Inflammatory pseudotumor mimicking chronic pulmonary embolism or pulmonary artery sarcoma: Report of five cases

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Abstract
Inflammatory pseudotumor (IPT), also known as plasma cell granuloma, is a rare lesion of unknown etiology that occurs in many organs, especially in the lung. Here we report five cases of IPT arising in pulmonary artery mimicking chronic thromboembolic disease, not previously documented in the literature. Those cases were identified at our institute among over 2500 pulmonary endarterectomy (PEA) specimens acquired from 2000 to 2017. The cohort included three men and two women with a median age of 41 years (range: 23–54). All patients presented with dyspnea and radiologic findings of pulmonary artery thromboembolism, some concerning for intimal sarcoma. The duration between disease onset and PEA ranged from 6 months to approximately 3 years. Histologically, all cases showed proliferation of spindle cells with marked inflammatory infiltrates composed predominantly of plasma cells, histiocytes, and small lymphocytes. Ancillary studies were performed in each case and ruled out other possibilities, such as sarcoma, lymphoma, plasmacytoma, IgG4-related disease, and infection. IPT arising in pulmonary artery presenting clinically as acute or chronic thromboembolic disease is very unusual, in which clinical data, radiographic findings, and histopathologic features have to be integrated for reaching the proper diagnosis.

Key words
ALK-1, chronic thromboembolic pulmonary hypertension, IgG4, inflammatory pseudotumor, pulmonary embolism, pulmonary endarterectomy

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INTRODUCTION

Inflammatory pseudotumor (IPT), or plasma cell granuloma, is a rare benign tumor-like condition first reported in lungs by Drs. Bahadori and Liebow,1 who described a localized proliferation of “mature plasma cells supported by a stroma of granulation tissue” simulating a neoplasm. IPT refers to an inflammatory, reactive, or regenerative process that often forms a mass and mimics malignancy clinically and radiologically. Histologically, IPT features polyclonal plasma cell infiltrates admixed with spindle fibroblastic/fibrohistiocytic proliferation that lacks nuclear atypia or increased mitotic activity. It should be distinguished from the inflammatory myofibroblastic tumor (IMT), a neoplastic process characterized by myofibroblastic proliferation with variable nuclear atypia and mitotic rate. ALK gene rearrangement is reported in approximately 60% of IMT and a subset of ALK-negative IMT may harbor other genetic rearrangements involving ROS1 or RET.2 While the terms of IPT and IMT have been used synonymously or interchangeably in old literature, it is now widely accepted that they are two different entities, and the diagnosis of ITP is a process of exclusion. IPT has been described in many anatomic sites, including lung, head and neck, genitourinary tract, breast, and liver. However, IPT occurring in vasculature has not been described in the literature. Our institution is a major center for the management of patients with the chronic thromboembolic disease (CTED) in the absence of resting pulmonary hypertension, as well as those with chronic thromboembolic pulmonary hypertension (CTEPH), having performed over 2500 pulmonary endarterectomies (PEA) from 2000 to 2017. Herein, we report five cases of IPT identified among those cases. The clinical, laboratory, imaging, and histopathologic findings of each case are summarized in Tables 1 and 2.

CASE DESCRIPTION

Case 1

A 41-year-old female with a past medical history of hypothyroidism presented to an outside hospital with abrupt onset malaise, diffuse myalgias, shortness of breath, and chest pain. She was diagnosed with acute pulmonary embolism (PE) by CT angiography (CTA) and deep vein thrombosis (DVT) by ultrasound. A ventilation perfusion (VQ) scan showed a complete absence of perfusion of the right lung (Figure 1). She had ongoing chest pain and follow-up imaging after 3 months of anticoagulation at another center was reported to show complete obstruction of the proximal right main pulmonary artery with eccentric filling defects in the left upper lobe felt to be compatible with chronic PE. The patient was subsequently referred to our institution for evaluation. Our review of the outside CTA revealed an infiltrating mass-like lesion that completely occluded the right main pulmonary artery (PA) with possible arterial wall involvement (Figure 1). Right heart catheterization yielded normal hemodynamics (Table 2). She was referred for surgical management of possible PA sarcoma. At surgery, she was found to have a mass invading the right main PA wall with complete vessel occlusion and extension into the subsegmental branches of all lobes on the right. She underwent a right main PA resection with Gore-Tex graft reconstruction and PEA of the right and left lungs. Small, subsegmental occlusions in the left upper lobe were removed with PTE. Grossly, the resected right PA measured 8.0 cm in size and showed a friable dark brown soft tissue mass filling the lumen (Figure 2a). Microscopic sections of these vascular contents revealed dense lymphoplasmacytic infiltrate with a few foci of bland-appearing spindle cell proliferation (Figure 2b). The lymphoplasmacytic infiltrate involved the artery wall and extended to the adventitia. The spindle cells were positive for smooth muscle action (SMA) but negative for ALK-1. The plasma cells were polytypic by immunostain, with less than 30% positive for IgG4 (Figure 2c–f). The material from the left upper lobe was consistent with the artery wall. Surgery resulted in partial reperfusion of the right middle and lower lobes. The patient was doing well 5 years after surgery on follow-up.

Case 2

A 54-year-old male with a past medical history significant for pituitary prolactinoma and intra-abdominal diffuse large B-cell lymphoma with an incidental asymptomatic right-sided PE diagnosed on CTA during routine screening for lymphoma recurrence (11 years after treatment) at an outside hospital. He was treated with 1 year of warfarin, then discontinued. One year later he developed new dyspnea and cough and was diagnosed with a PE described as occluding the right interlobar artery. Evaluation for DVT was negative. He was placed back on anticoagulation, but follow-up imaging demonstrated an increase in size of the obstruction with vessel distension, while evaluation for thrombophilia was negative (Table 1). At the time of referral, he described dyspnea only with heavy exertion. He did not have pulmonary hypertension on right heart catheterization. He underwent PEA due to suspicion of a possible neoplasm. At surgery, he was found to have a mass that grossly looked like a tumor rather than clot, with no abnormalities identified in the left lung. Grossly, the specimen consisted of multiple fragments of tan-yellow to brown hemorrhagic friable soft tissue. Microscopic examination showed abundant plasma cells mixed with scattered foamy histiocytes and spindle cells (Figure 3a). The spindle cells were positive for SMA while
Table 1: Summary of clinical and histopathologic findings

| Case no. | Age | Sex | Past medical history | Time from diagnosis | Thrombophilia | Histology | Immunohistochemistry results | Lymphoma workup | Other ancillary studies | IgG4/ IgG ratio | Follow-up | Outcome |
|----------|-----|-----|----------------------|---------------------|---------------|-----------|-------------------------------|-----------------|------------------------|----------------|-----------|---------|
| 1        | 41  | F   | Hypothyroidism       | 6 months            | Negative      | Plasma cell dominant          | (+): SMA, (−): ALK-I, S100 | Kappa, lambda, CD138, CD3, CD5, CD10, CD19, CD20 | EBV ISH; HHV-8 by LANA; molecular testing for syphilis; FISH for ALK-1 rearrangement | <30%         | 5 years   | Alive  |
| 2        | 54  | M   | Pituitary adenoma, DLBCL | 2 years            | Negative      | Plasma cell dominant          | (+): SMA, (−): ALK-I, S100, Desmin, CD31, CD34 | Kappa, lambda, CD3, CD20, IgA, IgM | EBV ISH; Warthin-Starry; GMS; AFB | <30%         | 6 years   | Alive  |
| 3        | 23  | M   | None                 | 3 years             | Positive      | Plasma cell dominant          | (+): SMA, CD68, (−): ALK-I, Desmin | Kappa, lambda | Congo red; Warthin-Starry; AFB; GMS | n.a.          | 2 years   | Alive  |
| 4        | 31  | M   | None                 | 6 months            | Negative      | Plasma cell dominant          | (+): SMA, CD68, (−): ALK-I, Desmin, CD31, CD34, factor VIII, p53 | Kappa, lambda | Iron; trichrome; elastin | n.a.          | 7 years   | Deceased |
| 5        | 47  | F   | None                 | 1 year              | Negative      | Stromal dominant              | (+): SMA, CD68, (−): S100 | CD3, CD20 | Trichrome; elastin | n.a.          | 4 years   | Deceased (unknown cause) |

Abbreviations: DLBCL, diffuse large B-cell lymphoma; F, female; LAC, lupus anti-coagulant; M, male; n.a, not performed or data not available; (+), positive IHC; (−), negative IHC.

*Time between acute PT diagnosis and surgery.

Thrombophilia evaluation included antithrombin III, Protein C, Protein S, and anticardiolipin IgG levels and lupus anticoagulant.
**TABLE 2** Pre- and postoperative pulmonary hemodynamics and preoperative imaging

| Subject | RAp (mmHg) | PA (mean) (mmHg) | Cardiac output (L/min) | PVR (WU) | Imaging |
|---------|------------|------------------|-----------------------|----------|---------|
| 1 Pre-op | 3          | 35/15 (24)       | 5.3                   | 2.6      | Invasive, obstructing mass R main PA |
| Post-op | 11         | 34/13 (22)       | 5.0                   | 2.2      |         |
| 2 Pre-op | 7          | 26/10 (16)       | 5.6                   | 1.2      | Enlarging mass R interlobar artery |
| Post-op | 8          | 22/12 (16)       | 5.8                   | 1.4      |         |
| 3 Pre-op | 7          | 82/25 (43)       | 7.1                   | 6.4      | Enlarging mass in L and R main PAs |
| Post-op | 6          | 33/14 (22)       | 7.6                   | 2.1      |         |
| 4 Pre-op | 24         | 51/34 (40)       | 2.4                   | 11.5     | Large bilateral proximal filling defects, bilateral lower lobe obstruction |
| Post-op | 7          | 46/18 (26)       | 4.0                   | 3.4      |         |
| 5 Pre-op | 3          | 53/20 (33)       | 5.7                   | 4.4      | Obstruction right main PA, disease LLL |
| Post-op | 10         | 39/16 (25)       | 5.2                   | 2.9      |         |

Abbreviations: L, left; LLL, left lower lobe; PA, pulmonary artery; PAp, pulmonary artery pressure; PVR, pulmonary vascular resistance; R, right; RAp, right atrial pressure; WU, Wood units.

**FIGURE 1** Radiology characteristics of Case 1. (a) An axial image from a contrast-enhanced chest computed tomography (CT) shows a complete filling defect in the right main pulmonary artery (arrows). It is atypical for bland pulmonary thromboembolic disease to result in unilateral occlusion of a main pulmonary artery. (b) A coronal maximum intensity projection (MIP) image demonstrates dilated bronchial arteries (arrows) coursing towards the right lung. (c) A frontal chest radiograph shows diminutive right hilar/pulmonary vessels compared to the left. (d) A VQ scan demonstrates the absence of perfusion in the right lung. Likewise, both CT images show diminished blood flow in the right lung.
negative for S100 and ALK-1. The patient was healthy and alive with a negative positron emission tomography (PET) scan 6 years after surgery on follow-up. Post-op perfusion imaging showed no change in perfusion and repeat CTA demonstrated acute thrombosis in the interlobar and right lower lobe artery. At 6 years follow-up the patient is reported to be asymptomatic and yearly PET imaging has been negative.

Case 3

A 23-year-old male with a history of sickle trait, prior pulmonary embolus with IVC filter placement 2 years prior, lupus anticoagulant, and multiple family members with venous thromboembolism presented with shortness of breath. He was not on anticoagulation at presentation to the outside hospital. CTA revealed a large filling defect occluding the entire left main pulmonary artery with minimal extension of the clot across the right main pulmonary artery. He was also found to have a non-occlusive popliteal clot and pulmonary hypertension. Catheter-directed tissue plasminogen activator (tPA) was unsuccessful. He was diagnosed with CTEPH, and discharged on warfarin. One year later he was referred to our center complaining of dyspnea on minimal exertion, a nonproductive cough, and a recent episode of hemoptysis. The evaluation revealed pulmonary hypertension with a pulmonary vascular resistance (PVR) of 6.4 Wood unit (WU) and CT scan showed an enlarging mass in the left

**FIGURE 2** Case 1. Macroscopic examination (a) shows a mass-forming lesion. H&E (b) reveals prominent plasma cells and lymphohistiocytic infiltrate. The scattered spindle cells are positive for SMA (c), and negative for ALK (d). IgG highlights markedly increased plasma cells (e), with only scattered immunopositivity for IgG4 (f); (b–f) ×200 original magnification. H&E, hematoxylin and eosin; SMA, smooth muscle action
PA extending into and partially obstructing the right main PA, concerning for sarcoma. An extensive amount of soft tissue material was identified and resected from the main PA and extending into the right main PA. Grossly, the specimen consisted of multiple yellow-tan to white fragments with areas of hemorrhage. Histologically, almost every section showed a marked inflammatory infiltrate comprised mostly of plasma cells, small lymphocytes, and histiocytes in a background of collagenous stroma with scattered proliferating spindle cells (Figure 3b). ALK-1, Desmin, and S100 were completely negative. Early postoperative hemodynamics were improved (PVR: 2.1 WU), but surgery did not result in reperfusion of the left lung. The patient was known to be alive 2 years after surgery but then lost to follow-up.

Case 4

A 31-year-old male with no significant past medical history was diagnosed with saddle PE by CTA and pulmonary hypertension by echo after presenting to an outside hospital with acute onset shortness of breath and chest pain. Ultrasound was negative for DVT. He was discharged on warfarin, but intermittently compliant. He was admitted to the hospital 4 months later with worsening symptoms (WHO Functional Class IV), including presyncope. He was diagnosed with CTEPH and transferred to our institution. Coagulation studies were negative for a thrombophilia. A nuclear perfusion scan showed perfusion confined to the upper lobes. CT scan showed a large intravascular filling defect extending into the left and right pulmonary arteries with near complete luminal occlusion, raising suspicion for sarcoma. Preoperative hemodynamics revealed severe pulmonary hypertension with a PVR of 11.5 WU. The patient underwent emergent PEA for presumed CTEPH or possible sarcoma. Grossly, both left and right resected materials consisted of irregular tubular structures. Histologic examination demonstrated solid spindle cell overgrowth on a background of cellular organizing thrombi with recanalization (Figure 3c). The proliferating spindle cells were cytologically bland, devoid of increased mitotic activity or necrosis, and were positive for SMA but negative for Desmin and ALK-1 by immunostains. Surgery resulted in improvement in right ventricular function on echocardiogram, improved PVR (3.4 WU), and reperfusion of both lower lobes on nuclear scintigraphy. The patient died 7 years after surgery, the cause of death uncertain.

Case 5

A 47-year-old female with morbid obesity presented to an outside institution with a 1-year history of progressive

FIGURE 3 H&E morphology of Case 2 (a), 3 (b), 4 (c), and 5 (d) shows similar histology to that seen in Case 1, except more confluent spindle cells in Case 5 (d); ×200 original magnification. H&E, hematoxylin and eosin
dyspnea. A CTA was reported as showing right greater than left thromboembolic disease and she was treated with tPA, followed by anticoagulation without improvement in symptoms. She was referred to another center 3 months later and CT imaging showed a large thrombus in the right PA as well as a large uterine mass consistent with leiomyoma, but no evidence of mass effect on the pelvic veins. Thrombophilia testing was reported as negative, but ANA was positive at 1:640 (no other antibody testing reported). The patient was referred to our center for management of her CTEPH. Pulmonary angiography revealed presumptive bilateral CTED, with total obstruction of the right PA, disease in the left lower lobe, and pulmonary hypertension by right heart catheterization with a PVR of 4.4 WU. With PEA a significant amount of organized and partially organized material was removed from the proximal right pulmonary vascular bed and basilar segments of the left lower lobe. Grossly, the PEA specimen consisted of multiple pink-and-white-tan irregular soft tissue fragments comprising mostly organizing thrombus on histopathologic examination. A few areas of prominent spindle cell proliferation were identified, accompanied by a marked inflammatory cell infiltration (Figure 3d). The spindle cells were positive for SMA. CD3 and CD20 immunostains revealed mixed B- and T-lymphocytic population. Hemodynamics were improved postoperatively with PVR 2.9 WU and perfusion imaging showed improved perfusion to the left lower lobes, but no reperfusion of the right lung. The patient died 4 years after surgery of unknown cause.

**DISCUSSION**

We report a series of five cases of IPT involving PA, identified from patients who underwent PEA at a single institution. The initial clinical manifestations and imaging were felt to be compatible with acute PE. However, follow-up imaging studies after initial diagnosis of acute PE were concerning for chronic PE (CTEPH/CTED), or PA intimal sarcoma due to complete obstruction of a proximal main PA, apparent growth in size, and/or questionable invasion into the arterial wall in four of the five patients.\(^3\) Because of the highly lethal nature of PA intimal sarcoma, the distinction between the two entities is important and mainly relies on histopathological findings.\(^4\) Histologically, all five cases showed typical features of IPT, without significant cytologic atypia, increased mitoses, or necrosis. In addition, ALK-1 was negative in four of five cases. Of note, in a separate case series study performed at our institute, we identified 47 PA intimal sarcomas from over 2500 PEA specimens (1.9%, unpublished data), seemingly more prevalent than IPT (0.2%) but both nevertheless quite rare.

Besides sarcoma, other differential diagnosis for IPT complicating pulmonary thrombosis include vasculitis and IgG4-related disease (IgG4-RD). Systemic vasculitis involving large PA mimicking CTED and leading to PEA has been reported.\(^5\) Histologically, it often shows mural or transmural mixed inflammatory infiltrate, necrosis, and/or granulomas. The prominent plasma cell infiltrates and myofibroblastic proliferation seen in our cases is unusual for systemic vasculitides, and the absence of stainable microorganism on histology sections argues against an infectious process. IgG4-RD is an immune-mediated fibroinflammatory condition very rarely involves pulmonary arteries or secondarily obstructing the PA from compression.\(^6\) IgG4-RD features storiform fibrosis, obliterative phlebitis, and dense lymphoplasmacytic infiltrates with Increased IgG4+ plasma cells (usually >30%) by histology. In our case series, IgG4 immunostain was performed in two cases and both did not meet the diagnostic criteria for IgG4-RD.

Although IPT has been described in the literature for more than 40 years, the etiology is largely unknown. Infection or postinfectious sequelae may play a role in some cases. For example, in the lower urogenital tract, IPT could well represent a reparative process resembling nodular fasciitis.\(^7\) Orbital IPT has been linked to infection, autoimmune disorders, and aberrant wound healing.\(^8\) In CTEPH/CTED, it is reasonable to hypothesize that local thrombotic injury stimulates inflammatory infiltrates, which in turn recruits myofibroblastic proliferation from intimal mesenchymal stem cells in the arterial wall. Indeed, studies have shown that multipotent mesenchymal progenitor cells or myofibroblast-like cells are predominant within endarterectomized tissues from CTEPH patients, contributing to the vascular lesion/clot.\(^9,10\) Overgrowth of those myofibroblasts likely exacerbates the thrombosis, enhancing further inflammatory infiltrates, forming a vicious cycle that precipitates mass-forming lesions manifested as IPT. Exactly how this process is initiated and enhanced may warrant further studies.

The major limitation of our study is the absence of detailed longitudinal follow-up. This is a consequence of UCSD serving as a surgical referral center for the United States, but longitudinal care is provided by their local physicians. In addition, while most patients had initial clinical presentations compatible with acute PE, the imaging done at time of the diagnosis at outside clinical facilities was not available to us for review. It is possible that in some of these cases IPT arose de novo in the pulmonary artery. However, the acute onset of pulmonary symptoms and presence of DVT in some of the patients supports the theory of acute PE as the inciting event.
CONCLUSION

In summary, we describe five unique cases of IPT in PA identified in PEA specimens of patients with CTEPH. Four of the five patients had preoperative imaging concerning for PA intimal sarcoma. The pathogenesis is still unknown but may be associated with thromboembolic disease as an unusual fibroinflammatory reactive process.

ACKNOWLEDGMENT

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

CONFLICT OF INTERESTS

The authors declare that there are no conflict of interests.

ETHICS STATEMENT

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. This study was reviewed by the UC San Diego Human Research Protections Program, and a waiver of informed consent was granted for this project.

AUTHOR CONTRIBUTIONS

The authors confirm contribution to the paper as follows: Study conception and design: Xiaoyan Liao, Grace Lin. Data collection: Xiaoyan Liao, Christine M. Bojanowski, Andrew Yen, Kim M. Kerr, Justin Dumouchel, William R. Auger, Michael M. Madani, Victor Pretorius, Huan-You Wang, Eunhee S. Yi, Grace Y. Lin. Analysis and interpretation of results: Xiaoyan Liao. Draft manuscript preparation: Xiaoyan Liao, Kim M. Kerr, William R. Auger, Eunhee S. Yi, Grace Y. Lin. All authors reviewed the results and approved the final version of the manuscript.

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How to cite this article: Liao X, Bojanowski CM, Yen A, Kerr KM, Dumouchel J, Auger WR, Madani MM, Pretorius V, Wang H-Y, Yi ES, Lin GY. Inflammatory pseudotumor mimicking chronic pulmonary embolism or pulmonary artery sarcoma: report of five cases. Pulmonary Circulation. 2022;12: e12004. https://doi.org/10.1002/pul2.12004