Author's response to reviews

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Pulmonary Artery Intimal Sarcoma: A Case Report

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Abstract
Pulmonary artery (PA) intimal sarcoma is a rare disease without any characteristic symptoms. It is difficult to earlier diagnose without any characteristic symptoms, and frequently was misdiagnosed as always mistaken for pulmonary thromboembolism. While surgery is the mainstay treatment for it, the prognosis is unfavorable. Here we report the case of a 54-year-old woman diagnosed with PA intimal sarcoma which was made during surgery or after autopsy. The defining feature of such sarcoma is local growth with slight ability to metastasize [1]. Herein, a case of PA intimal sarcoma, which with extensive metastases 10 months after surgery, which had extensive metastases 10 months after surgery is given and was presented and discussed.

Introduction
Intimal sarcoma of PA is a very rare tumor with poor prognosis. It was always mistaken for pulmonary thromboembolism, and in most cases the definite diagnosis was made during surgery or after autopsy. The defining feature of such sarcoma is local growth with slight ability to metastasize [1]. Herein, a case of PA intimal sarcoma, which with extensive metastases 10 months after surgery, which had extensive metastases 10 months after surgery is given and was presented and discussed.

Case Presentation
The patient was a 54-year-old female who presented with complaints in other hospital with of shortness of breath on exertion in past three months' duration. At physical examination found that slight jugular vein distention (JVD) engorgement was observed; auscultation at the left second intercostal space detected a grade III to VI systolic murmur which was heard at indicative of tricuspid area regurgitation; and no abnormalities were detected in abdomen—mild edema of both leg was found. Laboratory findings workups were within normal limits showed no abnormalities. Echocardiography showed that revealed the following results: the main PA did not widen, right PA (RPA) was almost completely obstructed by a low echogenic mass, left PA (LPA) was also partially obstructed and that by a low echogenic mass. Superior vena cava (SVC) was dilated, with the diameter of about 23mm. Right ventricle enlargement and tricuspid insufficiency were also revealed. Helical computed tomography (CT) scan showed that the RPA trunk was blocked by soft tissue mass which grew circumferentially, with only line-like contrast passing through it (Fig 1). LPA was thickened interorly, with favorable contrast perfusion to its branches. On the basis of these findings, the patient was diagnosed with PA thromboembolism and scheduled for surgery.
During operation, under general anesthesia, a local soft mass about 1.5×1.8×4.0 cm was completely resected from RPA. The diagnosis of intimal sarcoma was confirmed by pathological examination (Fig 2). Atypical spindle cells were observed. Immunohistochemical analysis showed vimentin (VIM) positive, CD34 weakly positive partially, while CD117, S-100, smooth-muscle actin (SMA), desmin (DES) and CD68 negative (figures not provided). After operation, the symptoms were relieved, and echocardiography showed normal blood flow in main PA trunk, RPA and LPA as well as their proximal branches. Then, the patient was discharged.

Ten months later, the patient was referred to our department with complaints of shortness of breath on exertion, restlessness during activity, recurring facial swelling, and pain in the right upper right quadrant. The patient was referred to our department. On physical examination no abnormalities were found except for swelling face and engorged jugular vein. CT scan showed a solid mass in right pulmonary hilum, without clear boundary to mediastinum. The mass could be enhanced asymmetrically, with low-density necrosis in the centre area invading SVC and RPA trunk (Fig 3). In addition, a low-density cycloid mass with clear boundary was detected on quadrate lobe of liver. Based on these findings, diagnosis was made as local recurrence of intimal sarcoma of PA, with suspected metastasis to the liver. 3-dimensional conformal radiation therapy (3D-CRT) to mediastinum was carried out. Total dose was 60Gy using 15MV x ray in 30 fractions. Dyspnea and facial swelling were relieved.

Thirteen days after radiotherapy, the patient complained progressive pain in the right upper right quadrant appeared again, with loss of appetite, anorexia, nausea and vomiting. Enlarged metastatic mass in liver was detected showed by CT. The treatment measure was transcatheter arterial chemo-embolization (TACE) was carried on for mass in liver using fluridine 1.0 gram, Epirubicin 40 mg and Mitomycin 6 mg. After treatment, the symptoms condition were relieved improved, and the patient was discharged again. Two months later, CT scan showed extensive metastases to lung, liver, and both adrenals. Then, the patient died 2 weeks later.

Discussion

Intimal sarcoma is a malignant mesenchymal tumor that arises from large vessels including aorta and PA. These tumors are characteristic of intraluminal growth with obstruction of the tract and seeding of emboli. Intimal sarcoma is a rare tumor, the incidence of PA intimal sarcoma is almost twice that of aortic origin. It mainly occurs in adults, with age ranging from 13 to 86 years, with female predominance, and the ratio of female to male is 1.3:1, whereas there is no gender variation in the incidence of aorta intimal sarcoma. The mean age of patients diagnosed with PA intimal sarcoma is 48 years while with aorta intimal sarcoma is 62 years.

PA intimal sarcoma arises from the intimal layer of right, left, and main PA. In rare cases, it extends retrograde to pulmonary valve and right ventricle. Approximately 40% patients develop a direct invasion or metastasis to lung, while systematic spread to kidney, brain, adrenal glands occurs in about 20% cases.

The patients with intimal sarcoma present in various presents in various ways without specific
symptoms. The most common initial symptoms are dyspnea, hemoptysis, and chest pain. Because of its rarity and insidious growth features, the diagnosis of sarcoma is always delayed or made at surgery or autopsy. PA intimal sarcoma is often mistaken for pulmonary thromboembolism as it was so in our patient.

Surgery offers the best way to prolong survival and is successful only if complete resection of tumor is performed. Postoperative chemotherapy has been reported effective in some cases, but its role in the treatment of PA intimal sarcoma is still not clearly defined. Radiation therapy and the duration of postoperative anticoagulation therapy have also not been demonstrated indispensably. The prognosis of PA intimal sarcoma is poor, and the survival is usually 12 to 18 months.

In the present case, the patient had gained a 10 months disease-free survival after surgery, even though the surgery was just a local mass resection, which confirms the positive role of surgery in the management of PA intimal sarcoma. When tumor relapsed in pulmonary hilum, local 3-D CRT to mediastinum had successfully controlled symptoms such as dyspnea and facial swelling for a while. This information suggests that intimal sarcoma of PA may be sensitive to radiation in some cases. In addition, TACE also relieved the liver pain for 2 months. Unfortunately, systematic metastases developed finally, and best supportive care was provided. The total survival time of this patient is about 15 months after the onset of symptoms.

**Conclusion**

In conclusion, PA intimal sarcoma is a rare and highly lethal tumor. While surgery can prolong the survival of patients, some other treatment adjuvant therapies such as chemotherapy and radiotherapy may also contribute to the management of this disease. Thus, these management should also be recommended. In addition, besides, systematic evaluation of metastasis should also be recommended as such tumor has the capacity to metastasize.

**Abbreviations**

PA=Pulmonary artery; RPA=right Pulmonary artery; LPA=left Pulmonary artery; SVC= Superior vena cava; CT= computed tomography; CD=cluster of differentiation; 3D-CRT=3-dimensional conformal radiation therapy; TACE=transcatheter arterial chemo-embdization.

**Consent**

Written informed consent was obtained from the relatives of patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.
Competing interests
The author(s) declare that they have no competing interests.

Authors' contributions
HQL collected the data, QQ drafted the manuscript, CHX revised and approved the final manuscript.

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References

Figure legends
Figure 1: A computed tomography scan of the chest shows the main PA was obstructed circumferentially.

Figure 2: Pathologic finding of surgical specimen shows abundant spindle cells (HE stain × 100).

Figure 3: A computed tomography scan of the chest shows the recurrence of PA intimal sarcoma.