Original Article

Appendectomy During Pregnancy in Sickle Cell Disease Patients

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ABSTRACT

Background/Aim: Acute appendicitis during pregnancy is the most common problem encountered in general surgery. However, limited data are available regarding acute appendicitis in pregnant sickle cell disease (SCD) patients. We aimed to study was the clinical presentation, management, and outcome in SCD patients who underwent appendectomy for suspected acute appendicitis during pregnancy. Materials and Methods: Between January 1998 and December 2006, 65 pregnant patients with clinically suspected appendicitis underwent surgery at our hospital of whom 11 were SCD patients. Data collected retrospectively included age, gestational stage, clinical signs and symptoms, operative findings, and complications. Results: Eleven out of 65 (16.9%) were SCD patients. The mean age in the SCD group was 22.5 years while the mean gravida and parity values were 2.0 and 1.1, respectively. The mean HbS, HbF, and HbA2 values were 71.1, 26.2, and 2.6%, respectively. Abdominal pain and vomiting were the most common presenting symptoms regardless of the gestational age. Abdominal and rebound tenderness were the most common physical signs. The mean operative time was 49 min (range: 35-125 min) and the average length of postoperative hospital stay was 4.7 days (2-8 days). There was no maternal death but one patient had spontaneous abortion during the first trimester, two days after the operation. Two patients had premature, postoperative delivery at 30 and 31 weeks of gestation. Conclusion: The clinical manifestations and diagnosis of appendicitis in sickler pregnant patients are similar to those in nonsickler pregnant patients.

Key Words: Acute appendicitis, pregnancy, sickle cell disease

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Sickle cell disease (SCD) is a genetically transmitted disorder of red blood cells. It is present throughout Saudi Arabia and is common in the Al Hassa area, with an incidence of 1.08%.^[1] Pregnancies in SCD patients present a clinical challenge as maternal mortality is 1-2% and perinatal mortality is 5-6%.^[2,3] Acute appendicitis is the most common extrauterine surgical emergency seen in pregnancy, and appendectomy is the most common nonobstetric procedure being performed on pregnant patients.^[4-7]

There are no data for acute appendicitis in pregnant SCD patients. We report our institutional experience with appendectomy for pregnant patients with SCD. Our aim was to explore the clinical presentation, management, and outcome in SCD patients who underwent appendectomy for suspected acute appendicitis during pregnancy.

MATERIALS AND METHODS

Between January 1998 and December 2006, 65 pregnant patients underwent appendectomy for clinically suspected appendicitis in the general surgery department of King Fahad Hospital-Hofuf (67,990 deliveries in the study period). Eleven (16.9%) of the 65 pregnant patients were SCD patients, who formed the study group. Clinical data collected retrospectively

included patients' age, gravidity, parity, gestational age, the presenting symptoms, percentage of sickle cell hemoglobin, extent of sickle cell crisis, blood transfusion, the time between the onset of symptoms and surgery, the severity of the inflammatory changes in the appendices, operative time, length of postoperative hospital stay, fetus birth weight, mode of delivery, and postoperative complications.

The criteria for the diagnosis of SCD were the absence of HbA₁, HbS >40%, and HbA₂ <3.5%. The diagnosis of acute appendicitis was based on clinical examination, and pregnancy was confirmed by ultrasound. The general surgeon performed the operation in the presence of an obstetrician who assessed all patients before and after the appendectomy. Neither urinary catheter insertion nor deep vein thrombosis prophylaxis was routinely performed. Antibiotics were administered postoperatively in complicated cases. All patients were operated in decubitus position and the appendix was accessed without difficulty through a McBurney's incision in all patients.

RESULTS

Details of the clinical cases are shown in Table 1. The mean age in the SCD group was 22.5 years; mean gravida and

Table 1: Details of the 11 clinical cases											
Case	Age	Gravida	Parity	Trimester	HbS	HbF	HbA2	Blood transfusion	Birth wt. (g)	Mode of delivery	Complication
1	20	2	1	1st	75.3	21.2	3.5	+	1255	-	Abortion
2	18	1	0	3rd	68.7	27.4	3.9	+	2890	LSCS	Wound infection
3	23	3	2	3rd	74.3	23.6	2.1	_	2920	AD	Premature delivery
4	24	4	2	2nd	71.9	25.4	2.7	_	2850	AD	Pyle nephritis
5	20	2	1	2nd	71.3	26.8	1.9	_	2895	ND	-
6	21	1	0	2nd	67.7	29.9	2.4	+	3150	AD	Crisis
7	26	2	1	1st	68.5	28.2	3.3	_	2880	AD	-
8	27	3	2	2nd	69.7	27.4	2.9	_	2990	ND	-
9	24	1	0	3rd	69.8	27.4	2.8	+	2850	ND	Premature delivery
10	24	3	2	2nd	73.9	24.2	1.4	+	3765	LSCS	Crisis
11	20	2	1	2nd	71.1	26.8	2.1	+	3150	LSCS	Wound infection

LSCS = lower segment cesarean section, AD = assisted delivery, ND = normal delivery

Symptoms and signs	Sickler p	patients	Nonsickler patients		
	Inflamed appendix n = 8 (%)	Normal appendix n = 3 (%)	Inflamed appendix n = 30 (%)	Normal appendix n = 24 (%)	
Abdominal pain in right lower quadrant	6 (75)	2 (67)	28 (93)	21 (88)	
Abdominal pain in right upper quadrant	2 (25)	1 (33)	2 (7)	3 (22)	
Nausea	3 (38)	1 (33)	12 (40)	10 (42)	
Vomiting	5 (62)	2 (67)	18 (60)	14 (58)	
Abdominal tenderness in right lower quadrant	7 (88)	2 (67)	25 (83)	19 (79)	
Abdominal tenderness in right upper quadrant	1 (22)	1 (33)	5 (17)	5 (21)	
Rebound tenderness	6 (75)	3 (100)	26 (87)	17 (71)	
Rovsing's sign	5 (62)	-	21 (70)	15 (63)	
Temperature (>37.5°C)	4 (50)	-	16 (53)	6 (25)	
Leukocytosis >16	6 (75)	1 (33)	25 (83)	15 (63)	

parity were 2.0 and 1.1, respectively. The mean HbS, HbF, and HbA2 values were 71.1, 26.2, and 2.6%, respectively. Two (18%), six (55%), and three (27%) patients were in their first, second, and third trimesters, respectively. Vomiting and abdominal pain in the right lower abdomen (distinct from sickle cell crisis pain) were the most common presenting symptoms regardless of the gestational age [Table 2].

Abdominal and rebound tenderness of the same site were the most common physical signs. Four (36%) patients developed pyrexia and seven (64%) patients had leukocytosis. The mean maximal temperature was 37.4° C and the mean leukocyte count was $15.9 \times 10^{\circ}$ /l. Appendectomy was performed within 24 h of the onset of symptoms in all patients in the first trimester. Four out of six patients were operated within 24 h of the onset of symptoms in the second trimester while all patients in their third trimester had symptoms for >48 h. The mean operative time was 49 min (range: 35-125 min) and the length of postoperative hospital stay was 4.7 days (range: 2-8 days).

Acute appendicitis was confirmed histologically in six patients, appendicle perforation in two cases, and three patients had normal histological findings. Blood transfusion was required in six (55%) patients to correct anemia and also to prepare for surgery. Wound infection occurred in two (18%)

cases, two patients developed crisis, and one (9%) patient developed pyelonephritis postoperatively. One patient in the first trimester had spontaneous abortion two days after the operation. Two patients in their third trimester had premature delivery in the immediate postoperative period. Three patients (27%) needed lower segment cesarean section (LSCS) and four patients (36%) needed assisted delivery. The mean birth weight of the babies was 2808.6 g; there was no maternal mortality.

DISCUSSION

Homozygous SCD is now known to be widespread and has broad clinical variability. A relatively mild manifestation of SCD (compared to the forms observed in other countries) has been noted in the Eastern Province of Saudi Arabia. [8-10] High maternal and perinatal mortality rates clearly indicate the high level of risk associated with pregnancy in SCD patients mainly in the African and Afro-American population. [11,12] Although recent data now suggests that appropriate medical care can help bring pregnancy to term safely in SCD patients. [12]

The reported incidence of appendicitis during pregnancy varies in the literature - from one in 705 to one in 2700 deliveries in the normal population. [13,14] The incidence in our series was one in 1046 deliveries (out of a total of 67,990

Table 3: Relation between time from onset of symptoms to operation and pathology of the appendix **Trimesters** Symptoms in sickler patients Symptoms in nonsickler patients <24 h (41 cases) <24 h (6 cases) >24 h (5 cases) >24 h (13 cases) First trimester 1 (i), 1 (nl) 3 (nl), 6 (i), 1 (p) 1 (nl) Second trimester 4 (i) 2 (nl) 9 (nl), 9 (i), 3 (g) 1 (nl), 2 (i), 2 (p) Third trimester 1 (g), 1 (p), 1 (i) 6 (nl), 2 (i), 1 (p), 1 (g) 4 (nl), 1 (i), 1 (p), 1 (g) nl = normal, i = inflamed, g = gangrenous, p = perforated

deliveries in the study period), which is consistent with previous reports. To the best of our knowledge, there is no data available regarding the incidence of acute appendicitis and appendectomy in SCD patients during pregnancy. The clinical diagnosis of acute appendicitis in pregnancy is difficult. [15-17] In our study, 75% of the SCD patients reported pain in their right lower quadrant, the same as reported in other studies for pregnant nonsickler patients. [15]

Vomiting was a common complaint (67%) and it was comparable to other reports for pregnant nonsickler patients with acute appendicitis. [18] Fever was not very helpful in diagnosing acute appendicitis in pregnancy. [13] In our study, only 50% of the inflamed appendix patients had fever, while none of the sickler patients had pyrexia in the normal appendix group. Pregnancy is associated with physiological leukocytosis, which may be as high as $16,000/\mu$ I. [18] Therefore, in the appropriate setting, the presence of leukocytosis > $16,000/\mu$ I in pregnant women may be an indicator of acute appendicitis. [19]

In our series, 75% of the acute appendicitis patients with SCD had leukocytosis >16,000/µl. In agreement with the findings of other studies, we found a significant difference in the leukocyte counts in patients having acute appendicitis compared to those with noninflamed appendices. [20] As with nonsickler patients, delaying the appendectomy in SCD patients with appendicitis beyond 24 h in their third trimester was associated with gangrene and perforation of the appendix [Table 3]. [21,22] Maternal mortality is rarely encountered in cases of acute appendicitis in pregnant nonsickler patients and there was none in our series as well. [15] The fetal loss and the rate of premature delivery were 9 and 18%, respectively, which is consistent with other reports. [13,19]

The incidence of complication in SCD patients in the present study is lower than in other studies. [23] This may be because of the milder form of SCD in this area (Al-Hassa) due to the presence of high levels of HbF in the affected population. The high HbF levels protect against several clinical features associated with SCD, but the association between HbF levels and the severity of the disease process is complex. [2,24] As in other studies, removal of the appendix from all patients was made easily through an incision over McBurney's Point. [25] Laparoscopic appendectomy has gradually gained universal agreement for the treatment of acute appendicitis; however, the controversy still exists over the safety and feasibility of a

laparoscopic appendectomy during pregnancy. [26,27]

CONCLUSION

In this relatively small study, no single symptom, sign, or laboratory finding is diagnostic for acute appendicitis during pregnancy in SCD patients. Hence, acute appendicitis must be considered in the differential diagnosis when pregnant SCD women complain of new-onset abdominal pain.

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