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Case Presentation

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Incidental Finding of Rare Mediastinal Neoplasm: Case Report

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Abstract

A case report of a 68-year-old patient who was examined for a suspected aggressive sarcoma in the posterior mediastinum, which was accidentally detected during orthopedic preoperative examination. Due to the need for biopsy, the patient was operated on at the cardiac surgery department, where the entire tumor was subsequently enucleated. Histological examination confirmed a left atrial paraganglioma with low proliferative activity.

Keywords: Paraganglioma; Mediastinal neoplasm; Enucleation; Left atrial tumor; Cardiac surgery

Introduction

A case report of a 68-year-old patient who was investigated for a suspected aggressive sarcoma in the posterior mediastinum, incidentally detected during preoperative examination. He was presented to our department for the need to biopsy an expansive tumor. During the operation we were able to resecate the whole neoplasm, and final histology described a paraganglioma. Paragangliomas are rare tumors arising from neural crest tissue that develops into sympathetic and parasympathetic paraganglia throughout the body [1]. The incidence ranges from 2-8 per million people per year. They are present in 0.1% to 1% of patients with hypertension and are present in up to 5% of patients in whom adrenal masses are present [1-3]. The average age at diagnosis is 24.9 years in hereditary cases and 43.9 years in sporadic cases [1,4]. Cardiac tumors are classified into primary benign or malignant tumors that arise from the heart or into secondary metastatic tumors that invade the heart. Secondary tumors are 20 times more common than primary cardiac tumors. Around 70% are benign tumors, most commonly myxomas, which are around 50-60% of the heart's primary benign tumors.

Pheochromocytomas/paraganglioma arise from chromaffin cells of the sympathetic nervous system and produce excess amounts of catecholamines, particularly norepinephrine. Approximately 90% of pheochromocytomas are in adrenal glands. Fewer than 2% arise in the chest. There are only tens of reported cases of heart paragangliomas. The tumor predominantly affects young and middle-aged adults. Approximately 60% occur in the roof of the left atrium [5]. The prognosis for patients with a benign tumor that is removed is good.

Case Presentation

In October 2023, a 68-year-old patient with a highly metabolically active, centrally necrotizing expansion in the posterior mediastinum was presented to our Cardiac Surgery Department. This was an incidental finding of an expansion lesion in the posterior mediastinum, which was found during a preoperative examination, before a planned hip TEP. The patient underwent PET/CT with verification of subcarinal expansion between the left atrium and the lung, measuring 104x75x63 mm. Left ventricular compression, dilatation of all cardiac compartments, pericardial effusion up to 12 mm. Furthermore, a minor left-right interatrial shunt was described. Nodes in the mediastinum and pulmonary hila and abdomen with low metabolic activity, multiple foci in the spleen were described, but they were suspected of being hamartomas. The thyroid gland was enlarged. The neck and supraclavicular joints without pathological enlargement of the nodes. Furthermore, an MRI of the brain was performed to exclude metastatic spread.

From laboratory tests, lung oncomarkers were determined, which were within the norm, only elevated chromogranin A to 143.3 ng/ml. Fibrinogen elevated to 5.72.AFP, bhCG, CA 19-9 levels were within the norm. CRP and procalcitonin within the norm.The patient underwent bronchoscopy for an attempt at node puncture, with cytology. This was performed without detecting malignant cells. Transthoracic echo was supplemented, which excluded significant pericardial effusion, but with described dilatation of the cardiac compartments. The patient was on warfarin for persistent atrial fibrillation and was being treated for arterial hypertension, hypercholesterolemia, hypothyroidism, and coxarthrosis, for which TEP was indicated. According to the previous examinations, the differential diagnosis primarily considered sarcoma, angiosarcoma and paraganglioma. To determine the further course of action and treatment, tissue sampling for histological examination was necessary. Due to the difficult anatomical location, our department was asked to perform a surgical approach and biopsy. The patient was fully informed about the risks associated with the procedure and agreed to the surgical procedure.

Before the operation, there were episodes of hypertension, the pressure did not exceed 160/100 mmHg. The surgery was also preceded by episodes of shortness of breath. The patient was admitted to our department in October 2023, the operation was scheduled on the next day of the hospitalization. The operation was originally intended to take a biopsy from the tumor. Under calm general anesthesia in the right lateral position, an anterolateral thoracotomy was performed. The pericardial sac was opened, where under pressure there was a leak of serosanguinous fluid in a larger volume, palpable bulky spherical tumor under the left atrial appendage. Due to the new finding, the patient was placed in a supine position, and a full sternotomy was performed. Cannulation of the ascending aorta and vena cava **(Figure 1)**. A clamp was applied and cardiac arrest was performed. The

left atrium was opened, where there was an arching tumor under the ceiling of the left atrium and the left wall. Subsequently, an incision was made in the ceiling of the left atrium, a bulky tumor of semi-solid consistency was enucleated with a finger in its entire extent en-block **(Figure 2).** The tumor size was 10x8x6 cm, it was sent for biopsy **(Figure 1)**. Re-suturing of the atrial wall from the excess of the original tissue, suture of the atrium. Subsequently, the patient was disconnected from the extracorporeal circulation and the bleeding was stopped. Drainage around the heart and into the left pleural cavity. Cerclage of the sternum. Suture in layers.

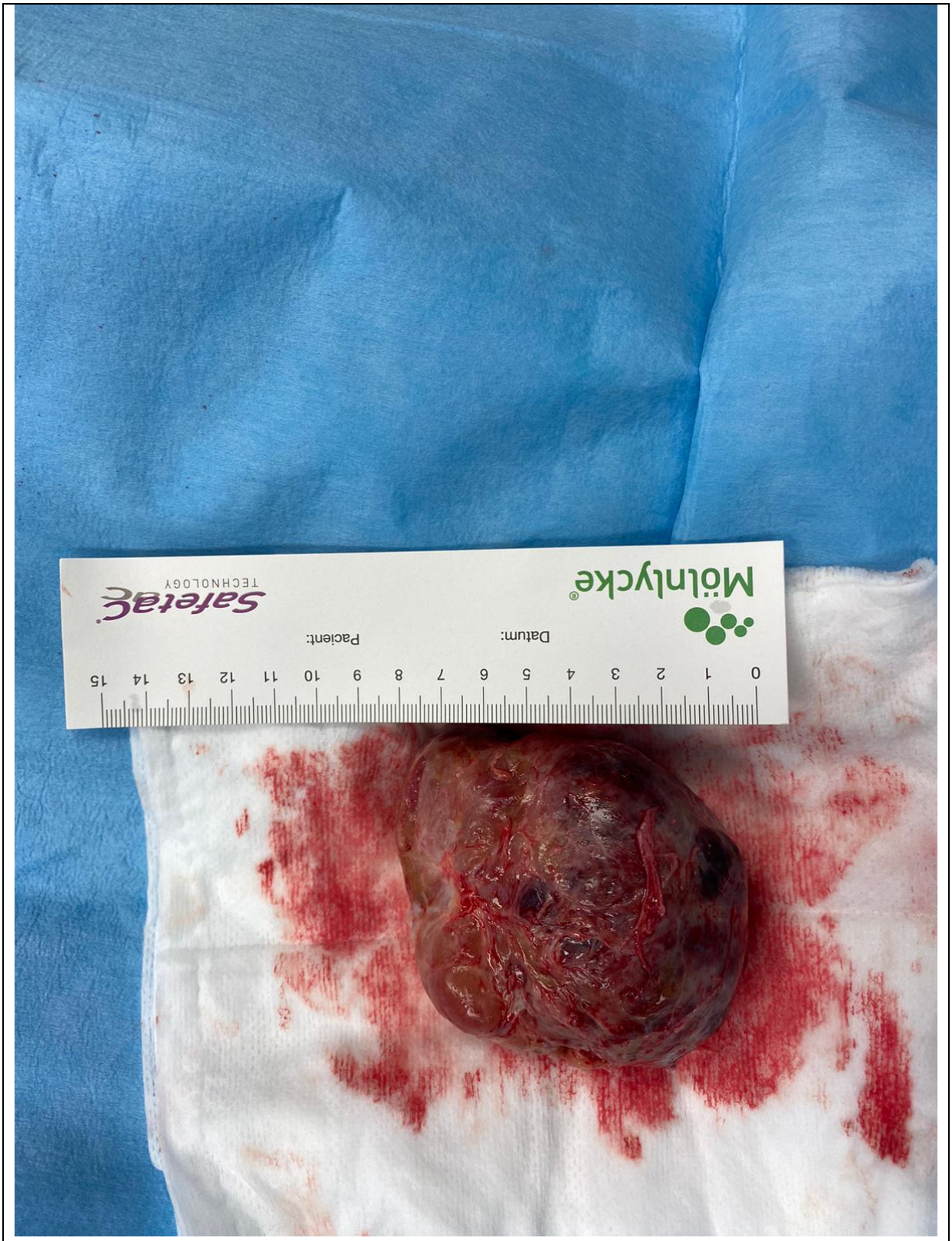

Figure 1: Enucleated Neoplasm.

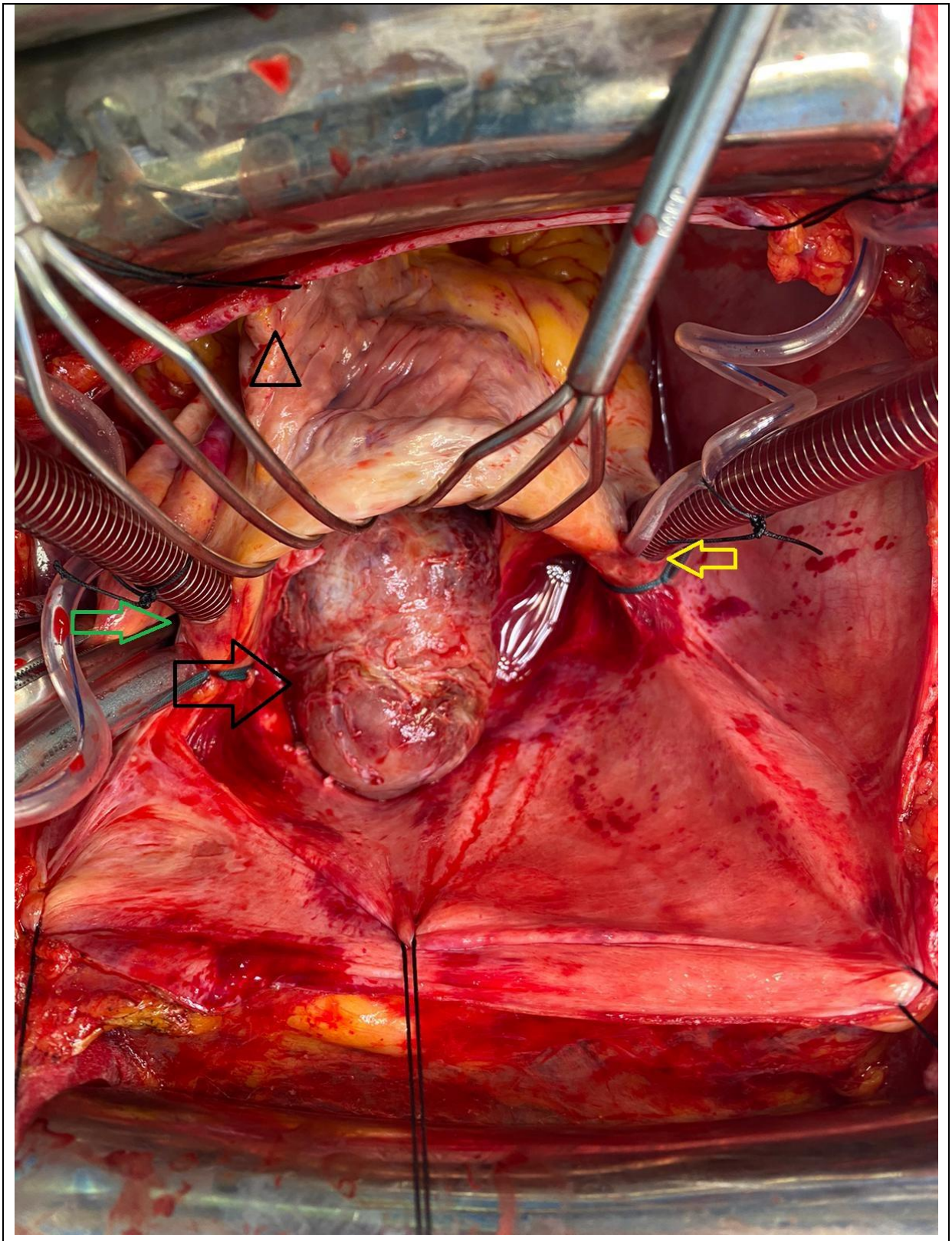

Figure 2: Neoplasm of the right atrium. Black pointer – Neoplasm, Yellow pointer - V. Cava inferior, Green pointer - V. Cava superior, Triangle - auricula of right atrium

Postoperative period without major complications. There were no more episodes of hypertension or shortness of breath. Postoperative Transthoracic Echocardiography (TTE) with description of minor

prolapse of the anterior leaflet of the mitral valve, dilatation of the left atrium. Multiple dilatated vascular structures around the left atrium and the left atrial appendage. Persistent small left-right shunt. According to the biopsy examination, the tumor reached the resection margin marked with ink. Immunohistochemically, synaptophysin, chromogranin with GATA3 antibody, S-100 protein and SOX 10, as well as SDHB protein were present. Histologically, a paraganglioma was detected, which was arranged in typical nest-like structures, which contained tumor cells with mild to moderate atypia.

According to the oncology council, the patient is after R1 resection of a rare tumor, for which there are no unified guidelines. According to the available data, despite the R1 procedure, the patient's prognosis is good. Adjuvant oncological treatment or resection is not indicated. According to CTAG, with a six-month interval without evidence of expansion.

Discussion

Pheochromocytomas and extraadrenal paragangliomas are rare tumors arising from neural crest tissue that develops into sympathetic and parasympathetic ganglia throughout the body [1]. In 2004, the WHO classification used the term pheochromocytoma exclusively for tumors arising from the adrenal medulla, and the term extraadrenal paraganglioma for similar tumors arising from other locations [1]. The incidence ranges from 2-8 per million people per year. They are present in 0.1% to 1% of patients with hypertension and are present in up to 5% of patients in whom adrenal masses are present [1-3]. The most common occurrence is in the 3rd to 5th decade of life. The average age at diagnosis is 24.9 years in hereditary cases and 43.9 years in sporadic cases [1,4]. The most common manifestations include symptoms associated with increased production of catecholamines, hypertension, headaches, increased sweating, palpitations, tremor, and facial pallor [1,5]. Diagnosis is associated with incidental findings and increased adrenal mass. Laboratory tests, in addition to increased catecholamine uptake, can determine metanephrines [5]. The prognosis for patients with a benign tumor that is removed is good. Recurrence rates range from 6.5 to 16.5%, according to available data [1,5]. According to the recommendation, tumor resection should be followed by repeated CT or MRI screening at intervals to monitor for possible recurrences [6,7].

The case described above, due to its accidental detection, fits into the mosaic of previously described cases of incidentally detected paragangliomas. While the literature is dominated by cases of younger patients, the case described above is in an older man [1-3,5,8]. Unlike other cases, which are associated with catecholamine production and more likely to be associated with hypertensive crises, the anatomical location of the tumor was crucial in our case [1-3,8,9]. Due to its location in the left atrium, dilation of all cardiac compartments was subsequently verified, which did not indicate remodeling in subsequent examinations. As in many other cases, our patient also had arterial hypertension, which in the preoperative period was accompanied by several high-pressure peaks that no longer occurred in the postoperative period [1-3,8-10]. In accordance with recommended guidelines, our patient undergoes annual CT screening to rule out recurrence or metastases [6].

Conclusion

This text summarizes the case of a patient with an incidental finding of a bulky rare tumor of the posterior mediastinum, which was enucleated in its

entirety during biopsy. Paragangliomas are extremely rare tumors that can recur after removal of the primary benign tumor, but based on current data, their long-term survival prognosis is good.

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