Angioleiomyoma: An unusual heart tumor

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ABSTRACT

Introduction: This is a case of a rare secondary heart tumor, manifesting with very unusual clinical symptoms. As cardiac tumors are very rare, secondary cardiac tumor angioleiomyoma originated from vena cava inferior was never described in literature, as far as we are concerned.

Case Report: Our patient was admitted to our hospital with very unusual symptoms—deterioration of the right eye vision, chorioretinitis, chills, and muscle aches. After various diagnostic tests, it turned out to be an embolism of the tumor cells, which originated from vena cava and the heart. After pathological tests the tumor turned out to be angioleiomyoma, originated from vena cava inferior to the right atrium.

Conclusion: There are no specific manifestations of angioleiomyoma, all the symptoms depend on tumor site and spreading. In cases like this, it is important to suspect not only thrombosis, which seems to be the most common option, but tumors should be also considered.

Keywords: Angioleiomyoma, Computed tomography, Heart neoplasms, Thoracic surgery, Vena cava inferior

INTRODUCTION

Cardiac tumors are very rare and constitutes only 0.001–0.28% of autopsy cases [1, 2]. Tumors, originated from the heart, are classified into three groups—primary, secondary, and metastatic tumors. Primary tumors are originated from heart tissues and localized anywhere in the heart. Secondary tumors are originated in other organs—most commonly lungs, breasts, blood, or skin, and spreads to the heart, they are 30–40 times more common than primary [3]. Also, tumors of the heart can be metastatic cardiac tumors in the presence of malignancies elsewhere in the body [2]. 75% of tumors are benign and 25% are malignant [4]. Metastases are discovered much more frequently than primary tumors of the heart, being diagnosed in over 10% of tumor patients at the autopsy [5].

Patients present nonspecific symptoms, depending on tumor site and infiltration. In most cases, the general symptoms of malignancy appear—subfebrile temperature, weight loss, exhaustion, muscle pain, night sweats, coughing, or leukocytosis [6, 7]. The most common cardiac symptoms are blood flow obstruction, which leads to heart failure or dyspnea, arrhythmias, especially atrioventricular block, and embolisms [5]. Also, it is common, that cardiac tumor is asymptomatic and is discovered accidentally, during an imaging examination [3].
Angioleiomyoma

Angioleiomyoma, also known as vascular leiomyoma, is a rare, benign, solitary tumor, which originates from the vascular smooth muscle. It could be found anywhere in the body [8]. Angioleiomyomas present commonly between people of age 30–50 [8, 9]. The etiology of these tumors is unclear. Suggested etiological features of these tumors are infection, trauma, venous stasis, and hormonal factors, especially associated with estrogens [9, 10]. Also, there may be genetical predisposition [9]. Histopathologically, these tumors are comprised of mature smooth muscle interwoven with vascular channels [10]. Morimoto in 1973 studied 241 cases of angioleiomyomas and classified them into three histological types:

1. **Solid**—the most common type, which is composed of compact smooth muscle and many small vascular channels. This type of tumor is three times more common in females than in males.

2. **Venous**—composed of loosely organized smooth muscle bundles, which intertwine with the thickened muscular walls of venous channels. It is more common type in male patients.

3. **Cavernous**—consists of dilated vascular channels and less smooth muscles. This type is the least common type, which occurs more in male patients [9–11].

Preoperative diagnostics of angioleiomyoma are difficult and often angioleiomyomas are found accidentally. The most common diagnostic instruments are imaging techniques—ultrasound, computed tomography (CT) scan, or magnetic resonance imaging (MRI). The diagnosis of angioleiomyoma is confirmed by mass biopsy and histopathological examination. In case of angioleiomyoma, the only possible treatment is surgical removal of tumor. The recurrence after surgical removal is very rare [8].

**CASE REPORT**

A 45-year-old woman was presented to our hospital with six months lasting chorioretinitis, chills, and muscle aches of unknown origin and deteriorating of vision in the right eye. The patient said that pain in the knees and hand joints was more common in recent years. Subsequently, the patient's eyesight began to worsen, she began to see flashes of light and eye floaters in the right eye (lat. oculus dextra (OD)), therefore the patient was brought to the department of ophthalmology. Physical examination revealed nonfebrile status, normal heart rate, and normal blood pressure. Ophthalmological examination disclosed swelling of the macular area of the OD, yellowish-gray areas spread temporally from the fovea, vision (V) OD=0.14 with refractive correction (cc) with sphere ( sph), −0.5=0.28; V oculus sinistra (OS)= 1.0; intraocular pressure OD = 19.3 mmHg; OS = 15.3 mmHg.

An electrocardiogram (ECG) was performed and revealed sinus rhythm with heart rate of 64 bpm, PR duration 190 ms, QRS duration 112 ms, QT/QTc ratio 404/409 ms. After collection of anamnesis and physical examination, it seemed that the problem could be rheumatological, neurological, or oncological disease.

Laboratory tests: blood test showed lowered number of platelets. Erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), antinuclear antibodies (ANA), and anti-neutrophil cytoplasmic antibodies (ANCA) were within the normal range. Immunoglobulin (Ig) G for toxoplasmosis was increased (>300 kIU/L); however, toxoplasmosis IgM was negative. IgG and IgM for *Borellia* spp. were found negative. Chest CT was performed and revealed signs of chronic pulmonary embolism (PE). Heterogeneous masses in the vena cava inferior (VCI) and right atrium were observed (Figure 1). Data on sarcoidosis and tuberculosis were insufficient. A structure reaching the stomach wall was visible between the spleen and the stomach—a more detailed assessment required CT of the abdominal organs. Abdominal CT was performed and showed VCI thrombosis extending below the renal blood vessels and suspicious formation in the right atrium. Also, there were several solid formations in the right lung—possibly metastatic involvement. On the left, under the diaphragm, a solitary derivative was observed. Lower extremity ultrasound was completely normal and did not reveal any thrombosis. Heart ultrasound showed masses in the right atrium that were thought to be thrombi (Figure 2). All the imaging tests revealed spread thrombosis. The patient was subsequently transferred to the cardiology department of our hospital for further evaluation and treatment.

At the mentioned department mammography, esophagogastroduodenoscopy, endoscopic ultrasound, pelvic MRI, and bone scintigraphy were performed and no pathological changes were found. Heart MRI was performed—a large construct extending into the VCI was seen in the right atrium, and another construct was observed in the right ventricle drainage tract, extending into the pulmonary artery trunk and right pulmonary artery. Derivatives in terms of MRI traits are partially similar to thrombotic masses, but the tumor component could not be ruled out (Figure 3).

The only option was diagnostic and simultaneously therapeutic surgery. During the surgery, in the working heart, the right atrium was opened, derivatives in the atrium leading to the VCI and the right ventricle were observed. Pulmonary artery was opened and it was observed that the derivative goes to the right pulmonary artery—it was opened and a partially fixed solid consistency was removed from the pulmonary artery. Moreover, the derivative in VCI was revised (it was easily detached from the blood-vessel wall; however, it was fixed distally). The derivative was detached and sent for urgent pathological examination (answer—data on oncology is insufficient, more detailed tests should be performed). The incision was extended downwards by performing...
a right subcostal incision, adhesions were dissected, VCI was accessed, and the derivative was removed. Perioperative transesophageal echocardiography was performed and no derivatives in the heart cavities were observed. Histological examination confirmed a rare benign tumor: angioleiomyoma, most likely originated from VCI. After the surgery, symptomatic treatment and antithrombotic therapy were provided for the patient. The CT scan was performed to assess the result of the surgery, no masses were found (Figure 4).

DISCUSSION

Angioleiomyoma, also known as vascular leiomyoma, is a rare, benign, solitary tumor that originates from the vascular smooth muscle (tunica media) [12]. Angioleiomyomas commonly presents for people of age between 30 and 50 [8, 9]. Moreover, male-to-female ratio of angioleiomyomas is about 1:1.7 [12]. It was consistent in our case, because the patient we present is female and was 45 years old when angioleiomyoma was diagnosed. This tumor usually occurs anywhere in the body, most frequently in the lower extremities [13]. We also found several cases of angioleiomyomas in the oral cavity, sinonasal tract or uterus; however, no cases with angioleiomyoma which originated from VCI. Yet we
found several cases, which found angioleiomyomas in other organs (e.g., in uterus) that extended into VCI [14]. Our patient was diagnosed with angioleiomyoma which originated from VCI extending to the right atrium and even to the pulmonary artery. A majority of these tumors are quite small, generally less than 2 cm in diameter, which was not consistent with our case. Classically, the most evident symptom of angioleiomyoma is pain with or without tenderness [15]. Pain and/or tenderness may be the presenting feature in 45–70% of all cases [9]. In our case, the patient was presented with six months lasting chorioretinitis, chills, and muscle aches of unknown origin and deteriorating right eye vision, which is likely to be the consequence of emboli from tumor. These mentioned symptoms were not common in patients with angioleiomyomas. Moreover, angioleiomyoma is seldom diagnosed before the surgery [12]. As it was in this case, in the terms of MRI, the derivatives in VCI was differentiated between the thrombotic masses and tumor components.

In this case, the discovery of the tumor was an accident. The patient was admitted to the hospital because of deterioration of her vision in the right eye, chorioretinitis. It was unclear, what is the origin of this deterioration. There were no symptoms related to cardiovascular system, just weakness. After eye examination, the thromboembolism was suspected. The CT scan was performed, there were signs of chronic PE in the lungs. Also, heterogenous masses in the right atrium and VCI were found. Heart ultrasound showed the same masses in the right atrium. In suspicion of PE and in the presence of deterioration in the right eye that may be originated because of thromboembolism, the thrombotic masses were suspected. But it was not clear, so oncological process was also a diagnostic option. After this, the heart MRI was performed. It has shown heterogenous masses in the right atrium, VCI, right ventricle drainage tract, and right pulmonary artery. It was still unclear, what kinds of masses were found. The differential diagnostics was basically between two options—massive spread thrombosis or oncological process, especially with anamnesis of benign uterus tumor. The only option to differentiate between these two options was diagnostic and simultaneously therapeutic surgery. Only then angioleiomyoma was found and confirmed by pathohistological test.

Perioperative and postoperative state of the patient was without complications. First follow-up visit was after three months post-surgery. Echocardiography showed dilatation of right ventricle and atrium [right ventricle end diastolic diameter (RVEDD) was 38 mm]. The quality of life questionnaire (MERIT-HF Study Group) was filled, the result was 66 points. The patient has regular appointments at a cardiologist, and was admitted to the heart failure program. After a year and a half after surgery, echocardiography and quality of life were assessed again—dilatation of right atrium and ventricle was observed (RVEDD was 42 mm). The quality of life questionnaire result was 67 points. The last visit was on August 3, 2020. The patient had these complaints—severe fatigue and shortness of breath during physical activities. The echocardiography was performed—it has shown no negative dynamics, ejection fraction was assessed 55%, also the quality of life was assessed, the result was 66 points. There was no recurrence of the tumor found, as the recurrence rate of angioleiomyomas is very low—only 0.4–7% [8, 16].

**CONCLUSION**

There are no specific manifestations of angioleiomyoma, all the symptoms depend on tumor site and spreading. The patient, who was admitted to the ophthalmology department because of deterioration of the right eye vision and suspicion of thromboembolism, was diagnosed with a very rare benign tumor—angioleiomyoma of the right atrium and vena cava inferior, and had no cardiac symptoms. All the imaging tests led us to spread thrombosis diagnosis, so in cases like this, it is important to suspect not only thrombosis, but tumors should be considered too.

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Author Contributions

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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