LETTER TO THE EDITOR

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Surgical treatment of primary pulmonary sarcoma disseminated to both pulmonary arteries using composite pulmonary trunk and bifurcation of abdominal aorta allograft

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Primary pulmonary artery sarcoma (PPAS) is a rare tumor originating from the intimal mesenchymal cells of the pulmonary artery with an incidence of 0.001–0.03% and is usually fatal [1]. The diagnosis is difficult and delayed in most cases. It usually presents in adults (almost equally in males and females), with median age of presentation of 50 years [2]. Newer imaging techniques could allow early diagnosis in patients with symptoms of pulmonary vascular obstruction [3]. Surgical resection improves clinical symptoms, offers the only chance of cure and is performed in many cardiothoracic clinics. The PPAS spreading to lobular branches of pulmonary artery (PA) represents an especially serious surgical challenge.

A 58-year-old female patient was referred to our unit in December 2016 complaining of dyspnea on moderate physical exertion and unproductive cough starting 2 months prior to admission. Transthoracic echocardiography revealed a mass formation of 62/24 mm in the pulmonary trunk, fused with the valvular leaflets. Third-degree tricuspid valve regurgitation was recorded as well. Aortic and mitral valves showed no significant change, with left ventricular ejection fraction (LVEF) of 62%, and right ventricular ejection fraction (RVEF) of 42%. Laboratory findings revealed no significant changes. Computed tomography angiography (CTA) showed a negative dynamic in comparison to CT data of a month earlier: the tumor spread from the bifurcation of the pulmonary trunk by 5 mm to the left branch of the pulmonary artery, to the bifurcation of the right branch of the PA and up to 10 mm to the valvular space of the right ventricular outflow tract (Fig. 1 A). Tumor emboli in the segmental branches of the PA were also seen. In the lung parenchyma, foci appeared to have increased in size: 4 mm in S6 on the right (+3 mm), 6 mm in S3 on the left (+1–2 mm), 7 mm in S6 on the left (newly formed). Positron emission tomography (PET) confirmed the presence of metabolic-active formation in the pulmonary truck spreading to both PA and secondary changes in S6 of the right and in S3 and S6 of the left lung. In this case changes in the lungs were assumed as secondary to primary sarcoma of the PA.

Surgery was performed on December 28, 2016 through median sternotomy. Before institution of cardiopulmonary bypass (CPB) and following a revision of the lungs, an atypical resection of the foci in S6 of the right lung and in S3 and S6 of the left lung was performed. Upon opening the pericardium, about 150 ml of fluids was drained with subsequent sudden disturbance in hemodynamics with systolic pressure dropping to less than 50 mm Hg, followed by an increase in the central venous pressure and asystole. Direct cardiac massage was performed and CPB was started. Under a total operative time of 380 min (myocardial ischemia time of 181 min) and hypothermia of 26°C, thromboendarterectomy from both PA branches with complete replacement of the pulmonary trunk by pulmonary trunk allograft (PTA, ring diameter 28 mm) was performed. Annuloplasty repair of the tricuspid valve by De Vega was also performed. Due to insufficient length of the right branch of the PTA, around 85 mm in length of bifurcation of the abdominal aorta allograft (BAAA, 20 mm in diameter) was anastomosed to the lobular branch of the PTA. To fit the diameter of one iliac branch of the BAAA, the right upper and lower lobular PA branches were sutured together for a suitable anastomosis, while the other iliac branch of the BAAA was ligated (Figs. 1 B, C).

Perioperative blood replacement therapy – 5 doses of erythrocyte mass, 5 doses of 10% albumin solution, 3 doses of Octaplas (a pooled human plasma solution, Octapharma Pharmazeutika Produktionsges m.b.H., Vienna, Austria). The patient was extubated after 13 h but remained in the intensive care unit for 5 days due to moderate respiratory failure. Postoperative chest X-ray showed subsegmental atelectases of both lobes on both sides, as a consequence

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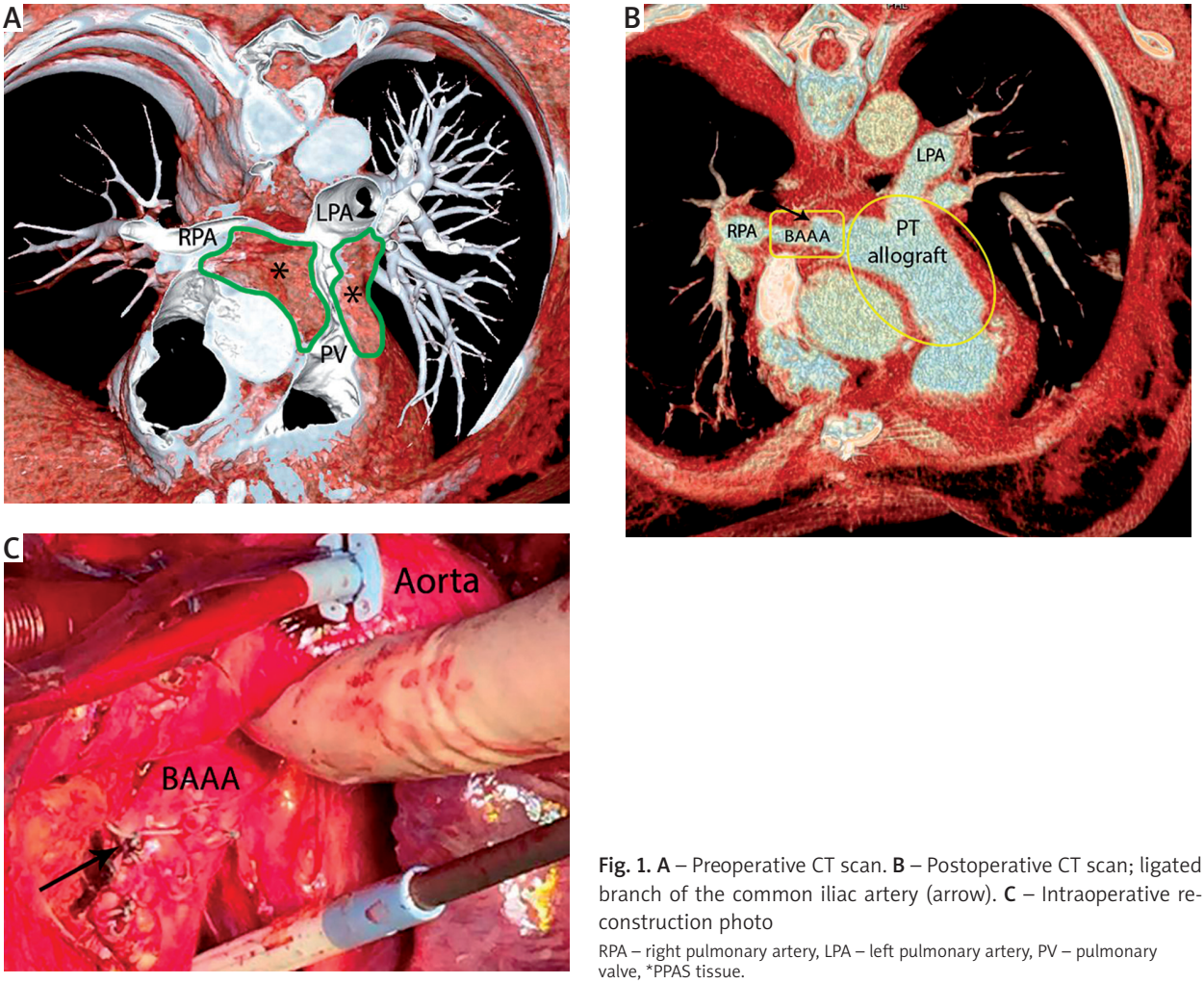

Fig. 1. A – Preoperative CT scan. **B** – Postoperative CT scan; ligated branch of the common iliac artery (arrow). **C** – Intraoperative reconstruction photo

RPA – right pulmonary artery, LPA – left pulmonary artery, PV – pulmonary valve, *PPAS tissue.

of thromboembolism of small branches of the PA in combination with inflammatory infiltration. Postoperative echocardiography (day 13): no detectable tumor mass in the PA, aortic, tricuspid and mitral valve – trivial regurgitation, LVEF 62%, PVEF 46%, maximum gradient across PA valve 3 mm Hg with mild regurgitation, average PA pressure 36 mm Hg. The patient received long-term antibacterial, anticoagulant therapy and was discharged on the 20th day after the operation in a satisfactory condition.

Pathology study confirmed the presence of PPAS, intimal type, with myofibroblastic differentiation, G3. Tumor growth in the margins of resection, and metastatic lesion of the right and left lungs were revealed (Fig. 2). Immunohistochemical study: SMA ++, Des (–), Vim +++, CD31 –/+, CD34 –/+, FVW –/+, Ki67 – 25%.

Postoperatively the patient initially received 8 courses of adjuvant polychemotherapy (docetaxel 120 mg and gemcitabine 100 mg) from February, 2017 to October, 2017 with a negative effect according to CT data (enlargement of lung foci from 5 mm to 22 mm). Subsequently, the patient was switched to another polychemotherapy (mesna 400 mg and ifosfamide 1.2 g/m^2) and received 6 courses, starting from October, 2017, with positive radiological results (decrease in lung foci from 9 × 14 mm to 6.3 × 9 mm in S5, from 5 mm to 3.5 mm in S9–S10, from 7 mm to 4 mm in S6 of the right lung).

Echocardiography 18 months postoperatively was not significantly different from that of the early postoperative period. The patient had a CT control on November, 2018, roughly 23 months after the operation, with further reduction in lung foci size.

The PPAS is an uncommon malignant tumor of the cardiovascular system. Moritz Mandelstamm first described this disease in 1923 [1]. Since then, around 400 confirmed PPAS cases were reported with a slight predilection for men with the majority of cases occurring in patients around 50–55 years old [2]. Symptoms vary depending on the tumor histology, size and location. Common symptoms include dyspnea, cough, chest pain, malaise, and anorexia. Occluded circulation induced abnormal coagulation, arrhythmias, and pericardial effusion tamponade. Signs indicative of neoplasia, such as weight loss, clubbing, anemia, and an elevation in the erythrocyte sedimentation rate, may occasionally be found. Systolic murmur of the PA valve area origin and signs of dysfunctional right ventricle can be frequently observed during physical examination. Usually, a chest radiograph can detect hilar enlargement and the echocardiography can locate the mass in the ves-

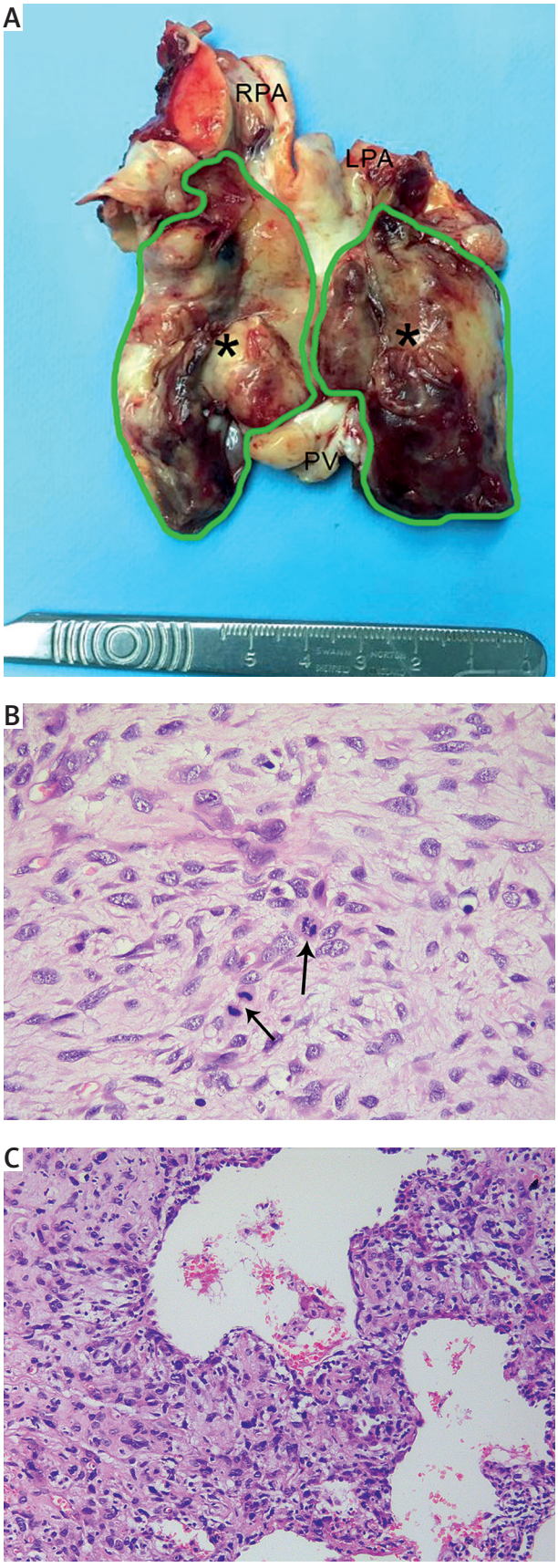

Fig. 2. A – Gross image of explanted tumor. **B** – Mitotic activity in the tumor (arrows), H + E, 400×. **C** – Tumor metastasis in the lung, H + E, 100×

sels. In clinical practice PPAS is extremely rare, whereas the prevalence of pulmonary embolism is fairly high. Due to similar symptoms and echocardiographic findings, PPAS is often confused with pulmonary embolism [2, 4]. The diagnosis of PPAS is often made during the operation when the tumor mass was thought to be chronic pulmonary emboli originally. Chest CT scan and cardiac MRI are helpful anatomically while PET scan has been proven to be more potent to differentiate these two diseases and to characterize the features of tumor mass in the PA. Due to limited effects of chemotherapy or radiotherapy, surgical resection has been proven to be the most beneficial alternative for the treatment of PPAS [4, 5].

Fures *et al.* stated that the effects of radiotherapy and chemotherapy after surgical treatment of PPAS are not clearly defined [6]. Mattoo *et al.* considered that if the disease was diagnosed early before the occurrence of distal metastasis or involvement of adjacent mediastinal structures, the intimal sarcoma could be cured by total resection [7].

In our case, the patient received 14 courses of polychemotherapy and there is no evidence of tumor recurrence in the pulmonary artery, along with a decrease in size of metastatic foci in the lungs.

The uniqueness of this case is the modification of composite allograft (PTA and BAAA) implantation following complete resection of the pulmonary trunk with both branches of the PA and thrombectomy due to severe intraoperative thromboembolism. Such a composite conduit is especially useful in the case of PA major branches' length deficiency and also if lobular branches are affected by PPAS. If lobular branches require a prosthesis, BAAA is a readily available biological bifurcation prosthesis.

We chose the BAAA over xenograft, xenopericardium patch, Dacron and PTFE prostheses as the plastic nature of the allograft was considered to reduce the risk of infection, hemorrhage and thrombosis.

In this case, resection of pulmonary metastases before CPB institution was performed to reduce the risk of bleeding into the pulmonary parenchyma following full heparinization.

We chose to ligate one branch of the BAAA instead of suturing it due to low pressure in the PA and to reduce myocardial ischemia time under CPB.

Overgrowth of PPAS into both branches of the PA up to the lobular branches cannot be considered as a contraindication to surgical treatment. In such cases multimodal therapy, comprising PA and lobular branches' replacement with a composite allograft (PTA and BAAA) in conjunction with adjuvant polychemotherapy can be considered as the treatment of choice. Composite allograft implantation, described in this paper, involves replacement of the PA with both its branches and is associated with low risk of bleeding and thromboembolic and infectious complications.

Disclosure

The authors report no conflict of interest.

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