SARCOMA OF THE PULMONARY TRUNK AND THE MAIN PULMONARY ARTERIES

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Introduction

Pulmonary-artery sarcoma is a rare vascular tumor with a poor prognosis. The tumors are often found incidentally or when the patients are highly symptomatic. Surgical measures offer the only chance for cure or symptomatic improvement, even in advanced tumor stages. We report a case of a sarcoma with infiltration of the pulmonary valve and obstruction of the pulmonary trunk and both main pulmonary arteries. Additional intravasal filling defects were found in both lungs. As the patient was quite young and highly symptomatic with recurrent syncopes, resection and prosthetic replacement was yet scheduled. Six months later a centrally located tumor necessitated a left sided pneumonectomy. The operation was solely indicated, because the peripheral filling defects had completely disappeared. Despite the major operative procedures the patient is doing well and has until now gained one year with a good life quality.

Clinical summary

A 63-year-old man was evaluated for worsening dyspnea and recurrent syncopes. Laboratory evaluations including plasma D-dimer level, ECG, and chest roentgenogram were normal. Transthoracic echocardiography revealed signs of right ventricular pressure overload. An intracavitary tumor or emboli in transit could not be detected. Venous duplex examination of the legs was negative for deep vein thrombosis.

Computed tomographic scan disclosed massive filling defects in the pulmonary trunk and an almost complete occlusion of the right and left pulmonary artery (Fig 1). The tops of solid intravascular masses were smooth and lobular and jutted into both extrapericardial pulmonary arteries. Additional endovascular filling defects could be seen peripherally in a left basal segmental pulmonary artery, in the anterior and posterior branches of the left upper lobe, and in the right basal artery. These filling defects measured 3 to 7 mm in diameter.

Right heart catheterization and pulmonary angiography confirmed these findings. The right ventricular systolic pressure was 95 mmHg, the systolic pressure in the distal pulmonary trunk was 60 mmHg, in the right pulmonary artery centrally to the level of the truncus anterior branch 40 mmHg, in the interlobar portion of the right pulmonary artery 15 mmHg, and in the left pulmonary artery at the level of the first branch to the anterior segment of the upper lobe 20 mmHg. Left heart catheterization and coronary angiography were normal.

Due to the morphological appearence of the lesions and the chronicity of symptoms despite the significant pressure gradients an endovascular tumor was suspected. A biopsy with myocardial biopsy forceps failed due to the firm consistence of the foreign tissue.

Thus operation was scheduled. After midline sternotomy the ascending aortic and the caval veins were cannulated. The superior caval vein was cannulated peripherally with a right angled cannula (Stöckert Instruments, München, Germany). Prior to installation of the cardiopulmonary bypass the pleurae were opened and the extrapericardial route of the main pulmonary arteries were dissected free. After crossclamping of the aorta and cardioplegic arrest, the pulmonary trunk was longitudinally opened, and biopsies were taken. Fresh frozen section yielded a myxoma.

The tumor was more likely to infiltrate the vessel wall than adhere to it. In addition, one leaflet of the pulmonary valve was infiltrated by the tumor. The pulmonary arteries were extrapericardially transsected. The tongue shaped top of the tumor protruding into the interlobar portion of the right pulmonary artery could be removed. The cut edges of the vessels were macroscopically free of tumor tissue. After transsection of the ascending aorta and the superior caval vein the pulmonary arteries and the pulmonary trunk including the pulmonary valve were in toto resected.

In a first step a 20 mm wide externally supported PTFE (polytetrafluoroethylene) vascular prosthesis (W. L. Gore & Associates, Inc., Flagstaff, AZ) was extrapericardially anastomosed end-to-end to the right and the left pulmonary artery. During intraluminal exploration of the pulmonary arteries and for the anastomoses two episodes of deep hypothermic circulatory arrest with an overall time span of 18 minutes were needed.

In a second step a 27 mm Shelhigh porcine valved conduit (Shelhigh, Inc. Union, NJ) was implanted as substitute for the pulmonary valve and trunk. The conduit was anastomosed end-to-side to the mid portion of the PTFE vascular prosthesis (Fig 2).

The transected ascending aorta and superior caval vein were re-anastomosed and the aortic cross-clamp was removed. Weaning from the cardiopulmonary bypass was without difficulties. The further operative and postoperative course was uneventful.

Histology: The intraoperative frozen section examination indicated a myxoma. The study of the paraffin-embedded specimens by three independently working pathologists yielded the diagnosis of a myxoid and regressively transformed thrombus. Only an expert for soft tissue malignancies made the diagnosis of primary sarcoma of vessel wall origin.

A first control-CT scan one month after surgery showed free vessels exept the known, but unchanged peripheral filling defects in both lungs. The patient received anicoagulation with phenprocumon. Another CT-control was made 6 months after surgery and revealed a left hilar tumor deriving from the interlobar portion of the pulmonary artery (Fig 3). The patient was again operated a via leftsided thoracotomy. After clamping the left branch of the pulmonary artery prothesis and both lung veins, the pulmonary artery and the interlobar portion of the artery were incised. An intra-arterially tumor destructing the vessel wall and reaching distally into the segment arteries of both lobes was found. As reconstructive measures were not possible pneumonectomy was performed. The postoperative course was again uneventful. CT-control 6 months after pneumonectomy (i.e. one year after central pulmonary artery resection) did not exhibit evidence of tumor recurrence.

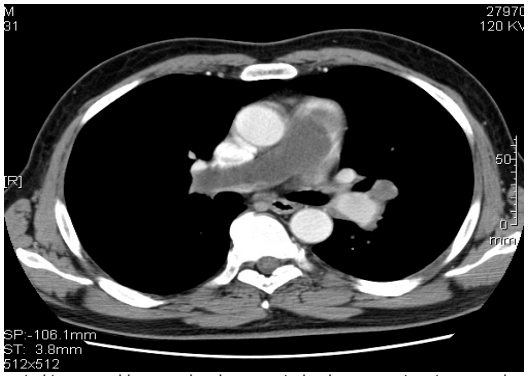

Fig 1. Computed tomographic scan showing a central pulmonary artery tumor and an additional filling defect in left upper lobe branches.

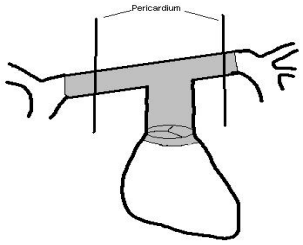

Fig 2. Operative schema. Right ventricle and central pulmonary arteries. The pulmonary trunk including the pulmonary valve as well as both main pulmonary arteries are resected. Reconstruction is done by implantation of a porcine valved conduit and an externally supported PTFE vascular prosthesis.

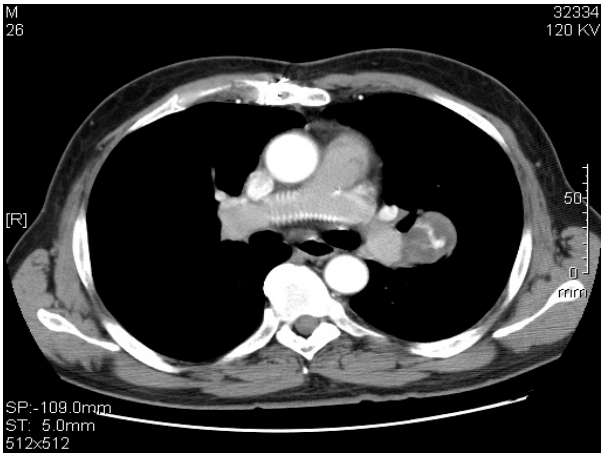

Fig 3. Computed tomographic scan six months after operation showing a left hilar tumor arising from the interlobar portion of the pulmonary artery.

Comment

Primary tumors of the pulmonary arteries are rare and usually represent sarcomatous neoplasms. The pathological features of pulmonary artery sarcoma have been repeatedly reviewed, but as histologic subclassification is of no clinical or prognostical use, the term "primary sarcoma of the pulmonary artery" is most appropriate.

The natural course of the disease is determined by local tumor growth and additional thrombosis both leading to progressive obstruction of the vessels. Due to slow progress it may cause few symptoms but sudden death will occur when near total occlusion by the tumor and/or an additional thrombus leads to acute right ventricular failure. This is underlined by the fact, that 60 % of the reported cases were identified at autopsy.

If characteristic symptoms of pulmonary artery obstruction develop, endovascular tumors are often mistaken for pulmonary embolism, leading to inappropriate diagnostic and therapeutic measures. Endovascular biopsy may be helpful. If concomitant thrombosis exists in the proximity of a tumor, biopsy can also result in the false diagnosis of pulmonary thromboembolism. Furthermore, sarcomas of vessel wall origin represent a wide spectrum of microscopic appearence and particularly hypocellular tumors can be misdiagnosed, as it was the case in our patient.

If an obstructing endovascular pulmonary tumor is diagnosed, a mean survival time of only 1.5 months was calculated. Surgery is the only potentially curative treatment modality independent of the histology. The operative strategy concerning endovascular pulmonary artery tumors differs essentially from the operative strategy in pulmonary artery obstruction by thromboembolism. While thromboendarterectomy is the adequate method to surgically treat chronic obstruction by thromboembolic disease, aggressive resection of tumor bearing arteries whenever possible represents the treatment of choice in endovascular pulmonary artery tumors. Complete resection will result in a median survival time of 19 months and sometimes even in long term survival.

Not all intravasal filling defects located distally to a central intravasal tumor are necessarily malignant. In our patient the small and peripherally located filling defects in the right segmental arteries had disappeared six months after the first operation. Based on this fact we indicated the second operation in the hope, minor pulmonary resection combined with arterioplastic procedures would be possible, but we ended in a leftsided pneumonectomy. This underlines the necessity of repeated CT-controls, to recognize local tumor recurrence earlier. Pneumonectomy in the case of potentially curative resection of a central intra-arterial pulmonary tumor in combination with unilateral peripheral filling defects is described but its role is not defined. The value of adjuvant chemotherapy or radiotherapy also remains unclear.

In conclusion, we believe that even extensive resectional procedures of the pulmonary trunk, the pulmonary valve, and both pulmonary arteries including secondary pulmonary resections are justified as far as the remaining pulmonary function warrants an acceptable lifestyle as it is the case in our patient. The remaining lifespan without surgical intervention is extremely short.

In the case of central pulmonary artery tumor additional peripheral filling defects may represent embolized tumor or thrombus. Frequent CT scan controls with the patient under oral anticoagulation are appropriate to discriminate re-operation.