

Pulmonary Artery Sarcoma—A Challenging Diagnosis: A Case Report

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ABSTRACT

Pulmonary artery sarcoma is a known but rare tumor with poor prognosis. It is a challenging diagnostic dilemma, as the main complaint of these patients is progressive dyspnea. Dyspnea is a common symptom in cardiorespiratory disease and therefore a high index of suspicion is necessary to make the diagnosis early and proceed with surgery and radiotherapy and/or chemotherapy. We report a case of primary pulmonary artery sarcoma that presented initially with weight loss, shortness of breath, palpitations, and dysphagia.

CASE REPORT

A 62-year-old woman presented with fatigue, dysphagia, weight loss, chest pain, palpitations, and progressive shortness of breath gradually reaching New York Heart Association functional class IV. Past medical history included macrocytic anemia (thalassemia trait), microprolactenemia, and bilateral cataracts. Her medications included bisacodyl, omeprazole, clonidine, codeine, paracetamol, and hormone replacement therapy.

Initially the main symptom of the patient was dysphagia and thus all the symptoms of the patient were attributed to the dysphagia as well as anemia. As a result, the medical team focused on the investigation of this symptom (they performed swallowed-barium tests and manometry of the esophagus) and underestimated the mild systolic murmur of the pulmonic valve, which was misdiagnosed as mild systolic murmur of the aortic valve. Despite the medications she was on for achalasia of the esophagus, all the symptoms remained, and only after a year was she referred to the cardiologists. Echocardiography at that time revealed a mass obstructing the right ventricular outflow tract. The differential diagnosis included pulmonary emboli, chronic thromboembolic disease, and pulmonary artery tumor.

A computed tomography (CT) scan (Figure 1), magnetic resonance image (MRI), and coronary angiogram were per-

formed to assist preoperative planning. Modified EuroSCORE predicted a mortality rate of 6%. The patient underwent exploratory median sternotomy under cardiopulmonary bypass. The surgical finding was an extensive tumor involving the main pulmonary artery, origin of the right pulmonary artery, and complete invasion of the entire left pulmonary artery (Figure 2) extending down into the left lobar branches. The involved pulmonary arteries had a very thick fibrotic component requiring endarterectomy as well as a more central sarcomatous component. The tumor was resected with an extensive tumor endarterectomy (Figures 2 and 3); however, as a fibrotic area in the right ventricle outflow was left, this was considered to be a palliative procedure.

She made an uneventful postoperative recovery and was referred for chemotherapy and radiotherapy. Now, 6 months later, she is asymptomatic and there is no evidence of recurrence of the tumor on the CT scan.

Histological examination revealed a pleomorphic pulmonary artery sarcoma (Figure 4) composed of highly atypical medium-sized epithelioid to spindle-shaped cells. Tumor giant cells and atypical mitoses were seen (25 mitoses/10 HPFs). Numerous hemangiopericytoma-like vessels were present. The tumor cells were positive for the

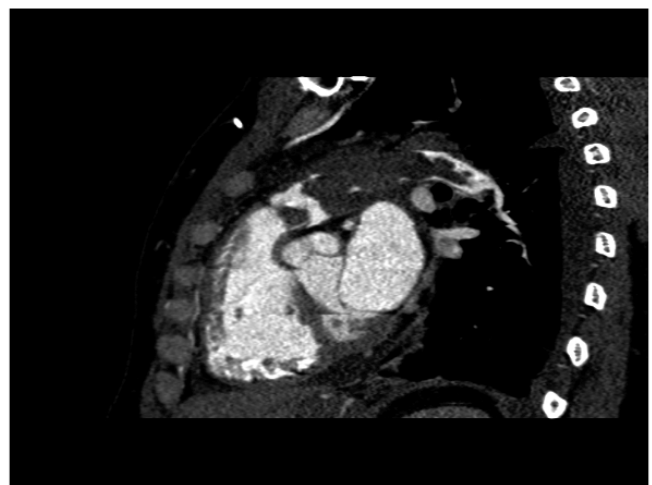


Figure 1. Chest computed tomography scan with intravenous contrast demonstrating a filling defect in the right ventricular outflow tract.

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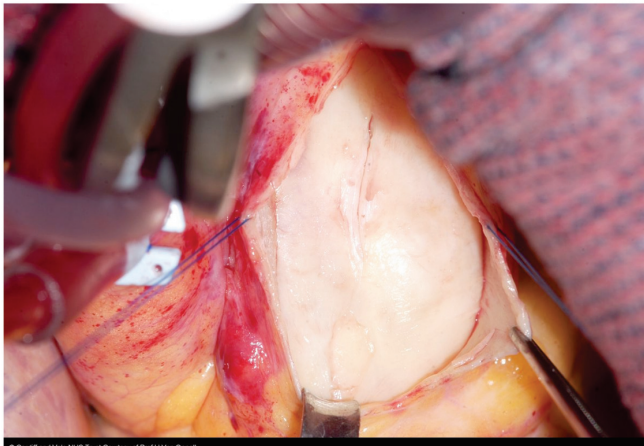


Figure 2. Exploration of the pulmonary artery revealing the pulmonary artery sarcoma.

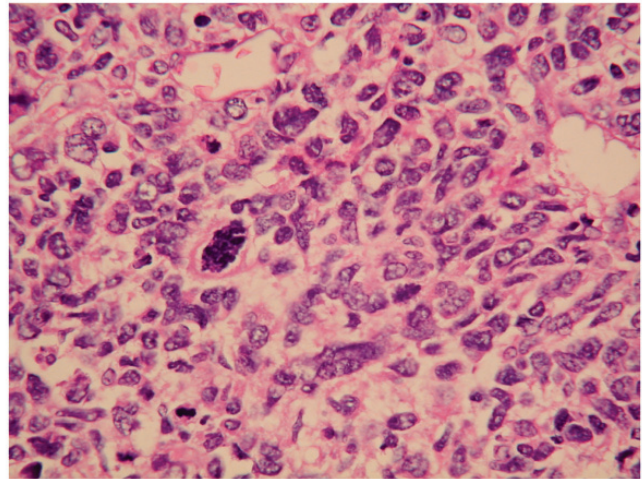


Figure 4. Histological section of the pulmonary artery sarcoma.

immunocytochemical marker vimentin but were negative for CD34 and CD31 (vascular markers), cytokeratin (epithelial marker), smooth muscle actin and desmin (smooth muscle markers), and CD99 and S100.

DISCUSSION

Mandglstamm [1923] described the first case of a pulmonary artery sarcoma and since then no more than 210 cases have been reported [Akomea-Agyin 1996; Minakata 2000; Hoffmeier 2001]. Pulmonary artery sarcomas arise from pluripotent intimal cells. They are most frequently located within the pulmonary trunk spreading into the proximal and distal pulmonary arteries. Typical onset age is 45 to 55 with a reported age range of 13 to 86 years [Farooki 1988; Pagni 1999]; the female to male ratio is 2:1. Presenting symptoms are progressive dyspnea, chest pain,

cough, intermittent hemoptysis, palpitation, clubbing, and fatigue, mimicking chronic pulmonary thromboembolic disease [Shmookler 1977; Hopwood 1979]. The differential diagnosis includes tumors of the lung and mediastinum, pulmonary embolus, pulmonary arteritis, congenital pulmonary stenosis, pulmonary infections, and fibrosing mediastinitis. Because of the rarity of these kinds of tumors they are often initially misdiagnosed as chronic thromboembolic disease, leading to unnecessary anticoagulation in these patients. Echocardiography (transthoracic and transoesophageal), CT, MRI, and right-heart catheterization are the most commonly used diagnostic tools. Preoperative histological diagnosis is generally not possible, although rarely biopsy may be obtained via CT-guided transthoracic aspiration [Parish 1996], alternatively via transvenous catheter suction biopsy with pulmonary angioscopy [Mattoo 2002], or by transbronchial biopsy [Daimon 2006]. Endarterectomy, via sternotomy and cardiopulmonary bypass, with dissection of the intima and most parts of the media, may result in complete tumor removal and is the most commonly used and widely accepted surgical strategy. Although in a unilateral disease it is possible to attempt a pneumonectomy [Mayer 2002], in a bilateral disease (extension of the tumor to the right and left pulmonary arteries) it is of limited value. Alternative surgical methods include total excision of the pulmonary trunk and reconstruction [Tobe 2004] or heart and lung transplantation for unresectable tumors [Talbot 2002]. Palliative treatment includes debulking of the tumor, endovascular stent [Totaro 2004], or bypass procedure [Calderon 1996]. The prognosis is poor. Without surgery survival is 1.5 months. Mean survival with surgical resection and chemotherapy and/or radiotherapy was previously reported to be 1 year [Kruger 1990], but reports indicate that the mean survival has increased to 3.5 years [Parish 1996]. The survival varies and in one case report the patient was alive 49 months after heart and lung transplantation [Talbot 2002], in another the patient was alive 62 months after successful resection of the tumor [Mayer 2002], in a third

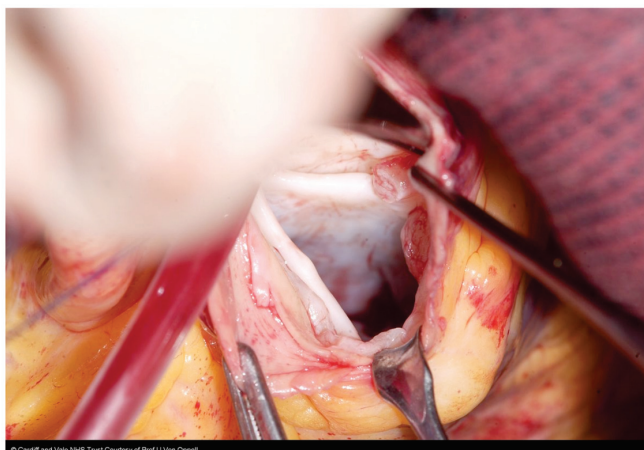


Figure 3. Pulmonary artery after excision of the pulmonary artery sarcoma.

report the survival was 60 months [Mattoo 2002], and in another it was 68 months [Huo 2006]. The maximum reported survival is 10 years [Baker 1985; Head 1992; Hoffmeier 2001]. Radical surgical resection remains the cornerstone of management offering definite diagnosis, improvement of symptoms, and the best outcome; however, if curative resection is not possible, palliative resection still provides good symptom relief and improved survival.

In our case, the diversity of the presenting symptoms led to the underestimation of the possibility of malignancy as the underlying cause, which resulted in a delay in diagnosis and treatment.

A high index of suspicion for early diagnosis is the key to successful treatment and prolonged survival, especially in patients with atypical presentations from the cardiopulmonary systems, associated with weight loss, anemia, shortness of breath, and, in our case, dysphagia.

REFERENCES

- Akomea-Agyin C, Dussek JE, Anderson DR, Hartley RB. 1996. Pulmonary artery sarcoma mimicking pulmonary embolism: successful surgical intervention. *Ann Thorac Surg* 61:1536-8.
- Baker PB, Goodwin RA. 1985. Pulmonary artery sarcomas. A review and report of a case. *Arch Pathol Lab Med* 109:35-9.
- Calderon M, Galvan J, Negri V, Verdin R. 1996. Right ventricular bypass for palliation of cardiac sarcoma. *Tex Heart Inst J* 23:178-9.
- Daimon T, Tajima S, Bando M, Ohno S, Hironaka M, Sugiyama Y. 2006. A case of primary fibrosarcoma of the pulmonary artery. *Nihon Kokyuki Gakkai Zasshi* 44:319-24.
- Farooki ZQ, Chang CH, Jackson WL, et al. 1988. Primary pulmonary artery sarcoma in two children. *Pediatr Cardiol* 9:243-51.
- Head HD, Flam MS, John MJ, Lipnik SS, Slater DL, Stewart RD. 1992. Long-term palliation of pulmonary artery sarcoma by radical excision and adjuvant therapy. *Ann Thorac Surg* 53:332-4.
- Hoffmeier A, Semik M, Fallenbergh EM, Scheld HH. 2001. Leiomyosarcoma of the pulmonary artery—a diagnostic chameleon. *Eur J Cardiothorac Surg* 20:1049-51.
- Hopwood D, McNeill G. 1979. Spindle cell sarcoma of the pulmonary trunk: a case report with histochemistry and electron microscopy. *J Pathol* 128:70-7.
- Huo L, Moran CA, Fuller GN, Gladish G, Suster S. 2006. Pulmonary artery sarcoma: a clinicopathologic and immunohistochemical study of 12 cases. *Am J Clin Pathol* 125:419-24.
- Kruger I, Borowski A, Horst M, de Vivie ER, Theissen P, Gross-Fengels W. 1990. Symptoms, diagnosis, and therapy of primary sarcomas of the pulmonary artery. *Thorac Cardiovasc Surg* 38:91-5.
- Mandelstamm M. 1923. *Über primäre Neubildungen des Herzens*. *Virchows Arch Pathol Anat* 245:43-54.
- Mattoo A, Fedullo PF, Kapelanski D, Ilowite JS. 2002. Pulmonary artery sarcoma: a case report of surgical cure and 5-year follow-up. *Chest* 122:745-7.
- Mayer E, Kriegsmann J, Gaumann A, et al. 2001. Surgical treatment of pulmonary artery sarcoma. *J Thorac Cardiovasc Surg* 121:77-82.
- Minakata K, Konishi Y, Matsumoto M, Aota M, Nonaka M, Yamada N. 2000. Primary leiomyosarcoma of the pulmonary artery mimicking massive pulmonary thromboembolism. *Jpn Circ J* 64:783-4.
- Pagni S, Passik CS, Riordan CH, et al. 1999. Sarcoma of the main pulmonary artery: an unusual etiology of recurrent pulmonary emboli. *J Cardiovasc Surg (Torino)* 40:457-61.
- Parish JM, Rosenow EC 3rd, Swensen SJ, Crotty TB. 1996. Pulmonary artery sarcoma. Clinical features. *Chest* 110:1480-8.
- Shmookler BM, Marsh HB, Roberts WC. 1977. Primary sarcoma of the pulmonary trunk and/or right or left main pulmonary artery—a rare cause of obstruction to right ventricular outflow. Report on two patients and analysis of 35 previously described patients. *Am J Med* 63:263-72.
- Talbot SM, Taub RN, Keohan ML, Edwards N, Galantowicz ME, Schulman LL. 2002. Combined heart and lung transplantation for unresectable primary cardiac sarcoma. *J Thorac Cardiovasc Surg* 124:1145-8.
- Tobe S, Nohara H, Yoshida K, Tanimura N, Kurokawa R, Kawata M. 2004. Right ventricular outflow tract reconstruction for primary pulmonary artery sarcoma. *Jpn J Thorac Cardiovasc Surg* 52:139-42.
- Totaro M, Miraldi F, Ghiribelli C, Biscosi C. 2004. Cardiac angiosarcoma arising from pulmonary artery: endovascular treatment. *Ann Thorac Surg* 78:1468-70.