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# Unusual presentation of primary myelofibrosis with spontaneous bleeding after laparoscopic adrenalectomy: A case report

Sara Colozzi<sup>a,\*</sup>, Benedetta Costantini<sup>b</sup>, Serena Rupoli<sup>b</sup>, Attilio Olivieri<sup>b</sup>, Monica Ortenzi<sup>a</sup>, Mario Guerrieri<sup>a</sup>

<sup>a</sup> Clinica Chirurgica Generale e d'Urgenza, AOU Umberto I-Lancisi-Salesi, Ancona, Italy

<sup>b</sup> Clinica di Ematologia, AOU Umberto I-Lancisi-Salesi, Ancona, Italy

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## ABSTRACT

**INTRODUCTION:** We described unusual presentation of primary myelofibrosis with spontaneous bleeding after laparoscopic adrenalectomy. This case is written following the SCARE criteria.

**PRESENTATION OF CASE:** A 73 years old Caucasian man underwent laparoscopic right adrenalectomy for a rapidly increasing expansive mass (3.5 × 2.5 cm) of the right adrenal gland. The night of intervention, patient underwent urgent explorative laparotomy, that highlighted massive haemoperitoneum, clots in the abdomen, on the splenic side, on Morrison's space, and at the confluence between right renal vein and inferior vena cava.

Surgical specimen examination showed extra-medullary myeloid proliferation of the right perirenal tissue.

**DISCUSSION:** Bone marrow biopsy was performed. The list of differential diagnoses included: chronic myelomonocytic leukaemia, atypical CML and primary myelofibrosis (PMF). After discharge, several results became available: conventional cytogenetics was normal, PDGFR- $\alpha$ , PDGFR- $\beta$  and FGFR1 mutations were negative but V617 F mutation of the JAK2 gene was positive. Therefore, the final diagnosis was pre-fibrotic primary myelofibrosis according to the 2016 WHO classification, Dynamic International Prognostic Scoring System (DIPSS) plus 2 (intermediate-2).

**CONCLUSION:** To our knowledge, we report uncommon case of primary myelofibrosis associated with extra-medullary myeloid proliferation of the right perirenal tissue.

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## 1. Introduction

Primary myelofibrosis (PMF) is a BCR-ABL1-negative myeloproliferative neoplasm characterized by anaemia, progressive splenomegaly, extramedullary haematopoiesis, bone marrow fibrosis, constitutional symptoms, leukaemic progression, and shortened survival. Constitutive activation of the Janus kinase/signal transducers and activators of transcription (JAK-STAT) pathway, and other cellular pathways downstream, lead to myeloproliferation, pro-inflammatory cytokine expression, and bone marrow remodelling [1]. Clinical presentation of this neoplasm includes typical and atypical localizations; in fact, it is found not only in bone marrow and spleen but also in extramedullary sites, soft tissues, gastrointestinal tract, genitourinary tract and others [2,3].

This case is written following the SCARE criteria [4].

## 2. Presentation of case

We described unusual presentation of PMF with spontaneous bleeding after laparoscopic adrenalectomy.

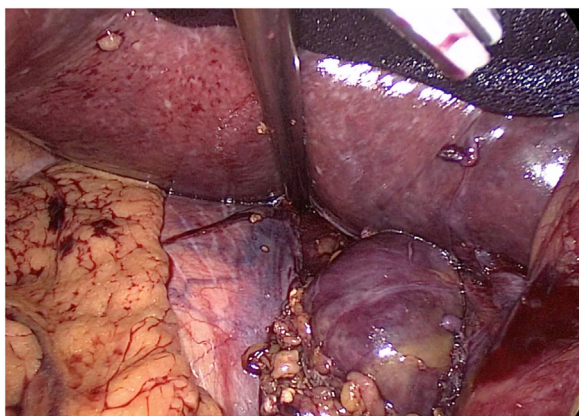
A 73 years old Caucasian man underwent laparoscopic right adrenalectomy for a rapidly increasing expansive mass (3.5 × 2.5 cm) of the right adrenal gland. It was an incidental finding on a computed tomography (CT) scan performed for dyspnoea. In the previous two years, the patient had a magnetic resonance (MR) and a positron emission tomography (PET) showing an increasing lesion with morphological features suggestive for adenoma, pheochromocytoma, tuberculosis (TB), and an enlarged spleen. Blood tests excluded secreting lesion or TB infection.

The list of comorbidities included hypertension on treatment with beta-blockers, benign prostatic hypertrophy treated with silodosin, chronic obstructive pulmonary disease (COPD) treated with steroids inhalers, hypothyroidism, previous appendectomy, unspecified previous surgery for head trauma.

An informed consent was obtained before surgery. According to standard procedure adopted in our Center for laparoscopic adrenalectomy [5,6], an open technique was used for laparoscopic

\* Corresponding author.

E-mail address: [sara.colozzi@libero.it](mailto:sara.colozzi@libero.it) (S. Colozzi).



**Fig. 1.** Intraoperative visualization of periadrenal mass.

access. Abdominal cavity exploration showed blood and clots on the surface of great omentum, in absence of any source of bleeding. After trocars placement, liver was retracted and posterior peritoneum was divided on the right side of the inferior vena cava to identify the adrenal gland (Fig. 1). After coagulation of adrenal vein, gland was dissected from its attachments and removed in a specimen retrieval bag. Haemostasis was accurately controlled, two drains were placed on the side of adrenal excision and on the splenic side, respectively, at the end of procedure.

After surgery, the patient was admitted on post-operative intensive care unit for parameters monitoring, due to COPD. The night of intervention the patient developed hypotension not responsive to fluids administration, tachycardia, anaemia not responsive to blood transfusions. Haematic fluid was present in drains bag. The patient underwent urgent explorative laparotomy, that highlighted massive haemoperitoneum, clots in the abdomen, on the splenic side, on Morrison's space, and at the confluence between right renal vein and inferior vena cava. No bleeding source was found. A lavage was performed with hot fluids and haemostatics were placed on the side of previous surgery. Two drains were placed under the liver and on the right adrenalectomy site.

First and second intervention were performed by the same surgeon (M.G.).

In the post-operative period, we observed a progressively increase of white blood cells count (WBC), until  $53.000/\text{mm}^3$  with basophilia, monocytosis and thrombocytosis, in the absence of any foci of infection; haemoglobin (Hb) was normal and blood transfusion was unnecessary. Clotting screening was normal. Spontaneous bleeding on abdominal wall started after the second surgery and required arteriography and embolization.

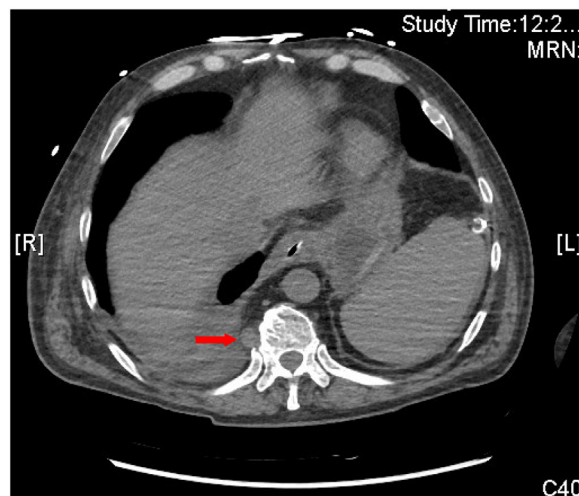
Due to an increase of WBC in the absence of signs and symptoms of infection, the patient was referred to haematology.

### 3. Discussion

The main differential diagnosis was chronic myeloid leukaemia (CML), but BCR-ABL1 mutation was absent (all transcripts were tested: *p190*, *p210*, *p230*). Following this result, a bone marrow biopsy was performed and it showed features compatible with a chronic myeloproliferative neoplasm, without excess of blasts, associated with extra-medullary myeloid proliferation of the right periadrenal tissue. A postoperative CT-scan confirmed extramedullary localization (Fig. 2).

The list of differential diagnoses included: chronic myelomonocytic leukaemia, atypical CML and primary myelofibrosis (PMF).

The patient was discharged from hospital 25 days after surgery.



**Fig. 2.** Paravertebral localization of extramedullary neoplasm.

After discharge, several results became available: conventional cytogenetics was normal, PDGFR- $\alpha$ , PDGFR- $\beta$  and FGFR1 mutations were negative but V617F mutation of the JAK2 gene was positive. Therefore, the final diagnosis was pre-fibrotic primary myelofibrosis according to the 2016 WHO classification [7], Dynamic International Prognostic Scoring System (DIPSS) plus 2 (intermediate-2) [8].

The patient is now being treated with hydroxycarbamide, to slowly reduce the leukocytosis, and erythropoietin  $\alpha$  40.000 units per week. As soon as the leukocytosis will settle, he will be considered for ruxolitinib.

### 4. Conclusion

To our knowledge, we report uncommon case of primary myelofibrosis associated with extra-medullary myeloid proliferation of the right periadrenal tissue.

### Declaration of Competing Interest

All authors declare no conflict of interest.

### Funding

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### Ethical approval

Ethical approval was not required.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of written consent is available for review by the editor-in-chief of this journal on request.

### Author contribution

Sara Colozzi and Benedetta Costantini: data collection and analysis; writing paper.

Serena Ruppoli, Attilio Olivieri, Monica Ortenzi and Mario Guerrieri: case management.

Mario Guerrieri: revision.

**Registration of research studies**

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