Contrast-enhanced CT evaluation of intra-abdominal vessels in sickle cell anemia

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Abstract

Background Sickle cell anemia (SCA) is a hereditary hematological disorder that affects millions of people worldwide. Abdominal crisis can result in significant morbidity and mortality if bowel infarction is present. Vaso-occlusive crisis is the most common pathological process accounting for morbidities. However, another mechanism was recently proposed for the arteriovascular changes associated with SCA. The terminology "sickle cell vasculopathy" was raised. Unlike the intra-cranial arterial vascular abnormalities associated with SCA (frequently reported findings of Moyamoya syndrome), there is scarce radiology literature describing the intra-abdominal vascular changes during or outside a crisis in SCA. Contrast-enhanced CT is a frequently used modality to assess abdominal vasculature and end-organ damage in acute abdominal crisis. Knowledge of the morphology of intra-abdominal vessels, especially the arteries may explain the underlying mechanism of abdominal crisis and potentially alter the management. The study aims to evaluate the structure of the intra-abdominal vasculature in SCA patients using contrast-enhanced computed tomography (CT) images.

Methods This retrospective study reviewed the medical records of SCA patients who underwent contrast-enhanced CT of the abdomen and pelvis between 2003 and 2020 irrespective of abdominal crisis status. The CT studies were reviewed for the presence of vascular abnormalities, ischemic bowel changes, or other end-organ ischemic changes.

Results Out of 509 patients, only 78 met the inclusion criteria. Two patients showed diffuse small caliber of the intra-abdominal arteries in the setting of abdominal crisis with end-organ ischemia but no bowel changes. None of the CT studies showed arteriovascular manifestations like advanced intra-cranial vasculopathy in SCA (Moyamoya syndrome-like changes) and none had vasculitis-like abnormalities. None of the patients had bowel ischemia, but six patients had renal and splenic infarcts in the absence of vascular abnormalities. One patient had venous varices secondary to superior mesenteric vein thrombosis.

Conclusions The vast majority of SCA patients demonstrate no noticeable arteriovascular abnormality on a contrast-enhanced CT of the abdomen. Diffuse intra-abdominal vasospasm is present in a very small percentage of SCA abdominal crisis patients.

Keywords Sickle cell anemia, Vasculopathy, Intra-abdominal arterial branches, Vasculitis, Moyamoya, Abdominal crisis, Vascular abnormalities

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Background

Sickle cell anemia (SCA) is a hereditary hematological disorder that results in abnormal hemoglobin S (HbS) [1]. According to the world health organization, 7% of the world's population carries a hemoglobinopathy gene, and 50% of those carry a SCA gene [2]. The prevalence of SCA is highest in Africa and the Mediterranean region reaching 200 per 10,000 in the Nigerian population. [2, 3]

In Saudi Arabia, the prevalence can reach 145 per 10,000 in the eastern region. [4] SCA carries a significant burden on health care systems as well as tremendous financial and educational impact on SCA individuals. [3] For that, a Saudi national premarital screening program was implemented to screen applicants for SCA among other diseases. [5, 6]

Vaso-occlusive crisis is the most common pathological process accounting for 90% of patients' morbidities. [3] It results from the occlusion of post-capillary venules by the abnormal sickled red blood cells (RBCs) [1]. Abdominal pain is one of the significant SCA crises, reaching up to 39.1% in Nigeria [7] and 16.8% in Saudi Arabia [3]. The morbidity and mortality are high if it results in bowel ischemia [8].

Recently, a second mechanism was proposed for sickle cell crises. SCA affects the structure of the blood vessels, and the terminology "sickle cell vasculopathy" was raised [9, 10]. The intra-cranial arteries in SCA can show advanced changes as abnormal segments of vascular narrowing and wall thickening intermittently with areas of vascular dilatation and tufts of abnormal vessels without the presences of inflammatory cells, which is known as Moyamoya syndrome. [11]

Similar structural vascular changes have never been reported outside the intra-cranial vasculature in SCA. Review articles mention vasculitis-like changes in the abdomen [12, 13] based on a single case report associating SCA with systemic necrotizing vasculitis. [14] Another recent case study reported abdominal manifestations to be related to the SCA crisis, however, the final diagnosis favored medium vessel vasculitis [15].

With the current literature gap in the abdominal arterial vascular changes in SCA, more studies should investigate and report these changes to avoid misdiagnoses in SCA crises. Appropriate assessment of vascular changes, especially of the abdomen, is critical as the supply of vital organs might be affected. Computed tomography (CT) images of the abdomen and pelvis with and without contrast are the mainstay of assessing abdominal manifestations in SCA [13, 16]. This study aims to evaluate the structure of the intra-abdominal vessels, especially the arterial vasculature in SCA patients using contrast-enhanced CT images.

Methods

Study design

This study received ethical approval from the Institutional Research Ethics Committee with reference number 164-23.

This retrospective study reviewed contrast-enhanced CT findings in patients with SCA. SCA patients' files were retrieved from the medical records for the period of 1/1/2003–31/1/2020, using the international classification of diseases (ICD-10) codes D57.0 and D57.1, corresponding to sickle cell anemia with crisis and sickle cell anemia without crisis, respectively. The medical records were reviewed for the presence of a contrast-enhanced CT of the abdomen and pelvis.

Inclusion criteria included: the diagnosis of sickle cell anemia and having a contrast-enhanced abdomen and pelvis CT in the arterial or the portovenous phase whether the patient is presenting with or without an acute abdominal crisis (by the history provided with the CT scan request form).

Exclusion criteria included: the lack of contrast on a CT examination, the sub-optimal assessment of the vessels on a contrast-enhanced CT either due to sub-optimal contrast opacification of the vessels or motion, and the inability to retrieve the CT images from the picture archiving and communication systems (PACS).

Imaging acquisition: All patients underwent a CT examination of the abdomen and pelvis, including the diaphragm to the symphysis pubis, in one of the following machines: GE LightSpeed Pro 32-slice scanner, Siemens SOMATOM Sensation 16-slice scanner, Siemens SOMATOM Definition AS 128-slice scanner, Siemens SOMATOM Definition Flash 128-slice scanner, and Siemens SOMATOM Definition 64-Slice scanner. Coronal and sagittal reformatted images were reconstructed from 0.625 mm thickness axial images. Reconstructed axial images ranged between 2.5- and 5-mm slice thickness.

Iodinated Contrast material (Xenetix 350 or Omnipaque 350) was administered at a rate dependent on the cannula size, contrast phase, and patient's age, averaging 2.8–3 ml/s for a 22-gauge cannula for a portovenous study. The total contrast volume ranged between 80 and 100 ml. There was a 45–50 s delay for the arterial phase examinations and a 70–75 s delay for the portovenous phase examinations.

Data collection and imaging assessment

Patient's medical records and PACS were utilized to collect the following data: sex, weight, height, age at the CT examination, contrast phase of the CT abdomen and pelvis, and CT indication.

The CT studies were reviewed by an abdominal radiologist, with nine years of experience, for the presence or absence of vascular abnormalities, ischemic bowel changes, or other end-organ ischemic changes.

The arterial vascular changes were noted for the location and the nature of abnormality: any caliber change, vascular wall thickening, luminal narrowing, beaded appearance, luminal thrombosis, or aneurysms. Any abnormalities were specifically observed for the large

and medium size arteries. Venous abnormalities such as thrombosis and varices were recorded.

The solid organ ischemia changes were defined as wedge-shaped areas of hypoenhancment or lack of contrast enhancement.

Bowel ischemia changes were defined as segmental or diffuse bowel wall thickening, areas of lack of contrast enhancement, air in the bowel wall (pneumatosis intestinalis) or the mesenteric veins, and -in severe cases- bowel perforation [12].

Statistical analysis

Data were collected using Microsoft Excel 360 spreadsheet. The descriptive data were populated with the use of means with standard deviations and frequencies with their percentages.

Results

The medical records yielded 509 patients. The number of patients who had a CT of the abdomen and pelvis performed was 84 cases constituting 16.5%, and only 78 cases (15.3%) had a contrast-enhanced examination in the arterial or portovenous phase and hence fulfilled

the inclusion criteria. The remainder of the patients were excluded (Fig. 1).

Most of the CT studies were conducted in the portovenous phase, accounting for 83.3% ($n=65$) of the examinations. Only 10.3% ($n=8$) were performed in the arterial phase, while a small portion, 6.4% ($n=5$), underwent both an arterial and a portovenous phase.

The cases which were performed for indications other than SCA crisis were 37.2% ($n=29$), whereas 26.9% ($n=21$) of the patients had an abdominal crisis, and 35.9% ($n=28$) had unknown presentations (Table 1).

As shown in Table 2, of all the included 78 cases of SCA with a contrast-enhanced abdominopelvic CT, only two patients showed a diffuse small caliber of the intra-abdominal arteries in the setting of abdominal crisis. Those patients didn't have any other vascular morphologic changes or luminal occlusion. The two patients had sequala of end-organ damage in the form of renal parenchymal scarring and one had liver infarction as well. However, they didn't have bowel ischemia changes.

None of the 78 cases had shown abnormal vascular configurations in the form of intermittent abnormal areas of luminal narrowing with wall thickening alternating
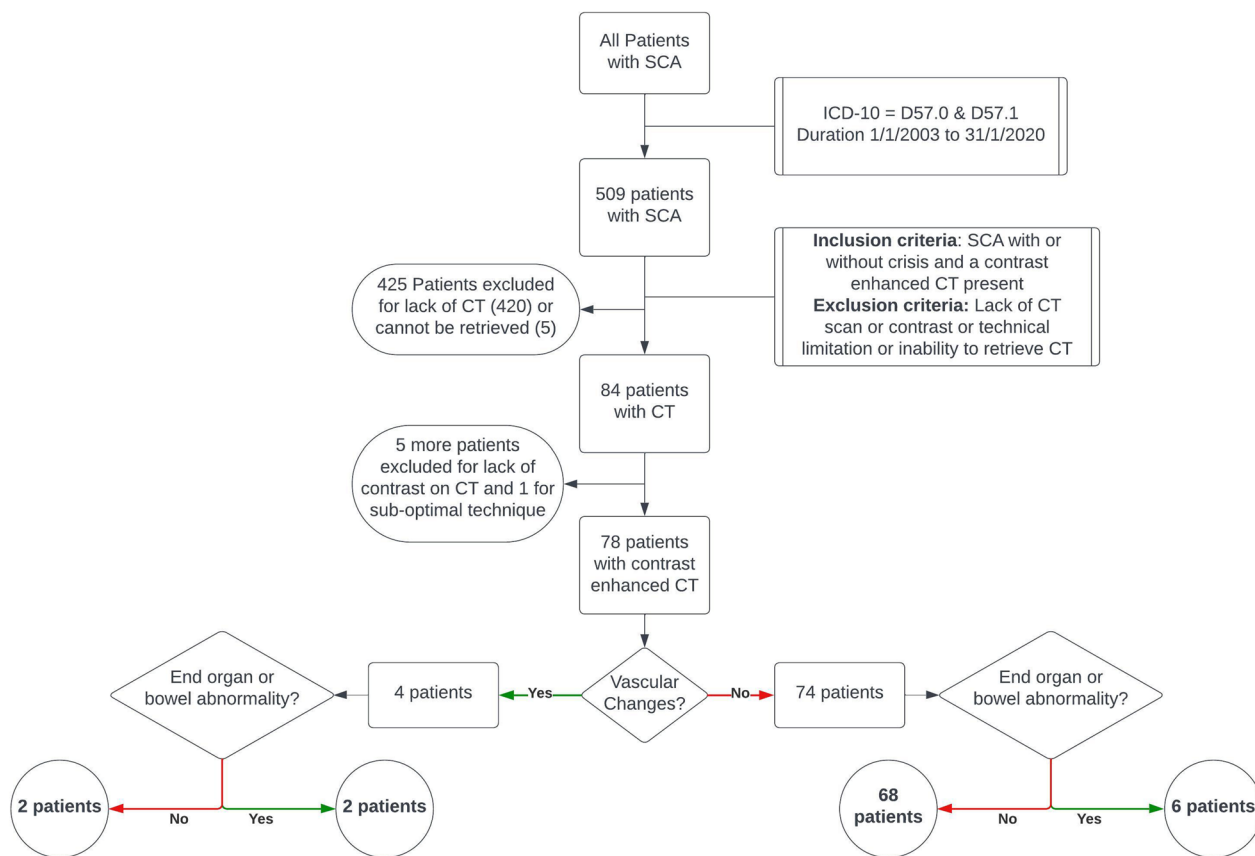
Fig. 1 Flow diagram of the study population selection process and their CT imaging findings

Table 1 SCA Patient Demographics (n = 78)

Patient's demographics	Value
Age*	25 ± 12 years
Sex**	Female: n = 43 (55.1%)
	Male: n = 35 (44.9%)
Height*	162.1 ± 15.5 cm
Weight*	50 ± 15.8 kg
CT indication	SCA Crisis n = 21 (26.9%)
	No Crisis n = 29 (37.2%)
	Unknown n = 28 (35.9%)

*The values are in mean ± Standard deviations (SD)

**The values are frequencies with % in brackets

with regions of vascular dilatation. None had abnormal tufts of vessels or aneurysms. None of the patients had bowel ischemia. Six patients had renal and splenic infarcts in the absence of vascular abnormalities (Figs. 2 and 3).

Two additional patients had incidental vascular abnormalities in the form of venous varices (Fig. 4) in the setting of superior mesenteric vein occlusion and an isolated left renal artery aneurysm.

Images A and B: coronal plane portovenous phase CT images of the abdomen and pelvis with positive oral contrast show normal caliber of distal jejunal branches of the superior mesenteric artery (SMA) (white arrows in A)

**Table 2** Vascular and end-organ changes in SCA patients in a contrast-enhanced CT examination

History of abdominal crisis	Normal CT	Vascular changes only	End-organ changes only	Vascular and end-organ changes	Bowel ischemia	Total
Positive crisis	15	1*	3	2	0	21
Negative crisis	27	1**	1	0	0	29
Unknown crisis	26	0	2	0	0	28
Total Cases	68 (87.2%)	2 (2.5%)	6 (7.7%) ***	2 (2.5%) ****	0	78 (100%)

*Intra-abdominal varices

** Left renal artery aneurysm
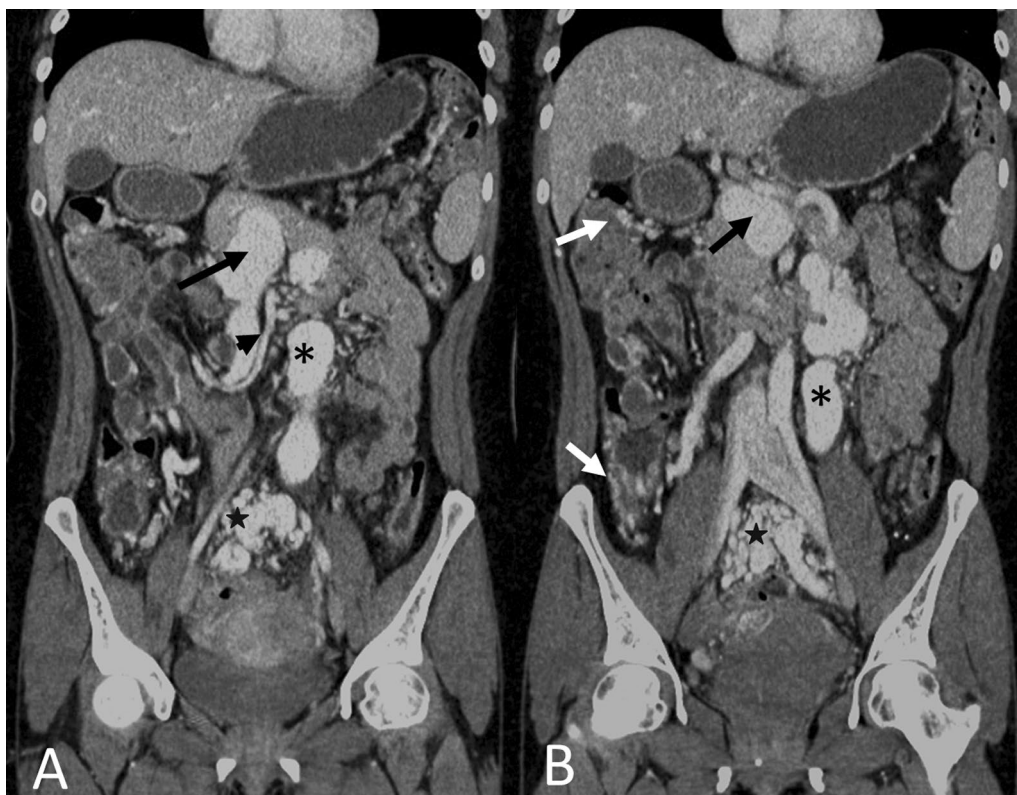
*** Two renal infarctions and 4 splenic infarctions

**** Diffuse arteriovascular luminal narrowing and vasoconstriction with 1 case of liver infarction and 2 cases of renal scarring
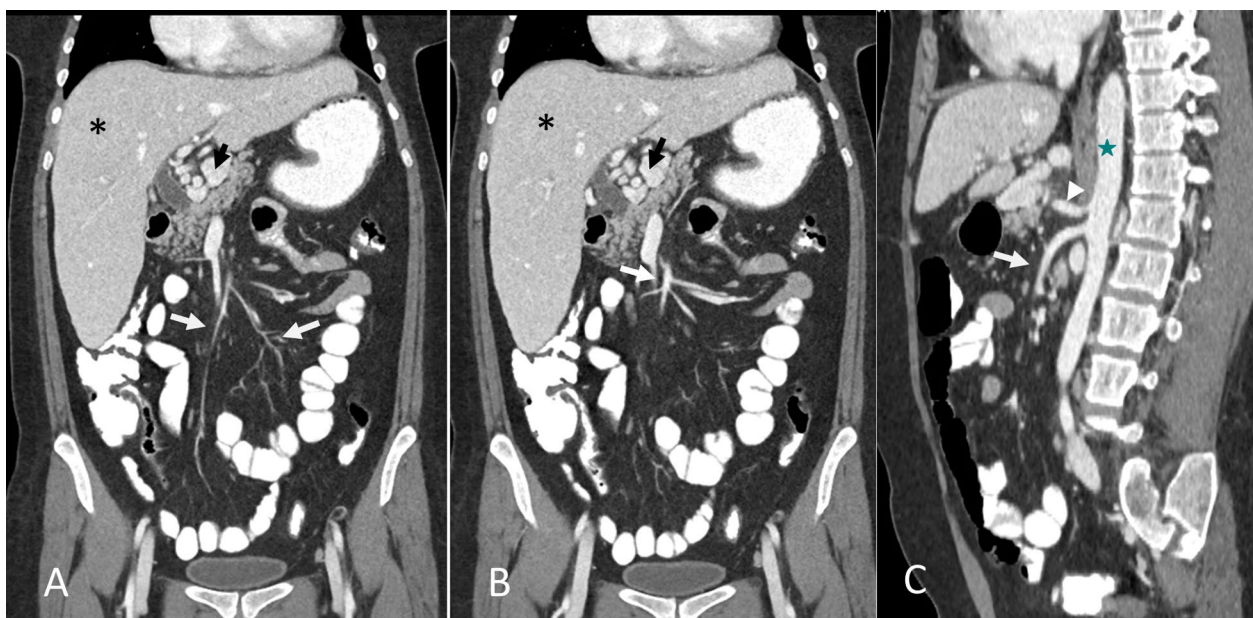
Fig. 2 A contrast-enhanced CT of a 21-year-old woman with SCA without abdominal crisis

Fig. 3 A contrast-enhanced CT of a 27-year-old woman with SCA who came with abdominal crisis

Fig. 4 A portovenous phase CT of a SCA patient with superior mesenteric vein (SMV) thrombosis

and the branching points of the SMA (white arrows in B) with no vessel wall abnormalities. The liver is enlarged with slight surface nodularity (black *) along with enlarged enhancing porta hepatis lymph nodes (black arrows).

Image C: a sagittal plane portovenous phase CT image of the abdomen and pelvis with positive oral contrast shows the normal caliber of the whole length of the abdominal aorta (*), origin of SMA (white arrow) and origin of the celiac axis (arrowhead) with no vessel wall abnormalities or occlusion.

Images A, B are coronal plane arterial phase abdominopelvic CT images while image C is a coronal plane portovenous phase CT image. The SMA jejunal branches (white arrows in A and C), left hepatic artery (black arrow in A), aortic bifurcation (white arrowhead in B) and inferior mesenteric artery (IMA) (black arrow in B) appear of normal caliber with no evident occlusions or vessel wall abnormalities. Peripheral areas of the enlarged spleen lack contrast enhancement (black asterisk '*'), suggesting splenic infarctions. Normal enhancement is observable in the small bowel loops, although mild submucosal edema is present (white symbol '*'). Additionally, moderate ascites is evidenced (white asterisk '*').

Images A and B are coronal plane portovenous phase abdominopelvic CT images of the abdomen and pelvis. Extensive pelvic venous collaterals are present (black symbol '*'), along with varices lining the walls of the bowel loops (white arrows). These observations are due to chronic SMV thrombosis, with collateral drainage occurring through the dilated inferior mesenteric vein (IMV) (black asterisk '*'). The IMV subsequently drains into the portal venous confluence (black arrow). The partially visible SMA (black arrowhead) appears of normal caliber, with no discernible occlusions or vessel wall abnormalities.

Discussion
This study focused on assessing the gross morphology of the intra-abdominal vessels especially the arterial branches by contrast-enhanced CT in SCA patients in the presence and absence of abdominal crisis. The study demonstrated that 97.5% of SCA patients, irrespective of crisis state, sex or age had normal intra-abdominal arterial morphology, including caliber, wall thickness, and luminal patency. Only 2.5% of the patients had diffuse vascular luminal narrowing during their abdominal crisis.

In the context of SCA, a notable discrepancy arises between the conspicuous absence of significant intra-abdominal arteriovascular alterations and the abundant intra-cranial vascular changes reported in Radiology literature. Despite the absence of overt clinical manifestations, intra-cranial vasculopathy is frequently observed by magnetic resonance imaging (MRI) in a substantial proportion of SCA patients, reaching rates of 30–40% which is known as Moyamoya syndrome in the advanced forms. [17]

The recent expansion in the pathophysiology of SCA might explain the discrepancy in the vascular morphology between the abdomen and the brain. The mechanism of disease now includes two phenotypes with different subsets of complications.

The first phenotype, "Viscosity-vaso-occlusion," is the result of the known sickling of the RBCs occluding the microvasculature including arterioles or post-capillary venules. Hence acute vaso-occlusive painful crisis occurs. Acute chest syndrome and osteonecrosis are the results of this phenotype, characterized by ischemia–reperfusion injury, inflammation, and infarction. [9, 10] Abdominal crises are probably accounted for in this pathophysiologic process, as evident in our study results and Gardner's previous work. [8, 18]

The second phenotype, "Hemolysis-endothelial dysfunction," is due to the ongoing hemolysis of the RBCs that depletes the nitric oxide (NO) stores and prevents further NO synthesis. NO is essential for vasodilation, and without NO, there is vasoconstriction, endothelial injury, and vascular remodeling. This results in ischemia of the end-organs. When this process predominates, it results in strokes, pulmonary hypertension, priapism, and leg ulcers. [9, 10] The resultant "Pan-vasculopathy" from the ongoing hemolysis in this phenotype may account for the extensive large-vessel vascular manifestations in the brain. However, the mechanisms discussed behind the SCA presentations are complex and often interrelated, making it challenging to justify a single event happening in isolation. [10]

Unlike the abundant literature on the intra-cranial vasculature of SCA, a single study assessed the vessels in abdominal crises and the presence of bowel ischemia in SCA patients by CT by Gardner and Jaffe. They looked at the various manifestations and components of SCA vaso-occlusive crisis. They evaluated whether the vessels were patent or not. They concluded that there were no macroscopic arterial or venous occlusions depicted by CT, but rather, the abdominal crisis is likely the result of microvascular occlusions. [8, 18] Our study is the second study that looks at various intra-abdominal vascular details of the medium and large arteries. We arrived to the same conclusion that the majority of the patients have normal intra-abdominal vessels on a contrast-enhanced CT.

Over the past decade, the working hypothesis was that the intra-abdominal vascular changes in SCA resemble those of vasculitis. To date, radiology review articles [12, 13] base this conclusion on a single case report from

1987. The case report mentioned an association between systemic necrotizing vasculitis and SCA. Their hypothesis was that SCA patients might be prone to immune complex disorders, including vasculitis [14].

None of our SCA patients demonstrated vascular changes that simulate vasculitis in form of beaded appearance with intermittent areas of luminal narrowing, wall thickening, or wall enhancement, nor did they have microvascular aneurysms. The lack of vasculitis-like changes in SCA in our study does not disprove the rare association between SCA and vasculitis [14], but it shows that when medium or large vasculitis changes are present in SCA, other differentials and management options should be considered [15].

The observed diffuse vascular luminal narrowing in 2.5% of our patients during their abdominal crisis could be due to diffuse vasoconstriction in a hypovolemic shock state or due to the administration of supportive therapy. An alternate hypothesis is explained by the vasospasm or failure of compensatory vasodilator mechanisms which is a known part of the proposed mechanism of SCA crisis [19].

Proper vascular imaging can be achieved by vascular angiography which is superior to CT in evaluating arteries. However, CT angiography (CTA) nowadays is a more favored noninvasive technique. CTA studies provide an excellent assessment of vessel caliber and luminal changes [20]. Our study observed a lower percentage of CTAs. This observation could be attributed to imaging guidelines that recommend the use of CT scans, both with and without contrast, for evaluating abdominal pain in SCA patients, without explicitly stating the necessity for CTAs [13]. Nonetheless, portovenous phase CTs remain "usually appropriate" in large-vessel vasculitis or "may be appropriate" in medium vessel vasculitis assessment per the American College of Radiology (ACR) appropriateness criteria, though inferior to CTAs. Portovenous studies add information regarding vessel wall thickness, enhancement, and end-organ damage [20]. Figures 2, 3, and 4 are examples from our studied cases that illustrate that the medium and large abdominal arteries can be assessed by portovenous scans.

Advancements in imaging acquisitions, such as dual-energy CT and spectral CT, offer potential solutions to the less-than-optimal assessment of arteries in conventional portovenous contrast-enhanced CTs. For example, dual-energy CT can provide higher vascular contrast enhancement with lower iodinated contrast dosages (iodine boost) in a low-keV setting, regardless of the contrast phase [21]. Photon counting is a new emerging technology that promises higher-resolution low signal-to-noise imaging with the use of lower radiation dosages [22]. However, to date, unlike the cardiovascular anatomy, abdominal vascular anatomy has not been studied extensively with this technology. Additionally, photon counting is not widely available for commercial use yet. Further research could help determine if these new techniques can assess even the smallest vessels for changes. [21] Such techniques, if applied to SCA patients, could help reveal other abnormalities that are not usually detected by conventional CTs.

This study's retrospective design introduces several inherent limitations. Among these is the absence of comprehensive information regarding the crisis status, for 35.9% of the patients. Furthermore, despite our institution's tertiary care setting, we encountered a limited number of SCA-related CT cases for analysis. Additionally, only a small subset of patients (16.7%) underwent an arterial phase during their abdominal CT examination. A multicenter study with a larger sample size, including patients in different crisis states, and the incorporation of advanced imaging modalities, could provide valuable insights into this area. Furthermore, exploring the correlation between intra-abdominal vasculature, other body part arteries, as well as intra-cranial vessels, may help elucidate the underlying mechanisms of "SCA vasculopathy." Ultimately, this research could lead to improved diagnostic and therapeutic strategies for patients suffering from SCA and its complications.

Conclusions

The vast majority of SCA patients have no obvious CT changes in intra-abdominal arterial morphology, regardless of the abdominal crisis status. Contrast-enhanced CT shows no macrovascular occlusions or vasculitis-like changes. This may support the hypothesis of microvascular occlusion in abdominal crisis. Furthermore, advanced imaging modalities should be considered to elucidate the pathogenesis of the abdominal crisis in SCA patients.

Abbreviations

ACR	American college of radiology
CT	Computed tomography
CTA	CT angiography
HbS	Hemoglobin S
ICD-10	International classification of diseases codes
IMA	Inferior mesenteric artery
IMV	Inferior mesenteric vein
MRI	Magnetic resonance imaging
NO	Nitric oxide
PACS	Picture archiving and communication systems
RBCs	Red blood cells
SCA	Sickle cell anemia
SMA	Superior mesenteric artery
SMV	Superior mesenteric vein

Author contributions

AB contributed to the data collection, analysis and the writing of this manuscript.

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Availability of data and materials

The data that support the findings of this study are not publicly available due to privacy and ethical restrictions. The study involves human participants and sharing the dataset publicly would compromise participant privacy rights and would not comply with the Health Insurance Portability and Accountability Act (HIPAA). However, further information about the data and conditions for access are available from the corresponding author upon reasonable request, ensuring compliance with privacy regulations.

Declarations

Ethics approval and consent to participate

This study has been approved by the research ethics committee with NCBE registration number: HA-02-J-008. The reference number for this ethical approval is 164-23.

Consent for publication

There are no individual person's data. Consent from individuals is not applicable.

Competing interests

There are no financial or nonfinancial competing interests.

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