CASE REPORT

A Rare Tumor - Primary Leiomyosarcoma of Pulmonary Artery. Case Presentation

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Rezumat

Leiomiosarcomul primar de arteră pulmonară este o tumoră extrem de rară, iar diagnosticul său este dificil de stabilit. Prezentăm cazul unui pacient de 63 ani, fost fumător, cu tuberculoză pulmonară în antecedente, diagnosticat în 2014 cu leiomiosarcom de arteră pulmonară. Intervenția chirurgicală (trombendanterectomie bilaterală de arteră pulmonară dreaptă și stângă) a fost efectuată în septembrie 2014, iar rezultatul examenului histopatologic al piesei rezecate a fost de leiomiosarcom. Chimioterapia adjuvantă (4 cure) a fost administrată în octombrie 2014 dar a fost întreruptă datorită reactivării tuberculozei. Șapte luni mai târziu, pacientul a fost diagnosticat cu metastaze cerebrale pentru care a efectuat radioterapie externă, tehnica "whole brain", în 2015. Radioterapia a fost întreruptă datorită deprecierii statusului de perfomanță neurologic. Leiomiosarcomul de arteră pulmonară este o tumoră pulmonară rară a cărui diagnostic este dificil de stabilit datorită simptomelor nespecifice, care adesea sunt interpretate greșit fiind relaționate cu trombembolismul pulmonar. În acest articol, prezentăm probleme de diagnostic, opțiuni terapeutice și prognostic precum și o revizuire a literaturii de specialitate.

Cuvinte cheie: arteră pulmonară, leiomiosarcom primar, tratament

Abstract

Primary leiomyosarcoma of the pulmonary artery is an extremely rare tumor and its diagnosis is very difficult to establish. We present the case of a 63-year-old male patient, previously smoker, with previous pulmonary tuberculosis, diagnosed in 2014 with leiomyosarcoma of pulmonary artery. Surgery (right and left arteriotomy and bilateral extended end arterectomy) was performed in September 2014 and histological examination of the resected mass was consistent with leiomyosarcoma. Adjuvant chemotherapy, 4 courses, was also, administrated in October 2014; chemotherapy was interrupted because of tuberculosis reactivation. Seven months later, the patient developed distant brain metastases for which we performed external beam radiotherapy in 2015, in "whole brain" technique. The radiation dose was incomplete because of neurological performance status decreasing. Leiomyosarcoma of the pulmonary artery is a rare tumor of the lung and its diagnosis is very difficult because the non-specific symptoms which are often misinterpreted as being related to pulmonary thromboembolism. The literature is reviewed and we discuss the diagnosis, option treatment and prognosis.

Keywords: pulmonary artery, primary leiomyosarcoma, treatment

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INTRODUCTION

Pulmonary artery sarcomas (PA) are uncommon; the first case was documented in 1923 by Mandel Staam. Leiomyosarcoma, fibrosarcoma and rhabdomyosarcomaare the most often histologically confirmed incidences. PA tumor is frequently misinterpreted as thromboembolism, as its correct diagnosis is difficult. Magnetic Resonance Imaging (MRI) is superior in providing an accurate diagnosis. Surgery is the only therapeutic strategy which may extend life expectancy. The survival rate in the absence of surgical care is of 1.5 months, survival for 5 years – 6%. In most cases, death occurs by heart failure, and rarely by distant metastases.

In what follows, we present the case of a 63-year-old patient, formerly a smoker, who presents recurrent episodes of pulmonary thromboembolism, starting a year prior to the diagnosis. His medical history includes: pulmonary tuberculosis (1983), superior left lobe (SLL) BK (+), bilateral inguinal hernia, operated 2007-2010, penicillin allergy, chronic venous insufficiency of legs, minimal aortic insufficiency, mild mitral and moderate tricuspid.

The patient presents for surgery to the Institute of Cardiovascular Diseases from Timisoara. In September 2014, surgical intervention is decided, and during operation, it is noted the right pulmonary artery, firm at palpating, with infiltrated arterial wall apparently occupied by intraluminal thrombosis. From the operatory protocol, we note longitudinal arteriotomy on the right pulmonary artery, extended beyond bifurcation. After the incision, a massive tumor mass is observed: almost intraluminal occlusive, yellowy, friable, fat, infiltrating the arterial wall and covering the vascular tunic, making up an intraluminal tumor mass lump of the pulmonary arterial tree. The tumor is removed piece by piece by retrograde excision up to the pulmonary trunk and to the tumoral mass in the lobar arteries.

Longitudinal arteriotomy on the left pulmonary artery, extended up to the bifurcation into the superior and inferior lobar arteries. After the incision, the same yellowy, fat and friable tumor mass is observed, without the profound infiltration of the arterial wall; a transversal incision and separation of the arterial intima are further pursued, with extended end arterectomy, followed by bilateral thromboendarterectomy of right and left pulmonary artery. Post-operative evolution is favorable. Post-operative echocardiography: left ventricle (LV) – normal, FE 55%, dilated right cavities PSAP40 mmHg, small quantity of liquid in the right pleura. The patient is discharged, being hemodynamically stable, with oral anticoagulant treatment: Syntron 4 mg, 1 pill per day, Cordarone 200 mg, 1 pill/day, Diurex 1 pill/day, ACC 200 mg, 2 pills/day, Aspacardin 6 pills/day. The histopathological report reveals malignant mesenchymal tumoral proliferation (chondrosarcoma, osteosarcoma chondro-myxosarcoma) and recommends immunohistochemical test (IHC). The IHC results establish the diagnosis of intimal leiomyosarcoma.

Chemotherapy (CMT) is initiated for the patient, within normal hematological and biochemical parameters: Gemcitabine (900 mg/m^2, day 1 and 8) and Docetaxel (75mg/m^2, day 8), associated with G-CSF without hematological toxicities or other acute secondary effects. After three CMT courses, in January 2015, a reevaluating abdomen and thorax CT scan is performed in the presence of contrast agent, which reveals a slight dilatation of the pulmonary trunk 38 mm, maximum caliber with homogenous aspect; a cavitary image of 16/20 mm on the SLL dorsal segment with clearly delineated hydroaeric level with thin wall, associating densification and perilesional alveolar filling area. Similar densification areas at the superior right lobe (SRL) and inferior left lobe (ILL) level with non-specific CT aspect; similar mediastinal adenopathies with comparable size, located in the Barety lodge, aortopulmonary window and precarinal. No pleural or pericardial effusion. Liver – no secondary determinations; suprarenal glands with normal aspect, no abdominal adenopathy, no ascites.

There is suspicion of SLL pulmonary tuberculosis. In January 2015, the patient is admitted to "Marius Nasta" Pulmonology Hospital, with cough with minimal sputum expectoration, dyspnea progressively aggravated in the last three months, without weight loss, fever or hemoptysis. Bronchoalveolar lavage: no tumoral cells, no BK negative bacterial elements or fungi, bronchial aspirateBAAR+ (9BAAR)/camp. The oncological treatment is interrupted and TSS initiation is decided, antituberculostatic regime without streptomycin in February, 7/7 HRZES associated with anti-anemics, anti-coughs, diuretics, anxiolytics, in oral anticoagulant treatment.

The medical commission at the Pulmonology Institute recommends reevaluating thoracic CT scan in two months in order to establish the resuming of the oncology treatment.

Normal biochemical and hematological constants, except for ESR=30 mm/h and Hb=11.2 mg/dl, Leucocytes= 11800/mm^3, thrombocytes = 552000/mm^3, INR=1.4.

During the tuberculostatic treatment, the neurological deficit occurs: right hemiparesis, confusion, suspicion of cerebral secondary determinations. Cerebral CT scan reveals: multiple nodular lesions with aspect

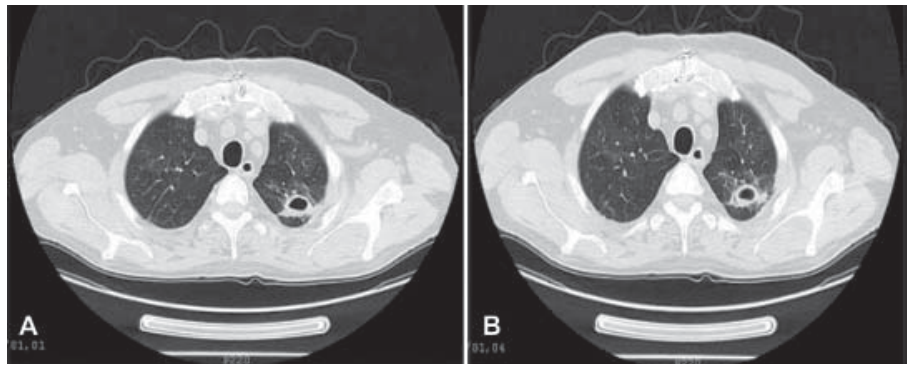

Figure 1 A,B. Cavitary images of 16/20 mm with clearly delineated hydroaeric level with thin wall, associating densification and perilesional alveolar filling area on the LSS dorsal segment.

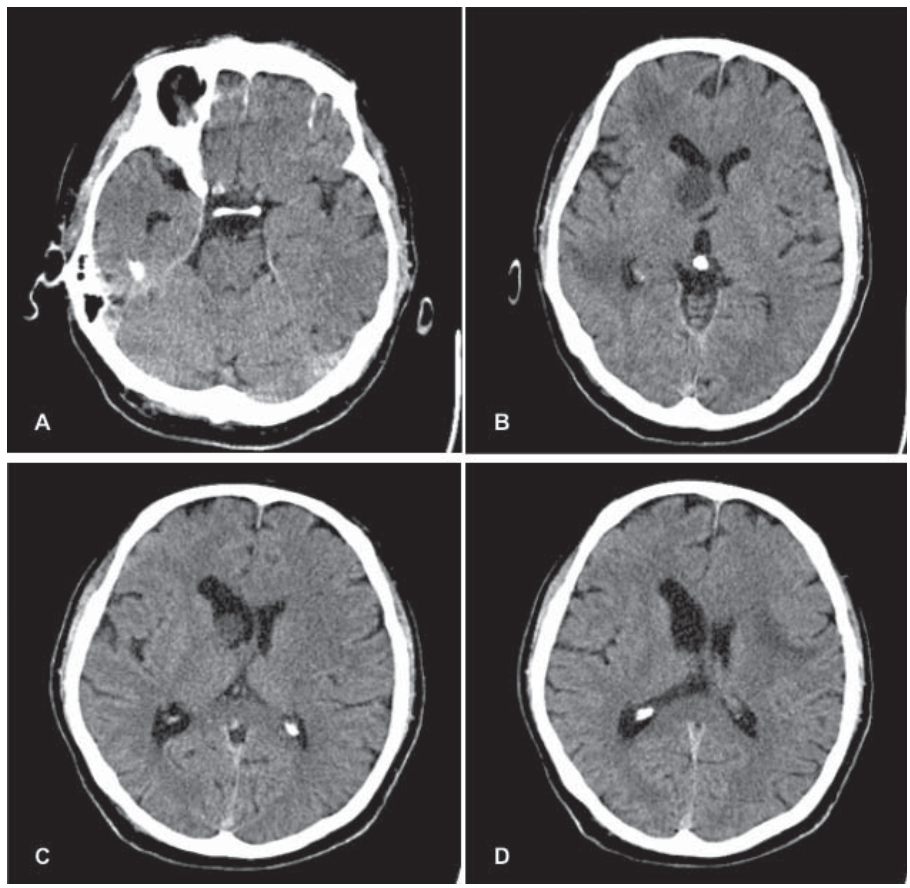

Figure 2 A,B,C,D. Cerebral CT scan: multiple nodular lesions with aspect of secondary determinations with peripheral iodophilia in the ring and edema of adjacent right white substance located supratentorially (frontal left, frontal and temporal right) and infratentorially at the right side of cerebellum, sized from 1 to 25 mm.

of secondary determination with peripheral iodophilia in the ring and edema of right adjacent white substance located supratentorially (frontal left, frontal and temporal right), and infratentorially, at the right side of cerebellum, with sizes varying from 10 to 25 mm. Thoracic CT reveals nodular lesion of ≈ 27 mm, partly excavated from the left apex, also visible in January 2015, transformed into a cavitary lesion with thin walls of 42 mm. Adjacently, inferior, in LSS, are identified two sub-centimeter nodular lesions and another one of 14 mm in the medium lobe, all having occurred after the January 2015 examination. Two lacunar lesions, one at the root of the pulmonary artery trunk and another one at its bifurcation are also observed, which were not identified during the previous examination. In rest – the image is superimposable with the January 2015 scan.

Cerebral depletive treatment is also initiated: Manitol, corticotherapy. The patient presents at Emergency County Hospital of Galati with recommendation of palliative radiotherapy, which is conducted using the "whole brain" technique: DT=15Gy/5 fraction/5 days, D/fraction = 300 cGy out of the proposed dosage of 30 Gy. Radiotherapy is interrupted due to decrease of neurological status performance, and the patient deceases within three weeks, after 7 months survival.

DISCUSSION

Pulmonary artery sarcomas (PA) have a rare incidence, the first case being documented in 1923 by Mandel Staam. Histologically speaking, the most often encountered cases are of leiomyosarcoma, fibrosarcoma or rhabdomyosarcoma.

Fifty more cases have been described in the literature up to the year 2000. The literature in the field reporting PA sarcomas is scarce. Incidence is of 0.001-0.003%, women being affected twice as often as men. From the data collected from the specialized literature, the average age is 52 in cases with this diagnosis. PA tumor is frequently misinterpreted as thromboembolism, which makes the diagnosis more difficult to establish. Clinical symptomatology includes dyspnea, thoracic pains, hemoptysis, and cardiac insufficiency specific symptoms. Many patients are initially cured for chronic pulmonary thromboembolism, and when the anticoagulant treatment is not efficient, additional investigations are recommended, which leads to the correct diagnosis. In the absence of the risk factors for venous thromboembolism and symptomatology persistence during treatment (fever, nocturnal sweating, weight loss, physical asthenia), suspicion of malignancy raises. The imagistic aspect of pulmonary thromboembolism and PA tumor is similar in CT scans. MRI is superior in determining the diagnosis. For an accurate diagnosis, transesophageal echocardiography (TEE) may also be used[1,2]. Differential diagnosis between pulmonary thromboembolism and PA sarcoma is difficult, and is often established either after resection or during autopsy. PET-CT is also useful in differentiating the tumor from thromboembolism.

The rate of survival in the absence of surgical treatment is of 1.5 months; five years survival – 6%. In most of the cases, death occurs by heart failure.

In 1972 Mofatetal[3] reported a few weeks to 3.5 years survival. Baker, in 1985[4] and Head in 1992[5] reported up to ten years survival after the complete surgical resection.

Prognosis is highly unfavorable in the cases of PA sarcoma[6]. In a review of 93 cases, Kruger et al.[7] reported an average survival of 1.5 months; in the cases of patients who underwent surgical excision, the survival was up to ten months. The treatment consists in surgical intervention, and other therapeutic options include adjuvant chemotherapy and radiotherapy.

Radical surgery performed as early as possible associated with cardio-pulmonary bypass represents the only strategy for potential healing.

It is important to obtain tumor free margins, as much as possible. Cryopreserved arterial allograft reconstruction and/ or pneumonectomy should also be considered as options.

In 2012, Kashima et al. published[8] a similar case of leiomyosarcoma of PA trunk of a 53-year male who underwent incomplete resection followed by chemotherapy. The patient survived for 20 months after surgery, and the autopsy revealed metastases only in his stomach.

In two cases out of three, metastases occur at the lungs level, and metastasis incidence is in 20% of the cases.

Doxorubicin chemotherapy is a therapeutic option in case of incomplete surgical intervention. Two of the most active agents are doxorubicin and Ifosfamide, the former being active in doses of 75 mg/m^2 or higher, while the combinations of Ifosfamide and Epirubicin or Doxorubicin and Ifosfamide may also represent viable options.

In the present study case, the chemotherapeutic option included Doxorubicin and Gemcitabine, but the patient only underwent four courses, chemotherapy being interrupted because of pulmonary tuberculosis reactivation.

CONCLUSIONS

PA sarcoma is a rare disease with an extremely unfavorable prognosis and a diagnosis difficult to establish. Surgery is the only therapeutic strategy which may extend survival.

Conflict of interests: The authors declare that there is no conflict of interests regarding the publication of this paper.

Author contribution: All authors contributed equally to this work.

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