Computed Tomography (CT) Angiography in Pre-Embolization Assessment of Location of Gastrointestinal Bleeding in Paediatric Patient with Granulomatosis with Polyangiitis (Wegener’s Granulomatosis) – Case Report

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Summary

Background: Acute gastrointestinal bleeding is an emergency with a high morbidity and mortality. Early diagnosis and appropriate intervention can be lifesaving and can prevent long-term complications.

Case Report: In this case report, we discuss and illustrate the role of CT angiography in the evaluation of acute, active gastrointestinal haemorrhage and show its usefulness prior to embolization. We describe a 15-year-old girl with granulomatosis with polyangiitis, formerly known as Wegener’s granulomatosis.

Conclusions: An accurate pre-embolization assessment of bleeding with CT angiography shortens the total diagnostic time, which results in prompt and more effective endovascular treatment.

We describe the clinical presentation of our patient and present diagnostic and interventional radiologic findings.

MeSH Keywords: Child • Embolization, Therapeutic • Emergencies • Gastrointestinal Diseases • Tomography, X-Ray Computed

Background

Granulomatosis with polyangiitis (GPA), formerly known as Wegener’s granulomatosis (WG), is an autoimmune systemic disease of an unknown aetiology. It was first described by Friedrich Wegener in 1936 and 1939 [1]. Currently, the eponym WG is no longer in use. During the International Chapel Hill Consensus Conference in 2012, WG was replaced by the more descriptive term GPA [2]. GPA is a rare disease, with an incidence of approximately 0.1: 100 000 in the paediatric population and a strong female and Caucasian predominance (63% and 69%, respectively) [3]. The median age at GPA diagnosis in the paediatric population is 14 years. When comparing paediatric patients to adults, children have similar clinical manifestations of GPA but a different frequency of organ involvement. Initial symptoms of GPA can vary widely; however, in children, the most common (between 80–91%) symptoms are ENT symptoms. Other symptoms of GPA are constitutional (malaise, fever, weight loss (89%), respiratory (79%), mucosal and dermatologic (64%), musculoskeletal (59%), and ophthalmologic (35%) [3]. The exact frequency of gastrointestinal (GI) symptoms in GPA is unknown, and it ranges between 16% and 42%, with the most frequent GI symptoms being abdominal pain, diarrhoea, and bleeding [3].

Acute GI bleeding is an emergency that requires fast detection and localization of the source of bleeding. The usefulness of CT angiography in diagnosing acute GI bleeding
has been assessed in many studies; however, most studies were conducted in adults [4–6]. The overall sensitivity and specificity of CT angiography for detecting active acute GI bleeding in adults is 85–89% and 85–96%, respectively [4–6]. The overall accuracy of CT angiography for detecting the cause of GI haemorrhage is approximately 80% [7]. CT angiography can detect flow rates as low as 0.3 mL/min, which is slightly lower than the threshold assigned to angiography (0.5 mL/min) [8].

In this case report, we discuss and illustrate the role of CT angiography in the evaluation of acute, active GI haemorrhage in a paediatric patient with GPA.

Case Report

A 15-year-old girl was admitted to the Department of Pulmonology, Allergology and Paediatrics, Medical University of Warsaw, due to a suspicion of diffuse alveolar haemorrhage. For the past 2 months, she presented with nasal congestion, temporary nasal bleeding, and then haemoptysis. She felt general malaise. She did not suffer from any chronic illnesses apart from Asperger’s syndrome.

On physical examination, she was pale, her heart rate was 150 beats/min, her respiratory rate was 40 breaths/min, and her oxygen saturation level was 91–97% on room air. Auscultation of her chest revealed crepitation. A skin examination revealed petechiae on the upper and lower extremities. Her haemoglobin was 6.5 g/dl, and her platelet count was 164×10⁹/l. A chest X-ray revealed massive bilateral diffuse opacities (Figure 1). She was diagnosed with GPA with an anti-neutrophil cytoplasmic autoantibodies (C-ANCA) titre of 1 in 1280 and evidence of necrosis (IgA, IgG, IgM, and C3 deposits in vessels were found in the skin biopsy). On the second day after admission, melena, haematuria, and proteinuria appeared. She was treated with methylprednisolone pulses (1.2 g per day), cyclophosphamide (every 2 weeks), and plasmapheresis (every other day). In spite of the treatment, one week after admission, the patient developed haemorrhagic shock due to massive GI bleeding. She underwent multiple transfusions (in total 18 units of blood packs and 23 units of cryoprecipitate) and bidirectional endoscopies. An upper GI endoscopy did not show any abnormal findings, whereas a limited colonoscopy was non-diagnostic due to a large volume of melena in the rectum and sigmoid colon.

CT angiography (CTDI 67.7 mGy) was performed in the Department of Paediatric Radiology, Medical University of Warsaw, to localize the site of GI bleeding. The scan included the entire abdomen and pelvis, from the diaphragm to the inferior pubic ramus. No oral contrast was administered.

Contrast medium (85 ml) was administered intravenously with a power injector at a rate of 4 mL/sec, followed by a chaser of saline (50 ml). Then, a multiphasic examination
Because of the very fast heart rate (due to hypovolemic shock), the pure arterial phase could not be performed. Contrast-enhanced scans were obtained manually with no ROIs and without using automated bolus triggering. The examination clearly demonstrated the origin and location of the bleeding. Extravasation of contrast was detected in one of the jejunal loops (Figure 2). A thrombus was also visualized in the inferior vena cava, just above the iliac veins. On the upper range scans, massive changes in the alveoli, suggestive of diffuse alveolar haemorrhage, were present (due to GPA) (Figure 3).

In spite of transfusions of packed red blood cells and fresh frozen plasma, the patient’s clinical state was deteriorating. Because there was a poor prognosis for further conservative or surgical treatment, the patient was moved to the interventional suite (2nd Department of Clinical Radiology, Medical University of Warsaw). Selective arteriography of the superior mesenteric artery (based on CT angiography) revealed intra-intestinal bleeding in the territory of the first branches of the superior mesenteric artery (Figure 4A). At the same time, embolization of the bleeding vessel was performed, using a 50% mixture of Histoacryl and Lipiodol. Angiographic control confirmed the effectiveness of the procedure (radiation dose 521.9 mGy) (Figure 4B). During the procedure, the patient received 4000 U of heparin. Subsequently, the patient was transferred to the intensive care unit, where further conservative treatment was applied. After 6 cycles of plasmaphereses, the patient’s clinical state improved. A follow-up chest CT scan (CTDI 3.4 mGy) revealed regression of the previously described massive changes in the alveoli, and the patient was discharged (Figure 5).

**Conclusions**

Acute GI bleeding is an emergency with a high morbidity and mortality. Early diagnosis and appropriate intervention can be lifesaving and can prevent long-term complications. CT angiography is a noninvasive and widely available diagnostic examination. Multiphasic imaging enables direct visualization of active bleeding in the GI tract. It can rapidly provide the referring physician with information on the source and location of an active GI haemorrhage, because the sensitivity of CT angiography for detecting active bleeding and its cause is very high. An accurate pre-embolization assessment of bleeding with CT angiography shortens the total diagnostic time, which results in prompt and more effective endovascular treatment.
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