Primary Intimal Pulmonary Vein Sarcoma with Expansion to the Left Atrium

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Abstract

We have recently treated a patient with pulmonary vein sarcoma. It is a 65 year old women who was admitted to the hospital because of the chest pain with propagations down her right arm, followed by suffocation and coughing out content with traces of blood and febrility >38°C. The primarily suspected pulmonary embolism was ruled out after the diagnostic heart CT scan and transthoracal and transeophageal echocardiogram which verified the existing of a tumor mass in the left atrium. The patient underwent an urgent surgery and the tumor mass was removed surgically from the upper right pulmonary vein. The path histological diagnoses revealed pulmonary vein intimal sarcoma. Pulmonary vein intimal sarcoma is one of the rarest sarcoma subtypes. To our knowledge, there is only one published case of such pathology in the literature.

Introduction

A 65 year old female patient was admitted in the Emergency Room of Montenegrin Clinical Hospital because of the chest pain with propagations down her right arm, followed by suffocation and coughing out content with traces of blood and febrility >38°C. She mentioned the poor physical exercise tolerance with occasional swelling of lower extremities. She was experiencing the discomforts for the previous 10 days [1-3]. The patient had been a smoker for a long time. She was aware of cardiac arrhythmia which is why she had been prescribed an oral anticoagulant therapy. She had been aware of her condition of thrombocytopenia too. Twenty years ago she underwent hysterecctomy with bilateral adnexectomy.

During the physical examination the patient was eupnoptic while resting, acyanotic, anicteric and afebrile. Her vital signs were: cardiac frequency (FR) 85/min, blood pressure (TA) 100/60 mmHg, respiratory frequency (FR) around 16-18/min, saturation (SO2) around 96%. Pulmonary auscultation revealed a weakened basal respiratory wheeze with sporadic late inspiratory bilateral basal crackles [4-7]. The auscultation also showed a rhythmic cardiac activity with clear tones and without wheezes. The extremities were without edemas, varicosity or deformities. EKG showed the sinus rhythm of around 85/min, normogram, without signs of ischemia.

Laboratory Test Results

Se 30...72, CRP 10...160, D-dimer 0,73, PV 14,2, INR 1,2, fibrinogen 81, Le 5,74, Er 3,96, Hgb 122, HCT 0,35, Tr 162. Pulmonary X- ray showed left basal pleural pericardial effusion. Chest CT scan describes a 35 mm thrombus in the main part of Art. Pulmonalis which extends to the branch for the upper lobe with consequential parenchyma consolidation ventrally bigger than 50 mm, pericardially along with the pericardium almost completely occupying the front FC sinus and dorsally with the lower branch of interlobar incisura up to 30 mm and medially almost at the very top of the lungs with the size of up to 20 mm with the complete hypo perfusion of the upper lobe with evidently positive air bronchogram.

Heart CT scan describes in the left atrium a big tumor-like change with the dimensions of 32x24mm, 32 mm long along which thrombotic masses are evident. The tumor covers both lower left
Several tissue fragments with the size up to
Tumor tissue made of groups and solid clusters of
tissue. Path histological diagnosis intimal sarcoma (gradus 2).
mediastinal lymph node is of preserved structure, without tumor
S100, CD 34 and HMB 45 negative. The path histologically analyzed
immuno histo chemical polymorphic, oval and hyper chromatic.
atypical spindle and oval cells. The nuclei are slightly to moderately
in slime.
40mm and with softer to medium hard consistency, partly covered
intestinal interventions on the valvular apparatus.
veins, with a long petiole the base of which was attached to the
atrium which was obstructing the confluence of lower pulmonary
left atrium and inter atrial septum, tumor jelly-like mass was
establishing of extracorporeal blood flow, with the incision through
dynamically significant stenosis on coronary arteries.

General endo tracheal anesthesia was induced and after the
establishing of extracorporeal blood flow, with the incision through
right atrium and inter atrial septum, tumor jelly-like mass was
identified with the diameter of around 30 mm on the top of left
atrium which was obstructing the confluence of lower pulmonary
veins, with a long petiole the base of which was attached to the
wall of left upper pulmonary vein. The tumor was not in contact
with the mitral apparatus structures and there was no need for interventions on the valvular apparatus.

**Patho Histological Results**

**Macroscopic:** Several tissue fragments with the size up to
40mm and with softer to medium hard consistency, partly covered
in slime.

**Microscopic:** Tumor tissue made of groups and solid clusters of
atypical spindle and oval cells. The nuclei are slightly to moderately
polymporphic, oval and hyper chromatic.

A. A moderate number of mitoses – focally between 5 and 10
    mitoses on 10 fields of big enlargements.

B. The tumor contains bleeding zones and small necrosis
    focuses.

C. Hypo cellular slimy degenerated areas were present.

**Immuno histo chemical**

Tumor cells are Vimentin and Actin positive; CK, EMA, Desmin,
S100, CD 34 and HMB 45 negative. The path histologically analyzed
mediastinal lymph node is of preserved structure, without tumor
tissue. Path histological diagnosis intimal sarcoma (gradus 2).

**Discussion**

Primary neoplasm of big blood vessels (aorta, pulmonary
artery and pulmonary veins, v. cava sup and inf) are extremely
rare. The pathogenesis of those tumors is still vague and signs
and symptoms are unspecific which makes differential diagnosis
difficult and postpones a final diagnosis. Around 400 cases of
primary tumors have been mentioned in reference books so far,
30 of them being cases of primary pulmonary vein tumors. The
majority of those tumors are leiomyosarcomas and there is one
case of myxosarcoma and one case of myxoidfibrosarcoma. These
pulmonary vein sarcomas are often located in the upper right and
lower left pulmonary veins. To our knowledge this is the second
published case of pulmonary vein non-myxoid intimal sarcoma
(libro sarcoma). The biggest group of patients with pulmonary vein
primary sarcoma includes 17 cases of leiomyosarcoma. Blood vessel
tumors appear on average when people are in their 40s and they are
equally found in both sexes (leiomyosarcomas mildly predominate
with females). The most common symptoms like cough, hemoptysis,
dyspnea, chest pain and pleural effusions are unspecific. Pulmonary
vein sarcomas, especially intimal sarcomas can be misdiagnosed
as pulmonary thromboembolism, which was initially the case
with our patient, because of the fact that it is rare and because of
unclear clinical progress. The consequences can be inadequate
treatments like a prolonged anticoagulant therapy or thrombolysis.
In the biggest series of pulmonary vein leiomyosarcomas, around
50% of the cases were diagnosed with the help of CT and/or MRI
angiography. It is important here to be mentioned that in majority
of such cases the tumor masses are thought to be originating from
left atrium. Bronchoscopy and transesophageal echocardiography
together with heart CT are important for proper diagnosing and
urgent management of those cases.

The difficulties in diagnosing are only one of the aspects related
to bad prognosis of primary pulmonary vein tumors, regardless
of the subtype. Only two cases were recorded when patients lived
three or more years after the treatment of leiomyosarcoma and
only one patient lived more than nine months after the treatment of
myxosarcoma. Both patients with leiomyosarcoma had a complete
surgical resection with clean remaining edges. After the resections
there was lots of case of relapsing which makes us conclude there is
a need for adjuvant or non-adjuvant therapy. Surgery with adjuvant
chemotherapy and radiotherapy can improve short term survival.
However the prognosis is not good with the rate of five year survival
between 0 and 6 percent.

The ideal management of pulmonary vein sarcomas is still
being discussed. Extensive surgical resection is for now considered
to be the most optimal solution. For patients with incomplete
resection or local relapsing heart transplanting is taken into
consideration as a valid solution. Although some authors do not
think that chemotherapy and radiotherapy are efficient, some other
authors hope that systemic chemo and radio therapy could increase
the survival rate and achieving therapy and palliative benefits could
avoid potential surgical complications.
References


