

## Case Report

# Splenic Rupture Secondary to Metastatic Undifferentiated Pleomorphic Sarcoma of the Spleen: A Rare Presentation of an Exceptionally Rare Entity

Maher Darwish<sup>a</sup> Ahmad AlAbdulkareem<sup>b</sup> Fawaz AlAbdulkareem<sup>c</sup>

<sup>a</sup>Department of Medicine, Mubarak AlKabeer Hospital, Jabriya, Kuwait; <sup>b</sup>Kuwait Institute for Medical Specializations, Ministry of Health, Andalous, Kuwait; <sup>c</sup>Faculty of Medicine, Kuwait University, Jabriya, Kuwait

## Keywords

Cancer · Spleen · Rare · Sarcomas

## Abstract

Splenic malignancies are uncommonly encountered in routine practice, and they are often only detected incidentally, posing a clinical challenge. Primary malignant neoplasms of the spleen are rare, and sarcomas are by far the rarest. The undifferentiated pleomorphic sarcoma type is an exceptionally rare and aggressive type with only a handful of cases reported in literature thus far. Herein, we present the case of a 49-year-old, with a previous finding of a heterogeneous splenomegaly on imaging for an unrelated medical complaint, presenting with an acute abdomen secondary to a ruptured spleen. The diagnosis of high-grade primary undifferentiated pleomorphic sarcoma of the spleen was made, and with further investigations, suspicious hepatic lesions were identified and later confirmed to be metastases.

© 2022 The Author(s).

Published by S. Karger AG, Basel

## Introduction

Primary splenic tumours are uncommon, and those of soft tissue origin are by far the rarest. Therefore, they are usually identified after investigations for unrelated medical complaints and are often only diagnosed retrospectively after a splenectomy. Only a few cases

of undifferentiated pleomorphic sarcoma of the spleen have been reported in literature [1]. Splenic rupture, a rare albeit potentially life-threatening cause of an acute abdomen, is a rare complication of malignant pathology. Herein, we describe a patient presenting with an acute abdomen due to a ruptured spleen, secondary to high-grade primary undifferentiated pleomorphic sarcoma of the spleen.

### Case

A 49-year-old Lebanese patient with a previous history of pleurodesis 4 years prior for a right pneumothorax presented with right-sided posterior paraspinal pain unresponsive to analgesics. Laboratory and radiographic investigations at the time were only notable for thrombocytopenia ( $103 \times 10^9/L$ ), for which blood film showed a picture of true thrombocytopenia, with large forms and activated lymphocytosis. Extended virology screen was normal. CT scan of his chest, abdomen, and pelvis was done and showed a peripherally located and regularly bordered lesion ( $1.3 \times 1.2 \times 2$  cm) of homogenous soft tissue density and evident contrast enhancement anteriorly at the right apex abutting the pleura and right subclavian artery. It also revealed heterogeneous splenomegaly (craniocaudal view: 16 cm) with distorted morphology due to variably sized hypoenhancing mass lesions, the largest ones located at upper ( $11 \times 9 \times 10$  cm) and lower ( $9.5 \times 8 \times 9$  cm) poles protruding out of surface. A positron emission tomography scan showed no hyperactive splenic foci, a mildly hypermetabolic right upper pulmonary activity corresponding to the previously noted lesion, and a markedly hypermetabolic activity at the proximal ascending colon, which was assessed by a colonoscopy and biopsy showed tubular adenoma of low-grade dysplasia. The patient at the time refused further investigations. Eight months later, he suffered a syncopal attack and presented with an acute abdomen and a drop in haemoglobin (7.7 g/L); CT showed splenic rupture, and he was managed conservatively. A bone marrow biopsy later revealed a hypercellular picture and a positive JAK-2 mutation. Elective splenectomy was done 3 months later (Fig. 1, 2).

Histopathological assessment was difficult to ascertain; the dominant mass showed non-distinctive appearances characterized by a discohesive proliferation of cytologically malignant pleomorphic epithelioid, polygonal or spindle-shaped cells, in some areas having copious eosinophilic cytoplasm and bizarre vesicular nuclei. The sample was then sent overseas for an external opinion; stains were negative for CD34, ERG, pan-keratin, AE1/AE3, desmin, CD68, CD21, CD31, ERG, D2-40, and HHV8. PU.1 and CD163 were positive only in non-neoplastic histiocytes. In the smaller mass from the splenic hilum, there was



Fig. 1. Intra-operative view of the spleen.



**Fig. 2.** Post-operative view of the resected spleen, highlighting the heterogeneous splenomegaly.

only some multifocal positivity for CD34. The splenic neoplasm was subsequently labelled as a primary undifferentiated pleomorphic malignant epithelioid and spindle cell sarcoma, high grade.

Three months post-splenectomy, a repeat CT showed suspicious, likely metastatic hepatic lesions within an enlarged liver (craniocaudal view: 18 cm). The patient was then treated with three cycles of doxorubicin and ifosfamide chemotherapy. A follow-up CT showed interval progression in his hepatic lesions, and the patient subsequently had microwave ablation for a right liver lesion and underwent a left hepatectomy which confirmed the metastatic nature of the lesions. Imaging 2 months post-hepatectomy, unfortunately, revealed newly developed hepatic lesions, suggestive of further metastases. The patient, currently more than 2 years since his initial presentation, undertakes regular outpatient follow-up at the Kuwait Cancer Control Center.

## Discussion

The vast majority of splenic neoplastic lesions are either secondary malignancies or metastases. Primary splenic lesions are uncommon, most are benign, and as such, primary splenic malignancies are rarely encountered in routine clinical practice. Among the primary splenic malignant diseases, vascular neoplasms are by far the most common, while sarcomas are the rarest [1]. Due to the infrequency of primary splenic sarcomas, their epidemiology is largely unknown and is mostly dependant on case reports. In the English literature, there have been less than 20 reported cases of undifferentiated pleomorphic sarcomas of the spleen, an extremely rare and aggressive primary splenic neoplasm. However, cases reported prior to 2000, before the criteria for histiocytic and dendritic cell sarcomas were established, may not truly represent undifferentiated pleomorphic sarcoma [2].

Undifferentiated pleomorphic sarcomas do not exhibit any morphological or immunohistochemical features characteristic of a specific line of soft-tissue differentiation. They tend to contain spindle cells with a storiform growth pattern, and show at least focal CD68 positivity; features that overlap but are not perfectly consistent with the findings we report herein [2, 3]. While our patient presented with an acute abdomen, a surgical emergency, these neoplasms may remain asymptomatic or present with constitutional and non-specific symptoms, complicating timely recognition and worsening the prognosis of an especially aggressive neoplasm. Additionally, these tumours exhibit the propensity for early invasion and distant metastases, as is the case in our patient, posing a significant clinical challenge. Indeed most cases reported thus far have a survival rate of less than 2 years, on par with pancreatic cancer, a notoriously aggressive tumour [2, 4].

In view of its rarity, the clinical behaviour and therapeutic responses of undifferentiated pleomorphic sarcomas of the spleen are yet to be determined. Surgical management is the treatment of choice, with splenectomies including laparoscopic splenectomies achieving negative histological margins. Chemotherapy and radiotherapy are treatment modalities that may be utilized in patients with advance disease and/or metastases, but their therapeutic effect is questionable and is poorly characterized [5]. Overall, the scarcity of available evidence, especially in relation to therapeutic options both surgical and pharmacological, compounded with the highly invasive nature of the disease with early and distant metastases, translates into dismal prognosis and this necessitates urgent attention.

## Conclusion

Primary splenic tumours are exceedingly rare, and the undifferentiated pleomorphic sarcoma type is an exceptionally rare and aggressive neoplasm. While the rarity of the disease complicates recognition, and despite the fact that little is known about the epidemiology including incidence, prevalence, and relevant risk factors, a degree of suspicion is warranted, especially in patients with persistent haematological abnormalities and/or abdominal complaints.

## Statement of Ethics statement

Written informed consent for the publication of the case including clinical photographs has been obtained from the patient. Institutional ethical approval is not required for this study in accordance with local or national guidelines.

## Conflict of Interest Statement

The authors have no conflicts of interest to declare.

## Funding Sources

This research received no grant from any funding agency in the public, commercial, or not-for-profit sectors.

## Author Contributions

Dr. Maher Darwish conceived and designed the paper. Dr. Ahmad AlAbdulkareem and Mr. Fawaz AlAbdulkareem were involved in data collection, both clinical and results of investigations done, and prepared and drafted the manuscript. Mr. Fawaz AlAbdulkareem retrieved and included the clinical photographs. Dr. Maher Darwish and Dr. Ahmad AlAbdulkareem edited, reviewed, and approved the manuscript. All authors read and approved the final manuscript.

## Data Availability Statement

All data generated or analysed during this study are included in this article. Further enquiries can be directed to the corresponding author.

## References

- 1 Ashmore DL, Dasgupta D. A splenic “cyst”: histology confirmed splenic sarcoma. *Ann R Coll Surg Engl*. 2020; 102(5):e105–6.
- 2 Farah LB, Chee M, Min Y, Siok C, Chien T. Primary splenic leiomyosarcoma as an exceptionally rare cause of ruptured splenomegaly: a case report and review of primary splenic sarcomas. *Hum Pathol Case Rep*. 2020; 22:200452.
- 3 Sieber SC, Lopez V, Rosai J, Buckley PJ. Primary tumor of spleen with morphologic features of malignant fibrous histiocytoma immunohistochemical evidence for a macrophage origin. *Am J Surg Pathol*. 1990;14(11): 1061–70.
- 4 Hashmi A, Podgaetz E, Richards ML. Laparoscopic resection of an undifferentiated pleomorphic splenic sarcoma. *JLS*. 2010;14(3):426–30.
- 5 Dawson L, Gupta O, Garg K. Malignant fibrous histiocytoma of the spleen: an extremely rare entity. *J Cancer Res Ther*. 2012;8(1):117.