Case Report

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Splenic Burst Secondary to Low-Grade Angiosarcoma with Fatal Prognosis: Aggressive Evolution and Multiple Metastases: Case Report

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ABSTRACT

Introduction: *Angiosarcoma is a rare endothelial malignant neoplasm with variable clinical behavior, predominantly aggressive, originating in endothelial cells. Its presentation in the chest wall is unusual and its diagnosis is often complex, and it can evolve into serious complications such as metastatic splenic rupture.*

Clinical case: *We present the case of a 47-year-old male patient who presented with a tumor in the left anterior rib cage. The first biopsy was inconclusive. The clinical evolution was accelerated, presenting from persistent bleeding from the surgical wound and subsequent massive hemoperitoneum secondary to a grade V splenic lesion due to splenic metastasis, where multiple metastatic lesions were also evident in the liver, intestine and peritoneum. An extension study such as computed tomography of the skull, thorax, abdomen and pelvis was performed, where metastatic lesions were evident, as well as in the skin, brain and lung. Patient who also presented severe systemic deterioration. Histopathological examination of the splenic specimen revealed vascular neoplasia compatible with low-grade angiosarcoma.*

Discussion: *This case underscores the rarity of the initial presentation, the biological aggressiveness of angiosarcoma even in low-grade forms, its capacity for hematogenous dissemination with an extensive metastatic pattern, and the inherent difficulties in managing surgical complications in cancer patients. Metastatic splenic rupture, although uncommon, represents a true surgical emergency requiring an immediate and aggressive multidisciplinary approach, even when the overall prognosis is guarded.*

Conclusion: *Splenic rupture due to metastatic angiosarcoma represents a serious complication in the course of advanced disease. This case highlights the importance of maintaining a high clinical suspicion in the presence of rapidly growing thoracic masses and reaffirms the value of damage control surgery as a palliative strategy in selected acute settings.*

Keywords: *angiosarcoma, low grade, metastasis, spleen, hemoperitoneum*

Introduction

Angiosarcoma is a malignant tumor of endothelial origin, characterized by a proliferation of atypical vascular cells that form irregular vascular structures. This entity was first described in the medical literature in the late 19th century. The term "angiosarcoma" was coined by the German pathologist Dr. Rudolf Virchow, who laid the foundations of modern pathology and made fundamental contributions to the understanding of vascular tumors [1]. The first reported cases were predominantly associated with the liver and skin, with a particular incidence observed in workers exposed to vinyl chloride and chronic arsenicalism, which allowed us to identify some of the first environmental risk factors associated with its development [2].

It represents approximately 1–2% of all soft tissue sarcomas [3]. Its annual incidence is estimated at 0.25 cases per 100,000 inhabitants, with a predilection for the scalp, skin, liver, breast, and deep soft tissues, and, to a lesser extent, solid organs such as the liver, spleen, lung, and heart; although it can affect any location in the body [4]. Primary presentation in the chest wall is particularly uncommon and often manifests as a painless, progressively growing mass [5].

The clinical behavior of angiosarcoma is highly variable, but it is generally associated with high aggressiveness, a marked tendency toward hematogenous dissemination, and a short overall survival rate[6]. The low-grade variant is often interpreted as having a better prognosis; however, several reports have described that even in these cases, an accelerated clinical course and a high degree of metastasis can be observed [7].

Splenic angiosarcoma is one of the rarest and most serious presentations among metastatic neoplasms, with fewer than 200 cases described in the literature [8]. It usually manifests clinically as abdominal pain, splenomegaly, and occasionally hemoperitoneum secondary to spontaneous rupture of the spleen; an entity traditionally associated with lymphoproliferative hematologic disorders and trauma [9]. This complication may be the first manifestation of the disease, requiring immediate surgical intervention [10].

This case is unique for three reasons: (1) the atypical initial presentation as a rib tumor, the biopsy of which was inconclusive; (2) the rapid clinical progression to a state of multiple metastases (liver, peritoneum, skin, lung, brain, and bone) complicated by hemorrhagic splenic injury; and (3) the histopathological finding of low-grade angiosarcoma, with an aggressive clinical course and fatal within a few months. This report seeks to contribute to the medical literature with an illustrative example of the unpredictable nature of angiosarcoma, even in its histologically less aggressive form.

Case Report

A 47-year-old male patient, originally from Caracas, with a personal history of smoking (6 cigarettes/day, pack-year index of 1.8) and a known allergy to penicillins, with no history of cancer. He presented with progressive enlargement of the left anterior chest wall, which had been present for 3 months. His only surgical history was varicocele surgery 15 years prior, which was undocumented. After initial imaging studies, he was referred to the Thoracic Surgery Service, where a left Chamberlain thoracotomy was performed with a biopsy of the lesion, the result of which was inconclusive. Subsequently, in the days following surgery, the patient presented bleeding through the surgical wound, which led to hospital admission by the Thoracic Surgery Service for management of complications.

Suspecting multiple metastatic disease, he was transferred to the Internal Medicine Department for evaluation and comprehensive management. During his stay, his general condition, abdominal distension, and initial signs of hemodynamic instability were evident. Diagnostic and therapeutic paracentesis was performed, which revealed the presence of hemoperitoneum, prompting an urgent consultation with the General Surgery Department.

On physical examination, the patient was in poor general condition, pale, hemodynamically unstable (BP: 90/70 mmHg; HR: 135 bpm; RR: 26 rpm; SatO2: 97% with a reservoir mask at 10 L/min), with capillary refill greater than 3 seconds. The chest showed asymmetry with hypoexpansibility, a thoracotomy wound with well-faced edges, with an underlying volume increase of 6x8 cm in the left anterior thoracic region and extensive perilesional ecchymosis. The abdomen was globular, distended, slightly depressible, painful on superficial and deep palpation, with a positive Gueneau de Mussy sign of peritoneal irritation. The neurological examination revealed drowsiness and partial disorientation.

The patient was taken to the operating table where an emergency exploratory laparotomy was performed. Massive hemoperitoneum (approximately 3000 cc) was found, revealing an enlarged spleen (10x10 cm) with grade V metastatic splenic rupture, as well as multiple nodular metastatic implants with a hemorrhagic appearance on the liver surface, intestinal loops, and parietal peritoneum. An exploratory laparotomy with splenectomy and abdominal drainage was performed.

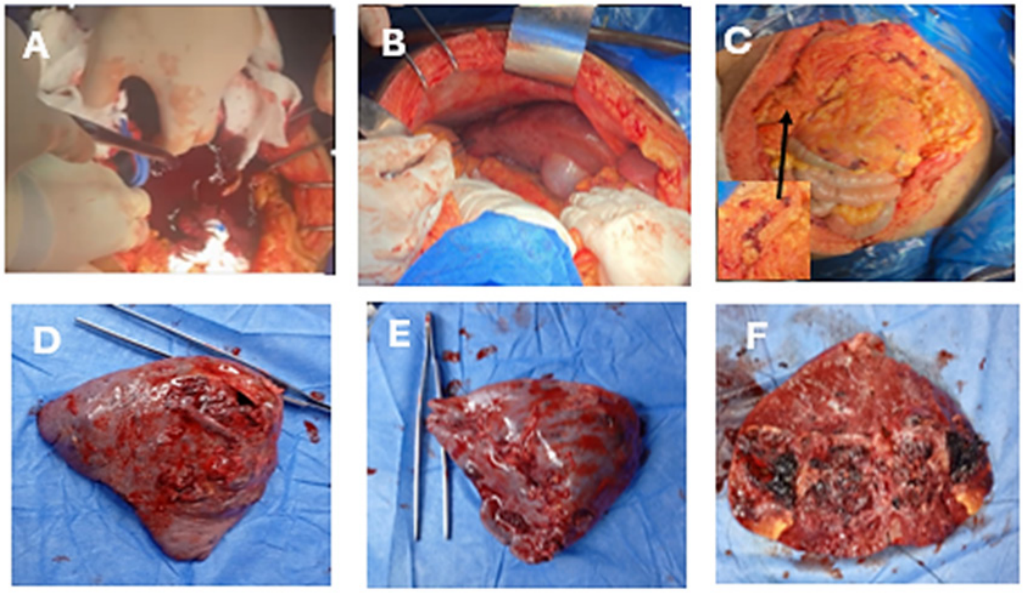

Figure 1. Exploratory laparotomy. *(A) Hemoperitoneum drainage; (B) Metastatic liver lesions; (C) Metastatic mesenteric and intestinal lesions; (D) Anterior surface of the spleen; (E) Posterior surface of the spleen; (F) Inferior surface of the spleen (base of the lesion ruptured).*

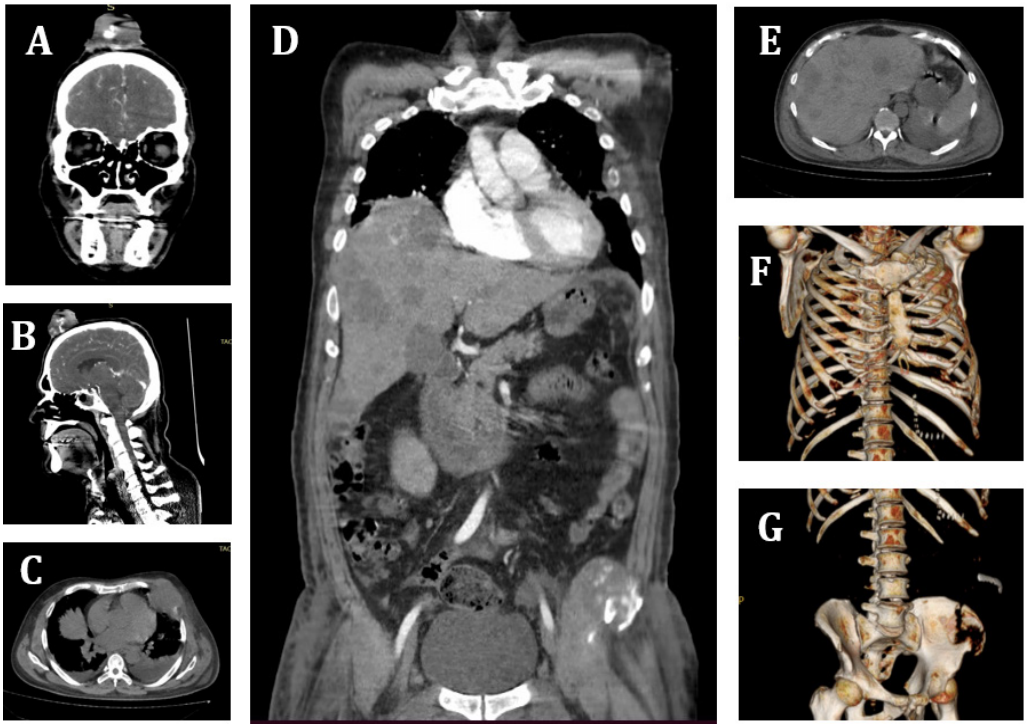

Figure 2. Cephalocaudal computed tomography. *Brain, coronal (A) and sagittal (B) sections show frontal and parietal subcutaneous masses, multiple homogeneously enhancing cerebral and cerebellar metastases and perilesional finger-like edema. Thorax and abdomen coronal (C, E) sections show multiple hypoenhancing liver masses with peripheral enhancement, solid right renal lesion measuring 7.3 x 6.2 cm, extensive lytic bone metastases with pathological fracture of T4. sagittal (D) section shows multiple diffuse solid nodular metastatic lesions in bilateral lung parenchyma, bilateral pleural effusion, primary mass measuring 6.5 x 6.2 cm in the left 4th intercostal space with extension to the pectoral muscles. Bone reconstruction (F, G) shows extensive lytic bone metastases with pathological fracture of T4 and left iliac crest.*

Due to severe anemia (Hb 6 g/dL) and hypovolemic shock, he remained in intensive care under mechanical ventilation and vasoactive support for 8 days, with transient clinical improvement that allowed the withdrawal of vasopressors and extubation. After initial stabilization, the patient was transferred to the hospital ward where multiple ulcerated skin lesions on the scalp were evident, an increase in volume in the left pectoral region measuring 10 × 10 cm, adhered to deep planes, grade III edema in the extremities and surgical wounds with persistent serous discharge, sustained functional deterioration and refractory oncological pain located in the dorsal region and lower extremities.

Cephalocaudal computed tomography with intravenous contrast revealed extensive disease dissemination. Findings included: frontal and parietal subcutaneous masses, and multiple cerebral and cerebellar metastases with homogeneous enhancement and perilesional finger-like edema, multiple diffuse solid nodular metastatic lesions in bilateral lung parenchyma, bilateral pleural effusion, a 6.5 x 6.2 cm primary mass in the left 4th intercostal space with extension to the pectoral muscles, multiple hypoenhancing liver masses with peripheral enhancement, a 7.3 x 6.2 cm solid right renal lesion, extensive lytic bone metastases with pathological fracture of T4 and left iliac crest.

Histopathological examination of the splenic specimen showed atypical vascular proliferation with irregular spaces lined by endothelial cells, focal necrosis, and hemorrhagic areas, concluding a low-grade angiosarcoma. Immunohistochemistry revealed positivity for CD31, CD34, and FLI-1, confirming the endothelial origin of the neoplasm and establishing the definitive diagnosis of low-grade angiosarcoma.

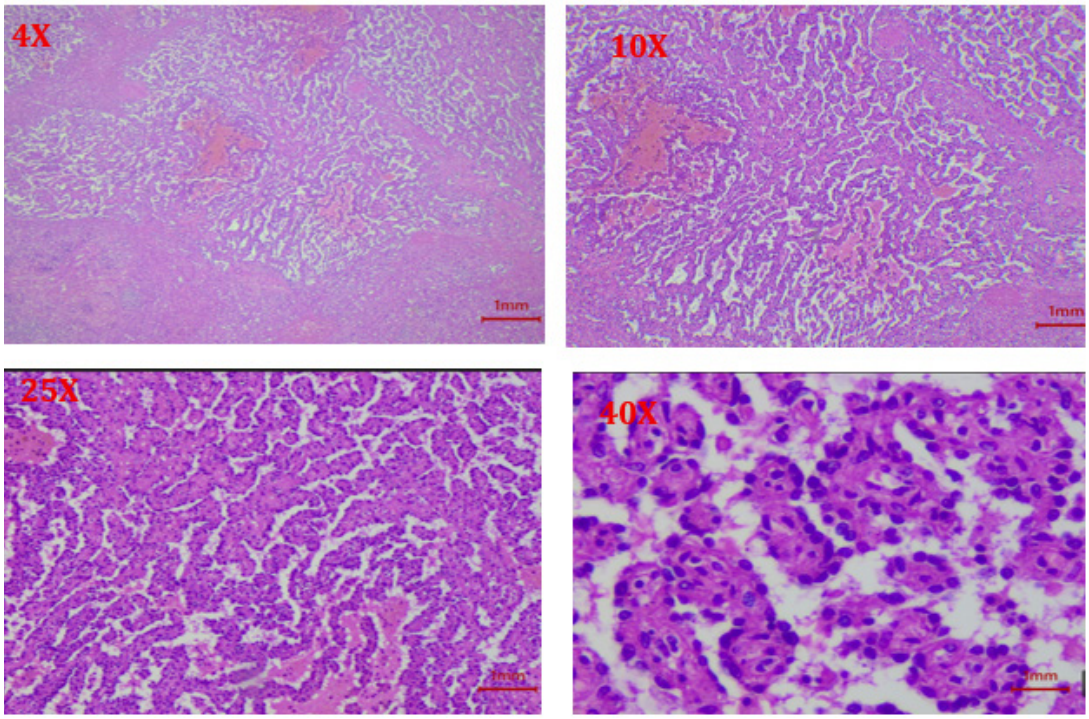

Figure 3. Histological study. Hematologic examination. *Irregular vascular proliferation infiltrating the stroma is evident, composed of anastomotic channels lined by endothelial cells with mild to moderate nuclear atypia and little mitotic activity. No significant areas of hemorrhage or necrosis are observed.*

Following initial postoperative stabilization, the patient was managed with a multidisciplinary approach that included the Oncology, Pain Medicine, and Palliative Care services. A stepped analgesic regimen was implemented, including major opioids (intravenous morphine) along with nonsteroidal anti-inflammatory drugs and neuromodulators for the control of refractory cancer pain. In parallel, advanced nutritional support measures and management of neuropathic symptoms secondary to the documented bone metastases were instituted.

Despite comprehensive management, the patient's general condition progressively deteriorated, with worsening multi-organ dysfunction, septic symptoms, and progression of brain metastases, leading to irreversible neurological deterioration. Management was reoriented exclusively toward palliative care, prioritizing the patient's comfort and dignity. Regrettably, the patient died three weeks after surgery as a result of multi-organ failure due to unresectable metastatic disease.

Discussion

This case provides clinical evidence of the remarkable biological aggressiveness and unpredictable evolution of angiosarcoma, a neoplasm that, despite its low incidence, is characterized by a high metastatic potential and a marked tendency toward locoregional recurrence, even after wide surgical resections and conventional oncological management [11]. Although the primary location in the chest wall is unusual, cases describing its appearance in deep soft tissues have been documented [12]. Visceral angiosarcoma, particularly splenic angiosarcoma, represents an extremely rare neoplasm with an unfavorable prognosis [13]. In the series published by Kanno et al., the median survival time was less than 12 months after diagnosis [5,6,11].

The literature suggests that although some tumors can be classified as low histological grade, this does not exclude aggressive clinical behavior [14]. This finding is consistent with our case, where the patient presented with an accelerated clinical course, multiple visceral and cutaneous metastases, and a fatal outcome within a short period.

The diagnosis of splenic rupture in cancer patients poses a considerable challenge, given that its manifestations can be confused with other causes of acute abdomen [15]. In this case, diagnostic paracentesis was instrumental in confirming hemoperitoneum, allowing for timely surgical intervention.

Hemoperitoneum secondary to splenic rupture is a rare, feared, and frequently fatal complication associated with splenic angiosarcoma [16]. In this patient, intra-abdominal bleeding reached approximately 3000 cc, requiring emergency splenectomy as the only viable option to control active bleeding, as well as a massive transfusion protocol. The procedure was based on the principles of damage control surgery, whose priority objective is the physiological stabilization of the patient rather than the definitive resolution of the oncological disease [17]. In this context, the intervention was essentially palliative in nature, given the advanced stage of the disease and the unresectability of the metastatic lesions. Despite surgical intervention and intensive support, the disease continued to progress, reflecting the limited efficacy of conventional management in this setting [18].

Therapeutic options for angiosarcoma include surgery, chemotherapy (doxorubicin, paclitaxel), radiotherapy, and, more recently, targeted and antiangiogenic therapies. However, the available evidence is scarce, and overall results remain poor. The diffuse and metastatic nature of this tumor, as observed in the case described, makes complete resection difficult and limits the efficacy of systemic treatments, relegating the therapeutic approach to patient comfort and dignity [19].

Another interesting aspect of this report is the initial thoracic presentation with a negative biopsy, which delayed definitive diagnosis. This fact highlights the need to repeat histopathological studies in suspicious vascular lesions and to maintain a high index of clinical suspicion.

Conclusion

In conclusion, this case underscores that low-grade angiosarcoma should not be interpreted as a benign entity. Despite its histology, it can exhibit clinical aggressiveness comparable to high-grade forms, with extensive metastatic spread and short-term fatal outcome.

Low-grade angiosarcoma can exhibit highly aggressive and lethal behavior. The initial costal location, the lack of diagnosis in the initial biopsy, and the extensive metastatic spread make this case exceptional. Early clinical suspicion and histopathological correlation are essential for identification.

Metastatic splenic rupture as the initial manifestation of chest wall angiosarcoma represents a serious, rare, and difficult-to-manage complication. This case highlights the clinical, surgical, and ethical challenges that arise in the management of oncologic emergencies in patients with advanced disease. Emergency splenectomy, although palliative, was a vital intervention to control bleeding and allow for temporary stabilization.

The report underscores the importance of maintaining a high level of clinical suspicion for rapidly growing chest masses and abdominal symptoms in cancer patients, as well as the need for informed and compassionate decision-making. Early integration of palliative care and a multidisciplinary approach are essential to ensuring patient-centered care, respecting their values and priorities in the context of an incurable disease.

Informed Consent

The authors declared that written informed consent was obtained from the patient for the publication of this case report and the images included within. .

Conflict of Interest and Funding

The author declares no conflict of interest and received no specific funding for this work.

Ethical Clearance

Not applicable

Use of Artificial Intelligence

The authors declared that no artificial intelligence (AI)-assisted technologies (such as large language models, chatbots, or image generators) were used in the preparation of this work.

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