



## Case report

## Pulmonary inflammatory pseudotumor causing lung collapse responding to corticosteroid therapy

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## A B S T R A C T

Pulmonary inflammatory pseudotumor (PIP) is a rare benign tumor that represents less than one percent of all tumors found in the lungs. Despite the benign etiology, PIP can cause significant clinical problems due to its growth rate and potential to compromise adjacent pulmonary and thoracic structures. Complete surgical resection is the preferred therapy for PIP to prevent recurrence, however, this is not possible in some patients due to the size or location of the tumor. We present the case of an 18 year-old male presenting with PIP in the proximal left mainstem bronchus causing complete left lung collapse. Surgical resection was not possible due to tumor location, and therefore the patient was treated with corticosteroids with marked response. Corticosteroid use has for PIP has been described in few other situations, and this case demonstrates the potential for this therapeutic option in patients with PIP who have a contraindication to surgical resection.

## 1. Introduction

Pulmonary inflammatory pseudotumor (PIP) is a benign tumor that can rarely be found in the lungs. Despite the benign etiology, PIP can cause significant clinical problems if it compromises adjacent pulmonary and thoracic structures. We present the case of an 18 year-old male presenting with PIP in the proximal left mainstem bronchus causing complete left lung collapse.

## 2. Case presentation

An 18 year-old male presented with 6 months of dyspnea on exertion that limited participation in competitive sports. He also noted concurrent non-productive cough but denied fevers, chills, or weight loss. He had no other significant medical history and denied tobacco, alcohol, or drug use. On physical examination, he was afebrile with stable vital signs. Auscultation of the lungs revealed decreased breath sounds in the entire left lung field. The remainder of the examination was normal.

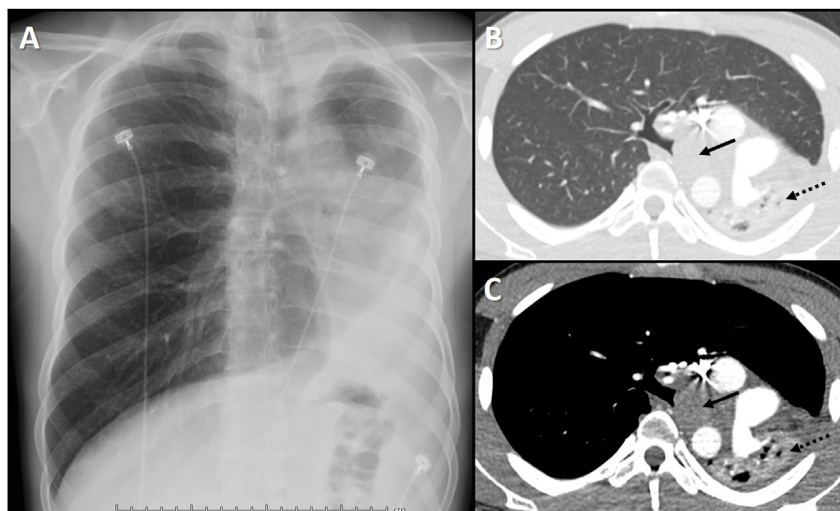
Chest radiograph showed left lung volume loss with compensatory hyperinflation of the right lung, and computed tomography (CT) showed a left mediastinal mass encasing the distal trachea and left

mainstem bronchus, with complete collapse of the left lung (Fig. 1). Pulmonary function tests (PFTs) showed severe obstruction with FEV<sub>1</sub> of 1.35 L (36% predicted), FVC of 3.59 L (83% predicted), FEV<sub>1</sub>/FVC of 36%, TLC of 5.62 L (92% predicted), and DLCO of 20 mL/min/mmHg (61% predicted). Bronchoscopy revealed near complete obstruction of the left mainstem bronchus by a mass with mixed endoluminal obstruction and extraluminal compression (Fig. 2). Biopsy of the mass revealed fibrotic tissue with a mixed chronic inflammatory cell infiltrate. Myofibroblasts stained positive for caldesmon, concerning for PIP. Additional specimens obtained by cervical mediastinoscopy and video-assisted thoracoscopic surgery revealed bronchial wall replacement by mixed inflammatory infiltrates with lymphocytes and spindle cells. Myofibroblast proliferation was confirmed with spindle cells staining positive for smooth muscle actin and caldesmon (Fig. 3). These findings were consistent with PIP involving the bronchial wall.

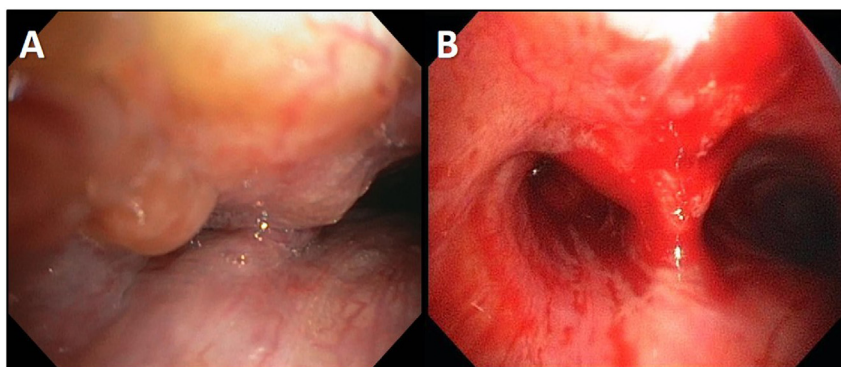
The patient was deemed to be a poor surgical candidate due to proximal tumor location involving the carina, which would prohibit resection with clean margins without compromising the patient's airway integrity and adjacent mediastinal structures. He was initiated on prednisone 60 mg daily for one month, and then tapered to 20 mg daily over the following 2 months. He had significant improvement in symptoms after 2 months of therapy with improvement in FEV<sub>1</sub> (3.09 L,

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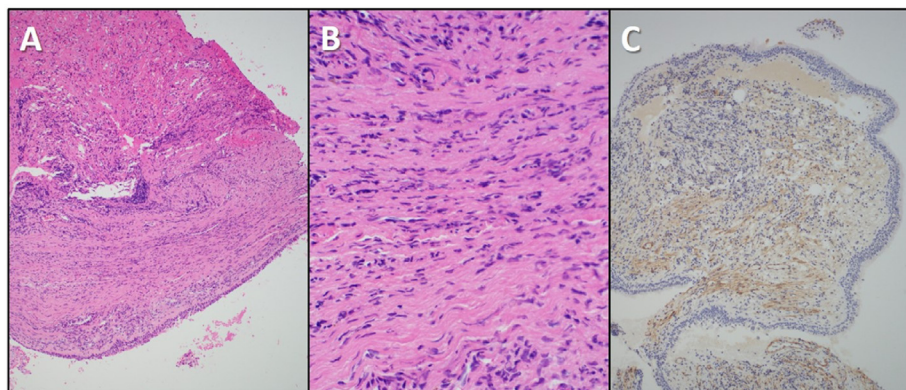
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**Fig. 1.** Chest X-ray (A) and CT (B and C) illustrating left lung volume loss with compensatory hyperinflation of the right lung. A left mediastinal mass encasing the distal trachea and left mainstem bronchus (solid arrows) results in complete collapse of the left lung (dashed arrows).



**Fig. 2.** Bronchoscopy Images. Panel A) Initial bronchoscopy at the distal trachea showing involvement of the trachea and near complete obstruction of the left mainstem bronchus by PIP with both endoluminal obstruction and extraluminal compression. Panel B) Repeat bronchoscopy after 3 months of steroid treatment showing restored patency of the left mainstem bronchus.



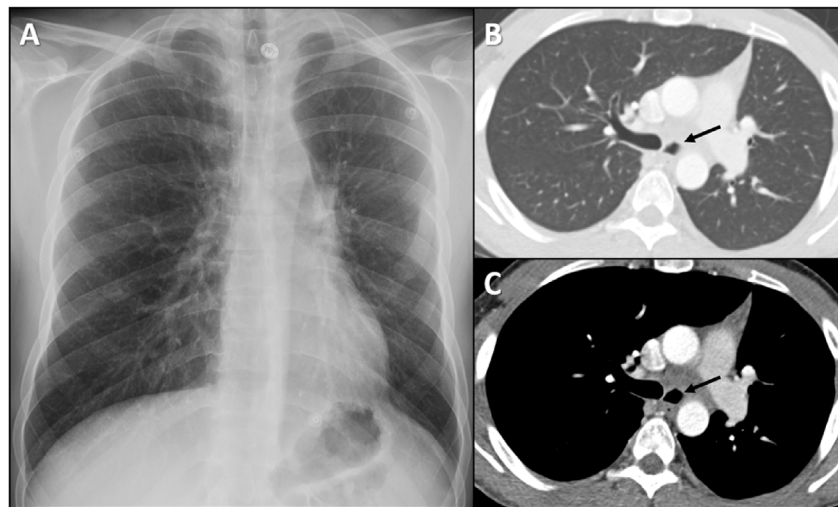
**Fig. 3.** Panel A) The biopsy demonstrates a polypoid fragment of soft tissue lined by respiratory epithelium (Hematoxylin and Eosin Stain, 10x magnification). Panel B) Higher power demonstrates bland spindle cells with chronic inflammatory infiltrates of predominantly lymphocytes and histiocytes (Hematoxylin and Eosin Stain, 40x magnification). Panel C) The spindle cells are positive for smooth muscle actin and negative for desmin.

74% predicted), FVC (4.74 L, 98% predicted), FEV<sub>1</sub>/FVC (69%), TLC (6.42 L, 98% predicted), and DLCO (24 mL/min/mmHg, 66% predicted). Subsequent chest radiograph and CT scan after 3 months of therapy showed significant decrease in size of the left mediastinal mass, a patent left mainstem bronchus, and complete re-expansion of the left lung (Fig. 4). Repeat bronchoscopy at that time also confirmed patency of the left mainstem bronchus (Fig. 2). As the patient regained baseline functional status, the prednisone dose was slowly tapered off with resolution of tumor confirmed by interval radiographic imaging. Total length of corticosteroid therapy was 14 months. At this time, approximately one year since steroids were discontinued, the patient remains asymptomatic.

### 3. Discussion

PIP is a rare benign tumor that represents less than one percent of all pulmonary tumors [1]. It may occur anywhere in the body, but is most commonly found in the lungs. Whether it is a true neoplasm or an inflammatory lesion with uncontrolled cell growth remains unclear. It is sometimes classified as a true neoplasm due to its tendency to recur and the presence of ALK gene rearrangement in a subset of cases [2].

Most patients diagnosed with PIP are under the age of 40. In a review of 28 cases of PIP diagnosed between 1977 and 2002 at a single institution in South Korea, Kim found the mean age of diagnosis to be 37.9 years [3]. In this case series, PIP was predominantly found in male patients (81.5%). Patients with PIP usually have non-specific symptoms



**Fig. 4.** Chest X-ray (A) and CT (B and C) after 3 months of steroid therapy demonstrating re-expansion of the left lung, decrease in size of the left mediastinal mass, patent left mainstem bronchus (arrow).

including cough, chest pain, fever, hemoptysis, and dyspnea. Laboratory studies are normal in most patients, but some may have anemia, thrombocytopenia, and an elevated sedimentation rate.

On imaging, PIP usually presents as a solitary, sharply circumscribed, peripheral mass greater than 3 centimeters in size with an anatomic bias for the lower lobes [4]. PIP masses generally do not cavitate nor do they have calcifications. In rare cases, PIP may present as an endobronchial lesion or invade the mediastinum or hilum. In a radiographic review of 61 cases, only 11 had extraparenchymal involvement including hilar, mediastinal, and airway invasion [4].

Pathologic evaluation demonstrates fibrous hyalinized bands interrupted by irregular patterned sheets of mononuclear cells including lymphocytes, histiocytes, and polyclonal plasma cells [5]. PIP is histologically classified into two categories, depending on its local invasiveness. Invasive PIP, as seen in this case, is usually diagnosed in younger patients, invades the chest wall or mediastinum, and is treated with lobectomy or pneumonectomy [1].

Complete surgical resection is the preferred therapy for PIP to prevent recurrence [6]. In a case review by Kim, open thoracotomy was performed in 23 out of 28 patients, with good response noted [3]. Melloni reported an association between completeness of resection and improved survival, which was independent of other clinicopathologic variables [7]. Corticosteroid therapy has been used with variable efficacy in patients who are inoperable, however the data is extremely limited in our review of the medical literature [2,3,8–12]. Steroids have been used in cases when surgical resection is not possible, such as when the tumor is too close to vital structures or if the patient is not a surgical candidate [2]. In two reported cases, prednisone 30 mg daily resulted in complete disappearance without recurrence of PIP masses after only 2–4 weeks of therapy [8]. Steroids have also been used in post-resection recurrence [9,12]. In one case report of PIP that recurred after surgery, steroid treatment with prednisolone 15 mg daily resulted in complete disappearance of the mass after 45 days of therapy [9]. In another case series by Shirakusa, three patients who failed surgical treatment were treated with steroids for 6–8 weeks, resulting in complete resolution with no recurrence after discontinuation of steroids [12]. Despite limited use in the medical literature, corticosteroid therapy has shown efficacy in treatment of inoperable PIP.

#### 4. Conclusion

In this case, an 18 year-old male presented with PIP in the proximal left mainstem bronchus causing complete left lung collapse. Surgical resection was not possible due to tumor location with involvement of

the main carina and proximity to mediastinal structures, which was prohibitive to obtaining clear resection margins. The patient was therefore treated with corticosteroid therapy, with a marked response. Steroid therapy has only been sparingly described in the literature. At this time, response rates to steroids and recurrence rates after steroid therapy are unknown, so it is not possible to advocate for steroid therapy as a first-line therapy. However, this case demonstrates the potential utility of steroids in patients with PIP who have a contraindication to surgical resection.

#### Conflicts of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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#### Appendix A. Supplementary data

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