

Pulmonary hypertension due to a pulmonary artery leiomyosarcoma: A case report

Hassan Adeli⁽¹⁾, Bardia Nemati⁽¹⁾, Mahboubeh Jandaghi⁽²⁾,
Mohammad Mahdi Riahi⁽³⁾, Fatemeh Salarvand⁽²⁾

Case Report

Abstract

BACKGROUND: Primary pulmonary artery sarcomas are very rare and their histologic type called leiomyosarcoma is even rarer. These tumors are frequently misdiagnosed as pulmonary thromboembolism in clinical settings. Many patients receive anticoagulant therapy without response, and many are diagnosed postmortem only. Most of the tumors reported in the literature have involved the right ventricular outflow tract and the main pulmonary trunk, often extending into the main pulmonary artery (MPA) branches.

CASE REPORT: A 64-year-old woman presented with weakness, fatigue, malaise, dyspnea, and marked elevation of pulmonary artery pressure was admitted to our hospital. She was initially diagnosed with chronic pulmonary thromboembolism, and chest computed tomography (CT) scan revealed lobulated heterogeneous left hilar mass extended to precarinal and subcarinal space. Magnetic resonance imaging (MRI) demonstrated a polypoid lesion at the trunk with extension to left MPA and its first branch. The patient was operated, and a yellowish-shiny solid mass in pulmonary trunk was seen intra-operation and pulmonary endarterectomy was performed. Her tumor was pathologically diagnosed as pulmonary artery leiomyosarcoma.

CONCLUSION: Clinicians must consider pulmonary artery sarcoma when making the differential diagnosis for patients with pulmonary artery masses. The clinical prediction scores and the CT and MRI findings can help identifying patients with pulmonary artery sarcoma.

Keywords: Hypertension, Leiomyosarcoma, Pulmonary Artery, Pulmonary Embolism

Date of submission: 23 Dec 2012, *Date of acceptance:* 22 May 2013

Introduction

Primary pulmonary artery sarcomas are very rare, and their histologic type called leiomyosarcoma is even rarer.¹ The incidence of primary pulmonary artery tumors is 0.001–0.03%, and are nearly always highly malignant and typically obtain their origin from the intima.² The underlying pathophysiology of these tumors of the pulmonary arteries is still unclear.^{2,3}

These tumors are frequently misdiagnosed as pulmonary thromboembolism in clinical settings. Many patients receive anticoagulant therapy without respond, and many are diagnosed postmortem only.⁴ Most of the tumors reported in the literature have involved the right ventricular outflow tract and the main pulmonary trunk, often extending into the main pulmonary artery (MPA) branches.⁵

Case Report

The patient was a 64-year-old woman presented with progressive generalized weakness and dyspnea for 6 months. She had no risk factors for thromboembolism. She had a history of headache, weight loss, and nonproductive cough prior to the admission and one episode of pre-syncope last year. Her vital signs at the time of admission were, temperature of 38°C, blood pressure of 120/70 mmHg, pulse of 80 beats/min and respiration rate of 24 breaths/min with O₂ saturation of 99%. On physical examination, no clinical evidence of deep vein thrombosis was found, and in cardiac auscultation, a systolic murmur (grade III/VI) was heard in lower left sternal border. Chest X-ray showed moderate enlargement of the right atrium and dilatation of right descending pulmonary artery (Palla's sign).⁵

1- Clinical Research Development Center, Qom University of Medical Sciences, Qom, Iran

2- General Practitioner, Clinical Research Development Center, Qom University of Medical Sciences, Qom, Iran

3- Qom University of Medical Sciences, Qom, Iran

Correspondence to: Fatemeh Salarvand, Email: salarmdfs@gmail.com

No abnormality was seen in her electrocardiography. Transthoracic echocardiography (ECG) showed a severe pulmonary artery hypertension (the predicted pulmonary artery pressure was 120 mmHg) with severe right ventricular hypertrophy, severe dysfunction, and moderate to severe tricuspid valve regurgitation. MPA was occupied by a large non-homogenous mass with very small flow from the left side of MPA and significant stenosis (peak gradient = 55 mmHg).

A contrast-enhanced computed tomography (CT) scanning of the chest showed lobulated heterogeneous left hilar mass occupying the precarinal and subcarinal space and invaded into left MPA and pulmonary trunk. Right ventricle and right atrium strain and enlargement, mild pericardial effusion and right sided pleural effusion were seen. Furthermore, magnetic resonance imaging (MRI) revealed a polypoid lesion at the trunk of pulmonary artery with extension to left main and the first branch (Figure 1).

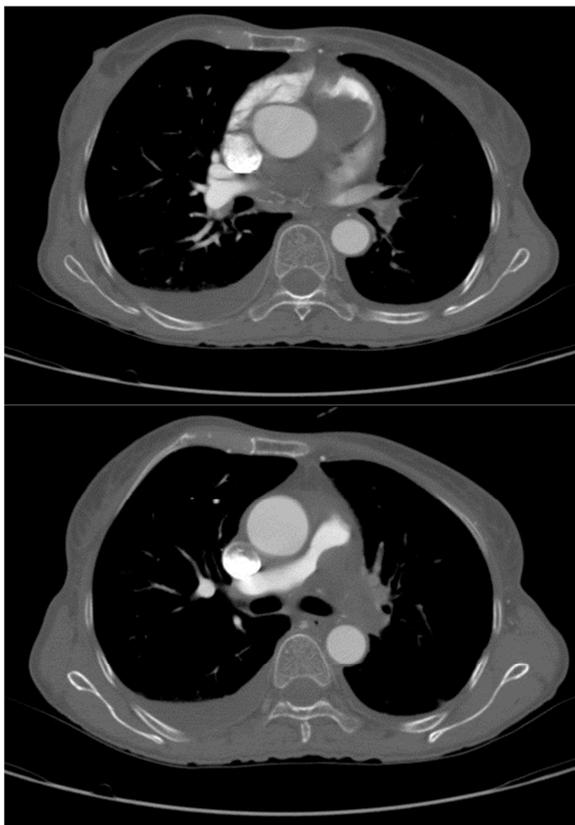


Figure 1. Magnetic resonance imaging reveals a polypoid lesion at the trunk of pulmonary artery with extension to left main and the first branch

No obvious extra-luminal extension was detected. With the primary diagnosis of the main pulmonary thromboembolism, the patient was

operated through mid-sternotomy. On opening the pulmonary artery, a soft yellowish-shiny mass was found. Gross examination showed creamy to gray color irregular tissue fragments with elastic consistency and hemorrhagic change totally measured $7 \times 6 \times 1.5$ cm.

Microscopically, the tumor was composed of spindle shaped cell with pleomorphism and ovaloid hyperchromatic to spindle nuclei was found. A pattern of tumor growth was observed in the lung parenchyma: tumoral cells arranged in parallel with whirling appearance forming storiform pattern.

Mitotic activity was evaluated as the number of mitotic figures per 10 high-power fields (HPF) was about 5–6 and bizarre cell and tumoral giant cells were present. Many cleft like vascular channel in addition to hemorrhagic myxoid and necrotic changes were present. Pathological diagnosis confirmed leiomyosarcoma confined to the excised pulmonary (Figures 2–4).

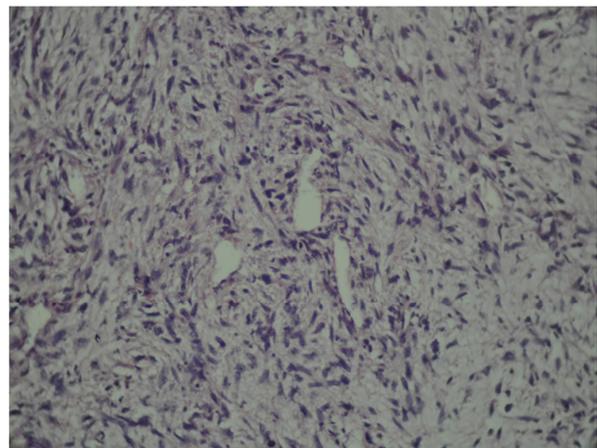


Figure 2. The intimate relationship of the tumor cells with the vessel walls is a clue to the diagnosis of leiomyosarcoma

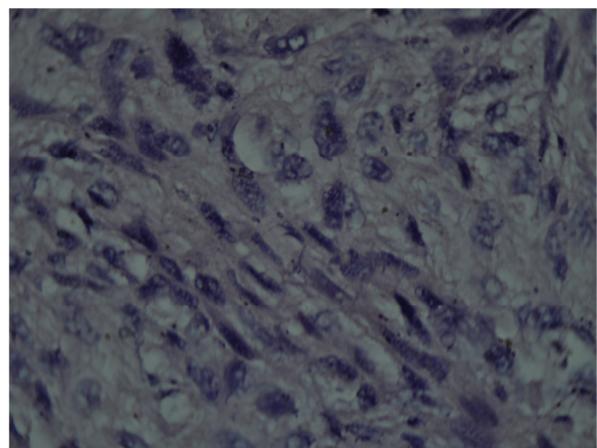


Figure 3. Cytologic features of leiomyosarcoma showing eosinophilic cytoplasm and blunt-ended nuclei

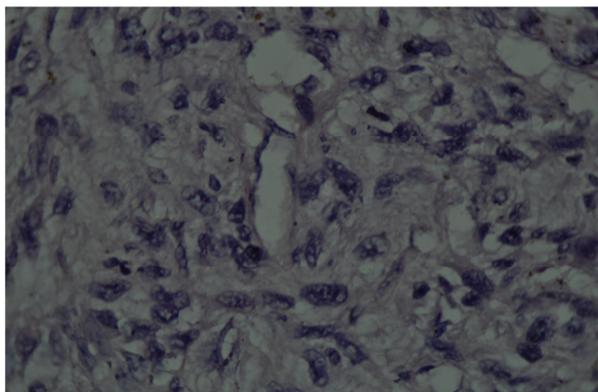


Figure 4. Cytologic features of leiomyosarcoma showing perinuclear vacuoles

Immunohistochemical staining was positive for smooth muscle actin, a marker for mesenchymal neoplasms. Immunohistochemical staining for factor 8, HMB45 and cytokeratin were negative while it was focally positive for CD68 and 5100. Tumor differentiation score was 2.3 with mitosis count of 13 per 10 HPF and score of 2.3.

Tumor necrosis: < 50% score: 1.2

And total score: 5.9. Grade: II.

The patient underwent pulmonary thromboendarthrectomy surgery with suspicion to pulmonary thromboemboly and a yellowish-shiny solid mass in pulmonary trunk was seen intra-operation and pulmonary endarthrectomy performed. Complete tumor resection was performed, and finally, the pathology result confirmed primary pulmonary artery sarcoma with smooth muscle differentiation compatible with leiomyosarcoma (i.e., intimal sarcoma).

Discussion

Primary leiomyosarcoma cases of the pulmonary artery are extremely rare, and most of them are initially misdiagnosed as pulmonary thromboembolism with symptoms of dyspnea, chest pain, cough, and hemoptysis.^{6,7} Both diseases are typically detected between the ages of 40 and 60, and women are involved twice as often as men. Physical examination, ECG and the chest X-ray may not reveal abnormal findings. However, cardiomegaly and radiological signs of a peripheral hypoperfusion can be present if a large tumor mass is obstructing a MPA vessel.⁸

Atypical features such as lack of predisposing factors for thromboembolism, persistence of symptoms or recurrence despite adequate anticoagulation, and unilateral distribution of a massive perfusion defect may evoke the diagnosis

of tumoral obstruction.⁹

The differential diagnoses include pulmonary artery sarcoma, thromboembolism, and lung cancer. The symptoms of the pulmonary embolism are nonspecific, and laboratory findings have a low diagnostic specificity and chest radiograph is generally nondiagnostic.⁴

The median survival time of the pulmonary artery leiomyosarcoma patients has been reported to be 1.5 months. It is believed that early and primary surgical resection is the best treatment of choice and can prolong the patient's life to 10–12 months.^{7,10}

The role of adjuvant therapy has not been yet clearly defined in the literature. The limited experience of any center in the treatment of these neoplasms makes it difficult to evaluate the relative importance of surgical excision and adjuvant therapy. Some investigators are in favor of adjuvant therapy and describe encouraging results.^{11,12}

Scores derived from explicit prediction rules that combine clinical findings at presentation with predisposing factors have proved useful in determining the clinical or pretest probability of pulmonary embolism. Three scores have been recommended as diagnostic criteria.⁴

Our patient experienced only a slight dyspnea before admission, and her physical examination on admission produced no findings suggestive of pulmonary embolism. It is suggested that pulmonary artery sarcomas should be strongly suspected in cases who present with mass lesions in the pulmonary arteries, but score low on the clinical prediction indexes. Several indicators on CT and MRI favor the diagnosis of pulmonary artery sarcoma over chronic thromboembolic disease.¹³ Chest X-ray showed moderate enlargement of the right atrium and dilatation of right descending pulmonary artery and MRI revealed a polypoid lesion at the trunk of pulmonary artery with extension to left main and the first branch.

Differential diagnosis also included primary and metastatic lung cancer. Our patient never smoked, and her serum tumor markers were all within normal ranges. It is said that the actual prevalence of pulmonary artery sarcoma is much higher than the estimated prevalence because of difficult diagnosis and often going unidentified without autopsy. The prognosis for patients with pulmonary artery sarcoma is poor, and in most cases, the mean survival time is < 2 years. Effect of chemotherapy or radiation therapy on prognosis is unclear and radical surgical resection seems to provide the only hope of long-term survival.¹⁴

Conclusion

Clinicians must consider pulmonary artery sarcoma when making the differential diagnosis for patients with pulmonary arteries masses. The clinical prediction scores and the CT and MRI findings can help identifying patients with pulmonary artery sarcoma.

Acknowledgments

The authors would like to thank Mrs. Fatemeh Hosseinzadeh for editing the manuscript.

Conflict of Interests

Authors have no conflict of interests.

References

1. Dumont P, Diot P, Aupart MR, Toumieux B. Leiomyosarcoma of the pulmonary artery. *Ann Thorac Surg* 1998; 66(6): 2089-91.
2. Bleisch VR, Kraus FT. Polypoid sarcoma of the pulmonary trunk: analysis of the literature and report of a case with leptomeric organelles and ultrastructural features of rhabdomyosarcoma. *Cancer* 1980; 46(2): 314-24.
3. Kramer TC. The partitioning of the truncus and conus and the formation of the membranous portion of the interventricular septum in the human heart. *American Journal of Anatomy* 1942; 71(3): 343-70.
4. Yamasaki M, Sumi Y, Sakakibara Y, Tamaoka M, Miyazaki Y, Arai H, et al. Pulmonary Artery Leiomyosarcoma Diagnosed without Delay. *Case Rep Oncol* 2011; 4(2): 287-98.
5. Delany SG, Doyle TC, Bunton RW, Hung NA, Joblin LU, Taylor DR. Pulmonary artery sarcoma mimicking pulmonary embolism. *Chest* 1993; 103(5): 1631-3.
6. Mayer E, Kriegsmann J, Gaumann A, Kauczor HU, Dahm M, Hake U, et al. Surgical treatment of pulmonary artery sarcoma. *J Thorac Cardiovasc Surg* 2001; 121(1): 77-82.
7. Kruger I, Borowski A, Horst M, de Vivie ER, Theissen P, Gross-Fengels W. Symptoms, diagnosis, and therapy of primary sarcomas of the pulmonary artery. *Thorac Cardiovasc Surg* 1990; 38(2): 91-5.
8. Moffat RE, Chang CH, Slaven JE. Roentgen considerations in primary pulmonary artery sarcoma. *Radiology* 1972; 104(2): 283-8.
9. Eng J, Murday AJ. Leiomyosarcoma of the pulmonary artery. *Ann Thorac Surg* 1992; 53(5): 905-6.
10. Mazzucco A, Luciani GB, Bertolini P, Faggian G, Morando G, Ghimenton C. Primary leiomyosarcoma of the pulmonary artery: diagnostic and surgical implications. *Ann Thorac Surg* 1994; 57(1): 222-5.
11. Head HD, Flam MS, John MJ, Lipnik SS, Slater DL, Stewart RD. Long-term palliation of pulmonary artery sarcoma by radical excision and adjuvant therapy. *Ann Thorac Surg* 1992; 53(2): 332-4.
12. Anderson MB, Kriett JM, Kapelanski DP, Tarazi R, Jamieson SW. Primary pulmonary artery sarcoma: a report of six cases. *Ann Thorac Surg* 1995; 59(6): 1487-90.
13. Meyer G, Roy PM, Gilberg S, Perrier A. Pulmonary embolism. *BMJ* 2010; 340: c1421.
14. Mattoo A, Fedullo PF, Kapelanski D, Ilowite JS. Pulmonary artery sarcoma: a case report of surgical cure and 5-year follow-up. *Chest* 2002; 122(2): 745-7.

How to cite this article: Adeli H, Nemati B, Jandaghi M, Riahi MM, Salarvand F. **Pulmonary hypertension due to a pulmonary artery leiomyosarcoma: A case report.** *ARYA Atheroscler* 2014; 10(2): 133-6.