guide treatment decisions [6].

We report a rare case of concomitant IPT of the spleen and invasive lepidic adenocarcinoma of the lung. To our knowledge, this is the first case report of these two rare tumors occurring simultaneously in the same patient. In this report, we describe the clinical, radiographic, and pathologic features of this unique case, and discuss the management and treatment options for these two rare tumors.

Case presentation

We present the case of a 63-year-old non-Hispanic white male with past medical history of migraines, benign prostatic hyperplasia, and prior 15 pack-year smoking history, who presented with several months of abdominal pain. In January 2016, the patient had an ultrasound (US) of the abdomen to further investigate the source of the pain. Abdominal US showed a 5.4 cm × 4.8 cm heterogeneous, well-defined, hypoechoic focus in the superior pole of the spleen with evidence of cystic changes. Subsequently, the patient underwent an abdominal computed tomography (CT) scan, which demonstrated a splenic lesion, suspicious for benign sclerosing angiomatoid nodular transformation, which is consistent with splenic hamartoma with cystic degeneration. At that time, no surgery was indicated or offered. However, a repeat abdominal CT was performed in 2019 for persistent abdominal pain and demonstrated a splenic lesion that appeared more aggressive in nature (Fig. 1). The patient underwent a comprehensive workup that included CT scans of the chest, abdomen, and pelvis which showed lung nodules. Owing to the patient's smoking history and new onset of lung nodules, the patient requested to be evaluated for his pulmonary complaints first but sought a second opinion. After a joint discussion with the cardiothoracic surgery and surgical oncology teams, the patient deferred further investigation owing to the lung nodules' small size and benign appearance, and decided to first undergo treatment for the splenic lesion. The patient was followed by surgical oncology from 2020. In 2021, a PET-CT scan was performed and demonstrated avidity in the 6-cm splenic mass (SUV 4.0), as well as a nonavid 1.8 cm × 1.1 cm right lower lung nodule. The case was discussed at a multidisciplinary tumor board. Despite the stable size of the splenic lesion over many years, subsequent imaging showed new, solid-appearing components, which were different from the prior cystic appearance, and it was deemed appropriate to perform a splenectomy. The patient underwent a laparoscopic splenectomy with removal of a retroperitoneal mass involving the left Gerota's fascia in March 2022. The histopathology report revealed spindle cell proliferation with collagen and abundant inflammatory cells.

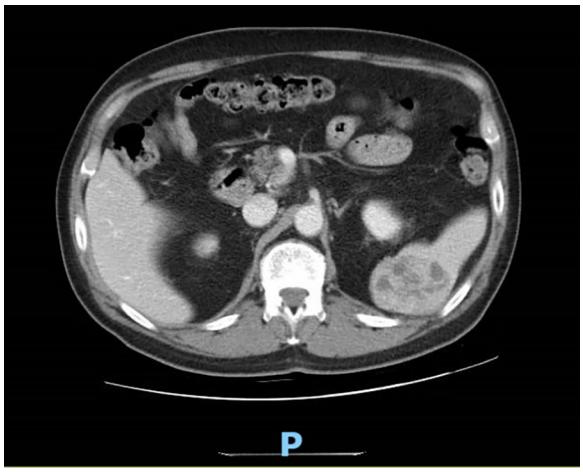

Fig. 1 Lower pole heterogeneous solid splenic mass that measures 6.2 cm × 4.6 cm × 6.2 cm

Prior to surgical intervention for the pulmonary nodule, preoperative pulmonary function testing (PFT) was performed. The predicted postoperative forced expiratory volume (poFEV1) was 2.95 L, and the predicted postoperative diffusing capacity of the lungs for carbon monoxide (poDLCO) was 23.33 ml/(minute mmHg). PFT demonstrated maximum vital capacity (VCMax) of 4.18 L. In June 2022, the patient underwent flexible bronchoscopy and robotic-assisted right thoracoscopy for evaluation and treatment of the right pulmonary nodule. On thoracoscopy, the right pulmonary nodule was identified, and a right lower wedge resection was performed to send for frozen-section analysis, which revealed adenocarcinoma. This prompted a completion right upper lobe superior segmentectomy with mediastinal lymph node dissection of levels 4 and 7–11 was performed (Fig. 2).

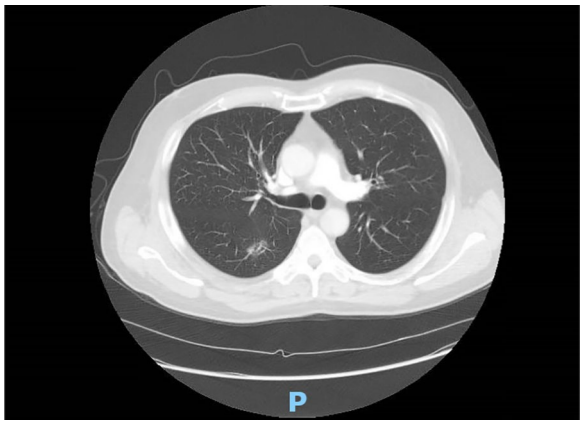

Fig. 2 Mixed cystic solid lesion in the superior segment of the right lower lobe abutting the fissure, with fissural retraction spanning approximately 2.2 cm in length

Intraoperatively, lung expansion was assessed and demonstrated no air leak. At the conclusion of the case, there was successful placement of a 28-French chest tube and connection to Pleur-evac. The postoperative course was complicated by a right-sided pneumothorax after chest tube removal, but the pneumothorax resolved with supplemental oxygen use through nasal cannula. The patient was discharged on postoperative day 3, with instructions for repeat chest X-rays and a 2-week follow-up visit. Final pathology was reviewed in an intradepartmental conference and was sent out for national cancer center revision, with a consensus opinion of lepidic adenocarcinoma of the right lung pT1c, N0, M0 (17 nodes negative), stage IA3 and splenic inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma. The final pulmonary margins were found to be negative. The patient remains recurrence free at 24 months postoperatively and will continue to complete serial CT chests every 6 months for his long-term follow-up plan. The splenic lesion required no further treatment. The patient completed his postsplenectomy vaccinations and will receive repeat imaging only if recurrent abdominal symptoms develop.

Discussion and conclusions

Inflammatory pseudotumor of the spleen is an extremely rare neoplasm composed of follicular dendritic cells. Since its first description by Cotelingam and Jaffe in 1984, only 48 cases of inflammatory pseudotumor-like sarcoma have been reported in the literature, with 67% of cases in the spleen [7]. Clinical manifestations of splenic IPT can vary, with some patients presenting with abdominal pain, bloating, abdominal mass, weight loss, fever, fatigue, and anorexia, while others may be asymptomatic [7]. It is important to note that splenic tumor infiltration can lead to marked peripheral thrombocytopenia owing to splenic platelet retention. The tumor is strongly associated with Epstein–Barr virus (EBV) [2].

Diagnosis typically involves imaging showing a well-defined lesion, though these tests often cannot rule out malignancy. Biopsy is considered if malignancy remains a concern, though it carries bleeding risks. For asymptomatic, small lesions, active surveillance with periodic imaging is often sufficient. However, larger or symptomatic lesions generally warrant intervention owing to potential complications such as rupture. Prognostic factors associated with a worse outcome include large tumor size ($\geq$6 cm), coagulative necrosis, high mitotic count ($\geq$ five mitoses per ten high-power fields), and significant cytological atypia [9]. A pooled analysis of the literature has reported local recurrence and distant metastasis rates of 28% and 27%, respectively [8, 9].

Surgical resection, typically laparoscopic splenectomy combined with regional lymphadenopathy, is preferred for IPTs that are symptomatic, large, or at high risk of rupture. This has shown potential for reducing postoperative recurrence and metastasis, thereby improving long-term survival rates [9]. Postsplenectomy patients require vaccinations against encapsulated bacteria and may need long-term antibiotics to prevent infection risks. Educating this cohort of patients on infection risk and proper preventive care is essential to ensuring long-term health.

Lepidic adenocarcinoma (LPA) is a variant of non-small cell lung cancer that typically originates in the lung periphery and demonstrates tracking alongside preexisting alveolar walls. Histologically, LPA tumors exhibit nonmucinous proliferation of type II pneumocytes and/or Clara cells along alveolar walls, often accompanied by involvement of vessels or the pleura [10]. LPA frequently presents as solitary or multifocal ground-glass lesions [12]. This characteristic finding is associated with improved resectability, favorable prognosis, and reduced propensity for nodal migration and extrathoracic involvement [11]. The presence of a small lesion size, ground-glass opacities, and air bronchograms on CT scans are considered favorable prognostic factors, while spiculations, pleural retractions, and lesions larger than 3.0 cm are associated with unfavorable outcomes [11–13]. The management of LPA generally involves surgical resection with an average 5-year survival rate of 90% [11]. The case presented here supports the existing literature, as the patient underwent video-assisted thoracoscopic surgery with wedge resection and segmentectomy for the $<$2 cm LPA lesion. This approach aims to achieve complete tumor removal while preserving lung function.

When discussing indications for surgery for splenic masses and less aggressive lung lesions, it is important to consider individual patient characteristics, tumor

features, and potential risks and benefits of surgical intervention. Splenic masses may require surgical excision, particularly in cases where there is suspicion of malignancy, significant symptoms, or risk of complications. As for less aggressive lung lesions such as LPA, the favorable prognosis associated with surgical resection supports the recommendation for early operations on low-grade pulmonary lesions.

Abbreviations

IPT	Inflammatory pseudotumor
VATS	Video-assisted thoracoscopic surgery
US	Ultrasound
CT	Computed tomography
PFT	Pulmonary function test
poFEV1	Postoperative forced expiratory volume
poDLCO	Postoperative diffusing capacity of lungs for carbon monoxide
VCMax	Maximum vital capacity
EBV	Epstein–Barr virus
PET	Positron emission tomography
MRI	Magnetic resonance imaging
SUV	Standardized uptake value
RUL	Right upper lobe
RLL	Right lower lobe
LPA	Lepidic adenocarcinoma

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Author contributions
Singer, Kiara: literature review, case report, draft, and manuscript preparation. Ranjbar, Tara: literature review, draft, and manuscript preparation. Aweeda, Marina: literature review, draft and manuscript preparation. Azab, Basem Nady: draft and manuscript preparation. Villa Sanchez, Manuel: draft and manuscript preparation.

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Not applicable.

Patient consent
Patient consent is available upon request.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of this written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that they have no competing interests.

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