CASE REPORT

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Multifocal Gastrointestinal Angiosarcoma: a Challenging Diagnosis?

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ABSTRACT

Angiosarcoma rarely involves the gastrointestinal tract. Herein, we describe the case of a 68-year-old man with haemoptysis and melena who was eventually diagnosed with multifocal angiosarcoma of the stomach, small bowel, lungs, and thyroid. The peculiarity was that the histological feature of the polypoid lesions removed at endoscopy was initially misinterpreted as benign hyperplastic polyps, whilst their neoplastic nature was clinically suspected only when the videocapsule endoscopy revealed the presence of multiple variable-sized nodules with apical erosion or active bleeding in the small bowel.
Based on the very low incidence, diagnosis of angiosarcoma involving the gastrointestinal tract may be misinterpreted by both the endoscopist and pathologist.

Key words: angiosarcoma - gastrointestinal tract - videocapsule endoscopy - endoscopy.

Abbreviations: GI: Gastrointestinal; GIST: Gastrointestinal stromal tumor; VCE: Videocapsule endoscopy.

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INTRODUCTION

Angiosarcoma is a rare neoplasm type that accounts for less than 2% of all sarcomas, which, in turn, are infrequent tumours [1]. Although they can develop anywhere in the body, angiosarcomas occur mainly in the skin, soft tissue and breast and rarely the gastrointestinal (GI) tract [1].

In the GI tract, angiosarcomas generally involve the liver and spleen, whilst a location in the small and large bowel has been rarely reported [1-8]. No standardized therapeutic approach is currently available for these sarcomas, and prognosis remains dismal. We report the case of an unsuspected diagnosis of multifocal haemangiosarcoma involving the stomach and small bowel, as well as the lung and thyroid.

CASE REPORT

A 68-year-old man was admitted to the Internal Medicine Unit because of recurrent haemoptysis episodes. He had worked for 40 years as a carpenter in the construction industry. Neither a family history of malignancy nor current diseases were stated. At entry, severe anaemia (haemoglobin: 6.8 g/dL) was detected, with a pulse of 124 beats/min, and blood pressure values of 110/70 mmHg, requiring transfusion of 4 units of packed red blood. Chest computed tomography was performed showing in both lungs multiple masses with central necrotic areas and peripheral ground glass aspect, associated with diffuse mediastinal lymphadenopaties suggesting a multifocal neoplasia (Fig. 1). In addition, two masses (3×4 cm and 5×3 cm) in the right lobe of the thyroid gland causing carotid and jugular vein dislocation were detected. Due to the presence of haemoptysis and the peripheral ground glass aspect of the lung lesions suggestive of a recent bleeding, a bronchial arterial angiography was performed confirming multiple nodular hypervascularized necrotic areas in both lungs, and revealed an arterio-venous fistula in the apical area of left lung which was embolized with Contour particles.

Two days later, the patient was referred to our Gastroenterological Unit because of melena onset. Upper endoscopy detected two adjacent polypoid bright-red, ulcerated lesions, with a diameter of 5 mm and 15 mm,

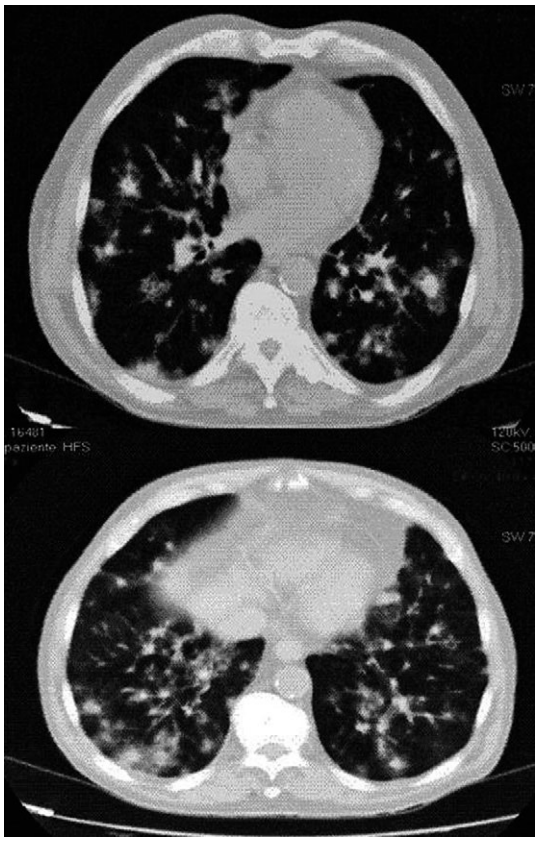

Fig. 1. Computed tomography showing multiple lung lesions.

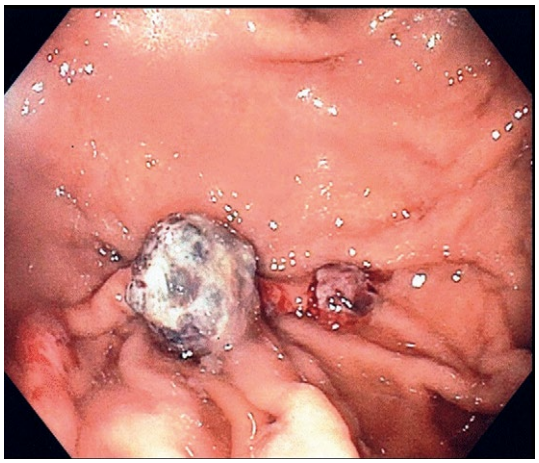

Fig. 2. Upper endoscopy showing ulcerated polypoid lesions in the stomach.

respectively, in the stomach (Fig. 2), as the potential cause of bleeding. A polypectomy was performed, and the histological assessment revealed a feature of hyperplastic polyps. However, melena persisted in the successive week, and haematochezia also manifested. A further gastroscopy was negative, whilst a colonoscopy revealed the presence of several clots in the lumen without active bleeding lesions. Therefore, a videocapsule endoscopy (VCE) was performed, revealing multiple variable-sized hyperaemic polypoid masses, with diameters varying from 3mm to 7mm, with apical erosions and signs of recent or active bleeding in the distal part of the small bowel (Fig. 3). Due to the suspected nature of intestinal nodules, with a macroscopic feature resembling that of previously removed polyps in the stomach, a histological re-assessment of the gastric polyp was solicited. The new histological examination with immunohistochemical staining revealed a haemangiosarcoma (Fig. 4). The patient died from haemorrhagic shock 75 days later, after receiving a total of 98 units packed erythrocytes, and a single dose of chemotherapy.

DISCUSSION

Angiosarcoma is a rare tumour, which infrequently involves the GI tract [1-9]. Both symptoms and endoscopic features of GI angiosarcoma are not specific, so that diagnosis is frequently unsuspected [2]. Bleeding episodes – either manifest or occult – as well as anaemia are not infrequent in patients with GI location of angiosarcoma due to its tendency to bleeding [2, 3], as occurred in our patient. A peculiarity of our case is that the initial histological assessment of the removed gastric polypoid lesions suggested benign, hyperplastic polyps. Therefore, it may occur that even an experienced pathologist could be unfamiliar with a GI angiosarcoma diagnosis due to its very low incidence. The neoplastic nature of gastric lesions was clinically suspected only when the VCE detected multiple variable-sized nodules with apical erosion or active bleeding in the small bowel. Indeed, at endoscopic observation, GI angiosarcomas may appear with different, unspecific features, ranging from flat bright-red

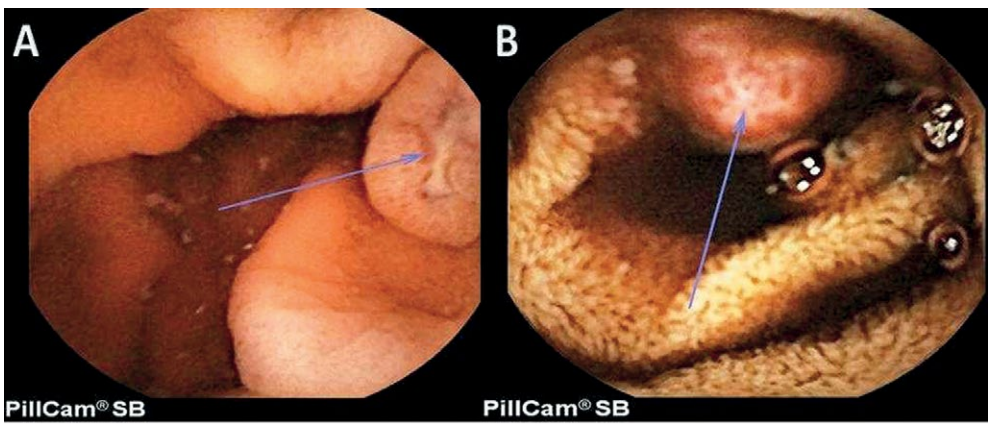

Fig. 3. Videocapsule endoscopy showing ulcerated nodules with fibrin (A) with active bleeding (B).

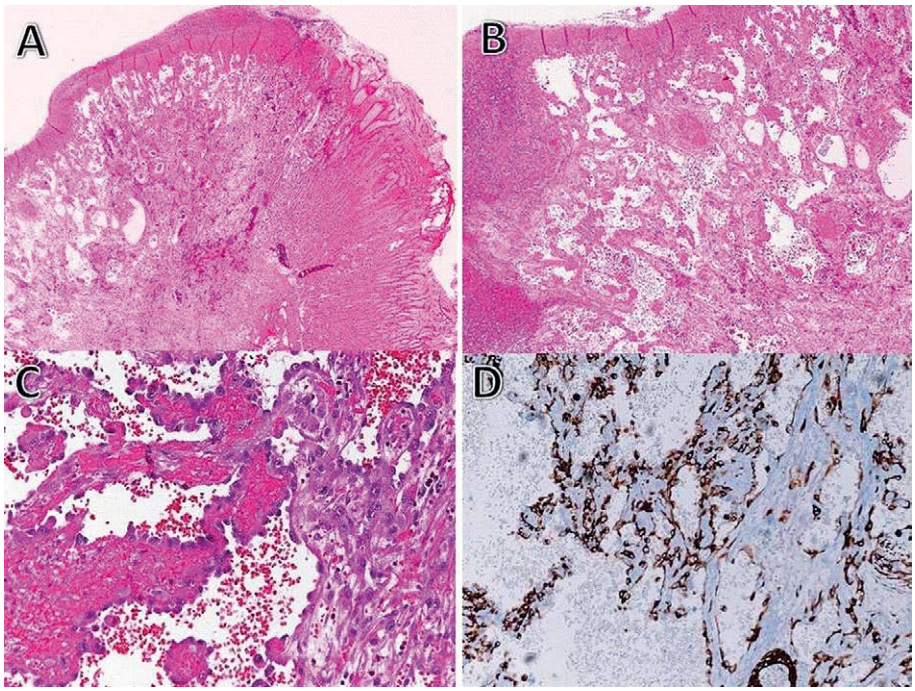

Fig. 4. A: Polypoid and ulcerated lesion of gastric mucosa (H&E, x20). B: The lesion is predominantly composed of anastomozing vascular channels of irregular size and shape, containing red blood cells (H&E, x40). C: The vascular spaces are lined by plump, atypical endothelial cells. A less differentiated, solid area is seen on the right (H&E, x200). D: Immunohistochemical staining for CD31, a sensitive and specific vascular marker (IHC, x100).

lesions, purpuric or haemorrhagic nodules, irregular masses or even as submucosal masses, so that differential diagnosis with other GI sarcomas or with GI stromal tumors (GISTs) is required [9, 10]. Endoscopists are generally untrained for a suspected angiosarcoma diagnosis. In our patient, masses were present also in the lung, thyroid, and mediastinal lymph nodes. Therefore, the initial origin of neoplasia remained unclear, the GI tract being the potential site of either primary or metastatic haemangiosarcomas [1, 6]. Of note, our patient had worked for a long-time as a carpenter in the construction industry, with exposure to toxic chemicals, such as another patient with multifocal angiosarcoma involving the GI tract described in Japan [8]. A professional exposure to vinyl chloride, thorium dioxide, or arsenic could have played a potential role [1, 7]. Unfortunately, a standardized therapeutic approach is not available for multifocal localization of angiosarcomas. The marked propensity to bleeding coupled with the very low susceptibility towards current chemotherapies account for the high aggressiveness of angiosarcomas, with an overall survival prognosis of less than 5 months.

CONCLUSION

We describe a rare case of unsuspected angiosarcoma with GI involvement, providing both gastric and intestinal features observed at upper endoscopy and VCE, respectively. Our case confirms the difficult diagnosis and the poor prognosis of this multifocal neoplasm in absence of a standardised therapeutic approach.

Conflicts of interest: None to declare.

Authors' contribution: V.D.F., A.Z. designed and wrote the report. A.B. and L.R. collected literature data. F.C. and A.P. performed histological assessment and evaluations.

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