

Case Report

A Case of Endometrial Stromal Sarcoma Revealed by Lung Metastasis 7 Years after Hysterectomy which Led to Histological Diagnosis of Uterine Fibroid

Moussa Bathily¹, Abdoul K Dembélé², Boubacari A Touré^{1,2}, Youssouf Badiaga¹, Cheick O Samaké¹, Fatoumata Diakité³ and Dapa A Diallo^{1,2*}

¹Department of Haematology-Medical Oncology, CHU du Point G, Bamako, Mali

²Sickle Cell Disease Research and Control Centre (CRLD), Bamako, Mali

³Department of Medicine, CHU Gabriel Touré, Bamako, Mali

Abstract

Endometrial stromal sarcomas are uterine malignant tumours. Recurrences as extragenital locations of these tumours are rare and, occur sometimes long time after the primary tumour treatment. Here we report a case of recurrence of endometrial stromal sarcoma revealed by lung metastasis in a 65-year-old woman who had, 7 years earlier, experienced a total hysterectomy whose histology concluded to the diagnosis of uterine fibroid. Its interest is clinical, diagnostic and prognostic.

Keywords: Diagnosis; ESS; Prognosis; Recurrence; Sub-saharan Africa

Introduction

Sarcomas Developed on Endometrial Stroma Sarcoma (ESS) are rare mesenchymal tumours of the uterus that account for between 0.2%

***Corresponding author:** Dapa A Diallo, Sickle Cell Disease Research and Control Centre (CRLD), University of Sciences, Techniques and Technologies of Bamako (USTTB), 03 BP: 186, Mali, Tel: +223 20223898; Fax: +223 20223899; E-mail: dadiallo@icermali.org

Citation: Bathily M, Dembélé AK, Touré BA, Badiaga Y, Samaké CO, et al. (2019) A Case of Endometrial Stromal Sarcoma Revealed by Lung Metastasis 7 Years after Hysterectomy which Led to Histological Diagnosis of Uterine Fibroid. J Clin Stud Med Case Rep 6: 064.

Received: January 29, 2019; **Accepted:** February 18, 2019; **Published:** March 05, 2019

Copyright: © 2019 Bathily M, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

and 1% of all malignant uterine tumours and 15% to 20% of primary uterine sarcoma [1]. The main clinical symptom of ESSs is abnormal vaginal bleeding. ESSs usually originate from the endometrium, but they can also develop from intra- or extra-uterine endometriosis sites [2]. These are tumours characterized by wide anatomopathological heterogeneity. Based on cellular atypia, mitotic index and tumour immunohistochemistry, a distinction is made between high-grade ESSs, also known as ketogenic chorion sarcoma, and low-grade ESSs [3,4]. High-grade malignancies ESSs often have an aggressive outcome and are associated with short survivals [5], while low-grade ESSs have a slow outcome and better prognosis. Recurrences of treated ESSs frequently occur in the vaginal, pelvic or peritoneal cavity and less frequently in certain sites far from the initial site such as the liver, bone marrow and lung [6]; according to several authors, lung metastasis can occur several years after the onset of ESS [1,7]. Here are reporting a case of endometrial stroma sarcoma revealed by lung metastasis in a 65-year-old woman who had, 7 years earlier, experienced a total hysterectomy whose histology led to the diagnosis of uterine fibroid. In our knowledge, it is the first case reported in sub-Saharan Africa. The interest of the case is clinical, diagnostic, prognostic and therapeutic.

Observation

Mrs. AT, a retired executive secretary aged 65 and residing in Bamako, Mali, was received in April 2017 in the pneumology department in Mali for intermittent chronic cough with whitish sputum. X-ray and Computed Tomography (CT) scans examinations led to a diagnosis of pulmonary metastasis of cancer based on the observation of “balloon-release” pulmonary nodular images (Figures 1 and 2). The pulmonary nodules were also associated with hepatic and abdominal-pelvic nodules. Following the diagnosis, Mrs. AT travelled to Dakar, Senegal, in May 2017, for explorations and treatment. After bronchial fibroscopy with aspiration, the histological examination of bronchial aspiration samples concluded that there was no malignancy. On 25 September 2017, Mrs. AT was referred to our consultation. The clinical examination at this moment found a patient in good general condition with a WHO performance status score evaluated at 1 [8], her conjunctiva were well coloured, abdominal palpation did find no organ hypertrophy and the gynecological examination was normal. Mrs. AT was 6th gesture, 5th pare, had four living children and, an antecedent of abortion; she had a history of oral contraception use in 2007 and the onset of her menopause the same year. On 26 May 2010, she underwent total hysterectomy with bilateral annexectomy, whose histological examination, without an immunohistochemical examination, concluded that she had uterine fibroid. She had hypertension that had been regularly monitored with amlodipine (Amcal® 5mg/day) since 2011. The initial biological tests conducted following our consultation were normal (whole blood cell count, serology of hepatic viruses B and C as well as HIV, carcinoembryogenic antigen, creatinine and serum electrolytes). In October 2017, we decided to conduct a biopsy of the right lung nodules for a histological examination. Using biopsy specimens embedded in paraffin, an immunohistochemical staining has completed the histological examination by using a panel of antibodies including polyclonal antibodies AE1/AE3, polyclonal

antibodies anti-desmin, monoclonal antibodies against EMA (Epithelial Membrane Antigen), vimentin, estrogen receptors, progesterone receptors, TTF-1 (Thyroid Transcription Factor-1), Ki-67, CD34, CD10, chromogranin and synaptophysin. Morphological aspect and immuno-histochemical staining authorized to consider the diagnosis of endometrial stroma sarcoma. Indeed, in the lung parenchyma, under the pleura, a lesion has been found consisting of small cells with a regular round nucleus and fine chromatin; cytoplasm of these cells was pale and its limits were discrete. Labelling by antibodies AE1/AE3 was positive, EMA (Epithelial Membrane Antigen) and TTF-1 (Thyroid Transcription Factor-1) were labelled by specific monoclonal antibodies on the epithelium lining and, negative within the tumour; chromogranin, synaptophysin and CD34 were negative. Actin and desmin were focally positive and there were some positive cells for CD10. Progesterone receptors were highly positive and there was a low positivity of lesions for oestrogen receptors. Ki-67 was < 1%. The patient was monitored under symptomatic treatments. She was seen again for consultation on 09/08/2018 after which, we noted a WHO performance status still equal to 1, as well as a tumour progression marked by 50% and 30% increase in the size of liver and lung nodules respectively without clinical impact.

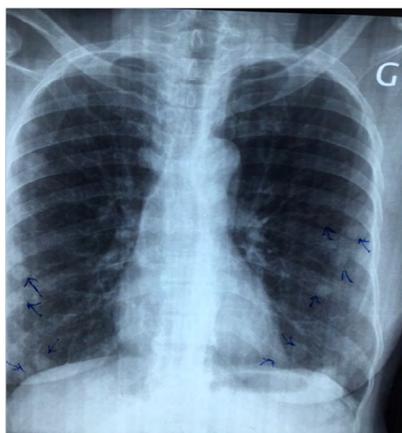


Figure 1: Chest x-ray photograph showing nodules scattered in the two pulmonary fields (arrows).

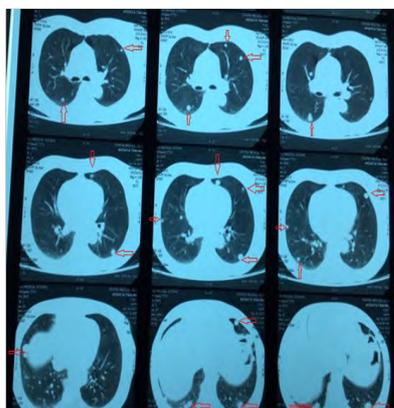


Figure 2: Chest CT scans showing nodules spread to both lungs on different sections (arrows).

Discussion

ESS is a rare tumour, accounting for between 0.2% and 1% of all malignant tumours of the uterus. Diagnosis is based on morphological and immunohistochemical or even molecular biology criteria [9]. Our patient had experienced a total hysterectomy with bilateral annexectomy in 2010, the histology of the operating material had not concluded malignancy, but it had not been completed by an immunohistochemical study; we cannot therefore a posteriori eliminate a false anatomopathological diagnosis. The diagnosis of recurrences of sarcomas far from the primary site of the tumour is generally difficult because of morphological aspects that are often not superimposed on those of the primary tumour. Thanks to immunohistochemistry, this diagnosis is facilitated if the right material is available for study. During our patient's medical treatment, the absence of malignancy of her lung nodules was noted in May 2017 based on the examination of bronchial aspirations. It was necessary to proceed in October 2017 or 5 months later, a biopsy that allow to affirm the malignancy of lung nodules. The false diagnosis made in May 2017 was clearly due to the use of inappropriate material for diagnosis. Most ESS recurrences take place within two years following diagnosis of the primary tumour, but very late recurrences have been described, particularly for low-grade lesions [4,7]. We were unable to obtain the hysterectomy part performed on our patient in 2010 for an immunohistochemical study due to lack of archiving, but two observations allow us to consider that our patient in 2010 had a low-grade ESS undiagnosed due to lack of immunolabelling: (i) the long period (7 years) between the treatment of the primary tumour and the diagnosis of recurrence, (ii) the histological and immunohistochemical characteristics of the lung tumours examined in 2017, revealing, in particular, the positivity of progesterone and oestrogen receptors and the low labelling of Ki-67. Low-grade lung sarcomas is an exceptional tumour whose incidence is estimated at one sarcoma per 500 lung carcinomas, or less than 50 new cases per year in France [10]. These tumours should be investigated for the existence of an extra-thoracic primary tumour, particularly uterine, which usually precedes the development of lung metastases by several years [5]. One of the singularities of the case we are reporting is the absence of gynaecological manifestations at the time of diagnosis of lung metastasis; some authors have reported ESS recurrences in the lungs without pelvic lesions [11]. These data clearly raise the issue of pathophysiology of ESSs recurrences and their extragenital locations, which has not yet been elucidated; the very variable time it takes to recur at extragenital sites, the often indolent course of the recurrences and the limited treatment options call for studies on the risk factors that promote their occurrence. Low-grade sarcomas are characterized by slow growth, with a 5-year survival rate estimated at between 80% and 100%. The specific mortality rate for low-grade ESSs is estimated at 15%-25% [12]. Their treatment is surgical for both primary tumour and metastases where possible. Radiation therapy can be offered for incompletely resected tumours, and it appears to be effective [13]. The case we are reporting could not be operated or irradiable even though the patient had a WHO stable performance status score of 1. Progesterone treatment has resulted in remission in some patients [14,15]; we did not offer this treatment to our patient due to the indolent clinical course of her disease.

This observation calls for systematic search for uterine sarcoma in front of low-grade lung sarcoma in a woman, as well as systematic immunohistochemical study of uterine tumours; it also calls for close monitoring of any woman who has experienced uterine tumour

hysterectomy regardless of the initial histology. The observation also raises the fundamental issue of risk factors for the spread of ESSs, which requires multicentre research studies.

References

1. Koss LG, Spiro RH, Brunshwing A (1985) Endometrial stromal sarcoma. *Surg Gynecol Obstet* 121: 531-537.
2. Hendrickson MR, Kempson R (1995) Pure mesenchymal neoplasm of the uterine corpus (4th edn). In: Fox H, Wells M (eds.). *Haines and Taylor, Obstetrical and Gynecological Pathology*, Churchill Livingstone, New York, USA, Pg no: 519-586.
3. Chang K, Crabtree G, Lim-Tan S, Kempson R, Hendrickson M (1990) Primary uterine endometrial stromal neoplasms. A clinicopathologic study of 117 cases. *Am J Surg Pathol* 14: 415-438.
4. Inayama Y, Shoji A, Odagiri S, Hirahara F, Ito T, et al. (2000) Detection of pulmonary metastasis of low-grade endometrial stromal sarcoma 25 years after hysterectomy. *Pathol Res Pract* 196: 129-34.
5. Michel G, Pfeiffer F, Duvillard P, Prade M, Castaigne D, et al. (1989) Sarcoma of the uterus. A clinical study apropos of 50 surgically treated cases at the Gustave Roussy Institute. Review of the literature *J Gynecol Obstet Biol Reprod* 18: 1024-1030.
6. Masand RP, Euscher ED, Deavers MT, Malpica A (2013) Endometrioid stromal sarcoma: A clinicopathologic study of 63 cases. *Am J Surg Pathol* 37: 1635-47.
7. Abrams J, Talcott J, Corson JM (1989) Pulmonary metastases in patient with low-grade endometrial stromal sarcoma: Clinicopathologic findings with immunohistochemical characterization. *Am J Surg Pathol* 13: 133-140.
8. Oken MM, Creech RH, Tormey DC, Horton J, Carbone PP, Davis TE, et al. (1982) Toxicity and response criteria of the Eastern Cooperative Oncology Group. *Am J Clin Oncol* 5: 649-55.
9. Tsuyoshi H, Yoshida Y (2018). Molecular biomarkers for uterine leiomyosarcoma and endometrial stromal sarcoma. *Cancer Sci* 109: 1743-52.
10. Nascimento AG, Uni KK, Bernatz PE (1982) Sarcomas of the lung. *Mayo Clin Proc* 57: 355-359.
11. Tuyaerts S and Amant F (2018) Endometrial Stromal Sarcomas: A revision of their potential as targets for immunotherapy. *Vaccines (Basel)* 6: 56.
12. Gadducci A, Sartori E, Landoni F, Zola P, Maggino T, et al. (1996) Endometrial stromal sarcoma: analysis of treatment failures and survival. *Gynecol Oncol* 63: 247-253.
13. Weitmann HD, Knoocke T, Kucera H, Potter R (2001) Radiation therapy in the treatment of endometrial stromal sarcoma. *Int Radiat Oncol Biol Phys* 49: 739-748.
14. Nakamura K, Nakayama K, Ishikawa M, Ishikawa N, Katagiri H, et al. (2016) Letrozole as second-line hormonal treatment for recurrent low-grade endometrial stromal sarcoma: A case report and review of the literature. *Oncol Lett* 12: 3856-3860.
15. Dupont NC, Disaia PJ (2010) Recurrent endometrial stromal sarcoma: treatment with a progestin and gonadotropin releasing hormone agonist. *Sarcoma* 2010: 353679.



Journal of Anesthesia & Clinical Care
Journal of Addiction & Addictive Disorders
Advances in Microbiology Research
Advances in Industrial Biotechnology
Journal of Agronomy & Agricultural Science
Journal of AIDS Clinical Research & STDs
Journal of Alcoholism, Drug Abuse & Substance Dependence
Journal of Allergy Disorders & Therapy
Journal of Alternative, Complementary & Integrative Medicine
Journal of Alzheimer's & Neurodegenerative Diseases
Journal of Angiology & Vascular Surgery
Journal of Animal Research & Veterinary Science
Archives of Zoological Studies
Archives of Urology
Journal of Atmospheric & Earth-Sciences
Journal of Aquaculture & Fisheries
Journal of Biotech Research & Biochemistry
Journal of Brain & Neuroscience Research
Journal of Cancer Biology & Treatment
Journal of Cardiology: Study & Research
Journal of Cell Biology & Cell Metabolism
Journal of Clinical Dermatology & Therapy
Journal of Clinical Immunology & Immunotherapy
Journal of Clinical Studies & Medical Case Reports
Journal of Community Medicine & Public Health Care
Current Trends: Medical & Biological Engineering
Journal of Cytology & Tissue Biology
Journal of Dentistry: Oral Health & Cosmesis
Journal of Diabetes & Metabolic Disorders
Journal of Dairy Research & Technology
Journal of Emergency Medicine Trauma & Surgical Care
Journal of Environmental Science: Current Research
Journal of Food Science & Nutrition
Journal of Forensic, Legal & Investigative Sciences
Journal of Gastroenterology & Hepatology Research
Journal of Gerontology & Geriatric Medicine
Journal of Genetics & Genomic Sciences
Journal of Hematology, Blood Transfusion & Disorders
Journal of Human Endocrinology
Journal of Hospice & Palliative Medical Care
Journal of Internal Medicine & Primary Healthcare
Journal of Infectious & Non Infectious Diseases
Journal of Light & Laser: Current Trends
Journal of Modern Chemical Sciences
Journal of Medicine: Study & Research
Journal of Nanotechnology: Nanomedicine & Nanobiotechnology
Journal of Neonatology & Clinical Pediatrics
Journal of Nephrology & Renal Therapy
Journal of Non Invasive Vascular Investigation
Journal of Nuclear Medicine, Radiology & Radiation Therapy
Journal of Obesity & Weight Loss
Journal of Orthopedic Research & Physiotherapy
Journal of Otolaryngology, Head & Neck Surgery
Journal of Protein Research & Bioinformatics
Journal of Pathology Clinical & Medical Research
Journal of Pharmacology, Pharmaceutics & Pharmacovigilance
Journal of Physical Medicine, Rehabilitation & Disabilities
Journal of Plant Science: Current Research
Journal of Psychiatry, Depression & Anxiety
Journal of Pulmonary Medicine & Respiratory Research
Journal of Practical & Professional Nursing
Journal of Reproductive Medicine, Gynaecology & Obstetrics
Journal of Stem Cells Research, Development & Therapy
Journal of Surgery: Current Trends & Innovations
Journal of Toxicology: Current Research
Journal of Translational Science and Research
Trends in Anatomy & Physiology
Journal of Vaccines Research & Vaccination
Journal of Virology & Antivirals
Archives of Surgery and Surgical Education
Sports Medicine and Injury Care Journal
International Journal of Case Reports and Therapeutic Studies

Submit Your Manuscript: <http://www.heraldopenaccess.us/Online-Submission.php>