Ultrasonographic and computed tomography manifestations of intussusception secondary to primary non-Hodgkin's lymphoma diagnosed in puerperium: report of a case and review of the literature

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Summary

Non-Hodgkin's lymphoma is infrequently diagnosed during pregnancy and puerperium. A 21-year-old woman, para II, gravida II, on the seventh postpartum day arrived at the Emergency Gynaecologic Clinic complaining of colicky abdominal pain associated with bilious vomiting. A large tender mass of firm consistency was palpated in the midline towards the left hypochondrium. A CT scan of the upper abdomen showed thickened small bowel with halo, specifically at the junction of the jejunum with the ileum, resembling imaging of intussusception. Also, blocks of mesenteric and paraortic lymph nodes and multiple lymph nodes inducing enlargement of the anterior mediastinum were found. These findings rendered a picture of lymphoma in Stage at least III. Subsequent exploratory laparotomy confirmed our preoperative diagnosis. Histopathological examination of the lymph node biopsies established the diagnosis of a primary non-Hodgkin's lymphoma. Judging from the extent of the non-Hodgkin's lymphoma in our patient seven days after delivery the disease probably already existed during the last trimester of pregnancy. This study demonstrates a remarkable delay in diagnosis of non-Hodgkin's lymphoma in pregnancy. The diagnostic dilemmas of non-Hodgkin's lymphoma during pregnancy are emphasized and the literature is reviewed.

Key words: Non-Hodgkin's lymphoma; Pregnancy; Puerperium; Intussusception; Ultrasonography; Comptuted tomography; Diagnosis.

Introduction

The diagnosis of any malignancy during pregnancy is uncommon [1] and its diagnosis during puerperium, particularly in an advanced stage, demonstrates the difficulties in detecting the neoplasia during pregnancy.

Hodgkin's disease is the fourth most frequently diagnosed cancer in pregnancy (1 in 1,000 to 1 in 6,000 pregnancies). In contrast, the incidence of non-Hodgkin's lymphoma during pregnancy is extremely rare, with fewer than 100 cases reported in the English literature [2, 3]. The scarcity of reports of non-Hodgkin's lymphoma associated with pregnancy in comparison with Hodgkin's disease could be explained by the differences in age distribution of these diseases [4]. Non-Hodgkin's lymphoma has an age-dependent incidence pattern with a sharp increase in frequency beyond the reproductive age [4, 5]. In contrast, Hodgkin's disease exhibits two peaks in incidence, the first in early adulthood and the second after the age of 55 [4].

Based on a few case reports, it seems that most non-Hodgkin's lymphomas complicating pregnancy are of an aggressive type and disseminated in nature [4]. A large variety of complaints that are similar to those commonly found in pregnancy and other diseases make prompt diagnosis of non-Hodgkin's lymphomas in pregnancy a difficult task [6].

In this report we document a case of a puerpera with primary non-Hodgkin's lymphoma, in advanced Stage, presented initially to the gynaecologists with the clinical picture of intussusception. The ultrasonographic and computed tomography manifestations of the disease are presented. Judging from the extent of the non-Hodgkin's lymphoma in our patient seven days after delivery, this study demonstrates a remarkable delay in diagnosis of non-Hodgkin's lymphoma during pregnancy, which was potentially harmful to the patient. The diagnostic problems of non-Hodgkin's lymphomas during pregnancy are emphasized and the literature is reviewed.

Case Report

A 21-year-old woman, para II, gravida II, presented to the Gynaecologic Emergnecy Clinic of "George Gennimatas" General State Hospital of Athens in February 2001, complaining of colicky abdominal pain associated with bilious vomiting. The patient was at the seventh postpartum day and had suffered from abdominal pain with occasional episodes of vomiting for one month. In addition, she mentioned that 20 days prior to her labor she had visited the State Maternity Hospital of Athens on a daily basis because of abdominal pain; however she was not hospitalized because she was not in labor

Revised manuscript accepted for publication May 23, 2002

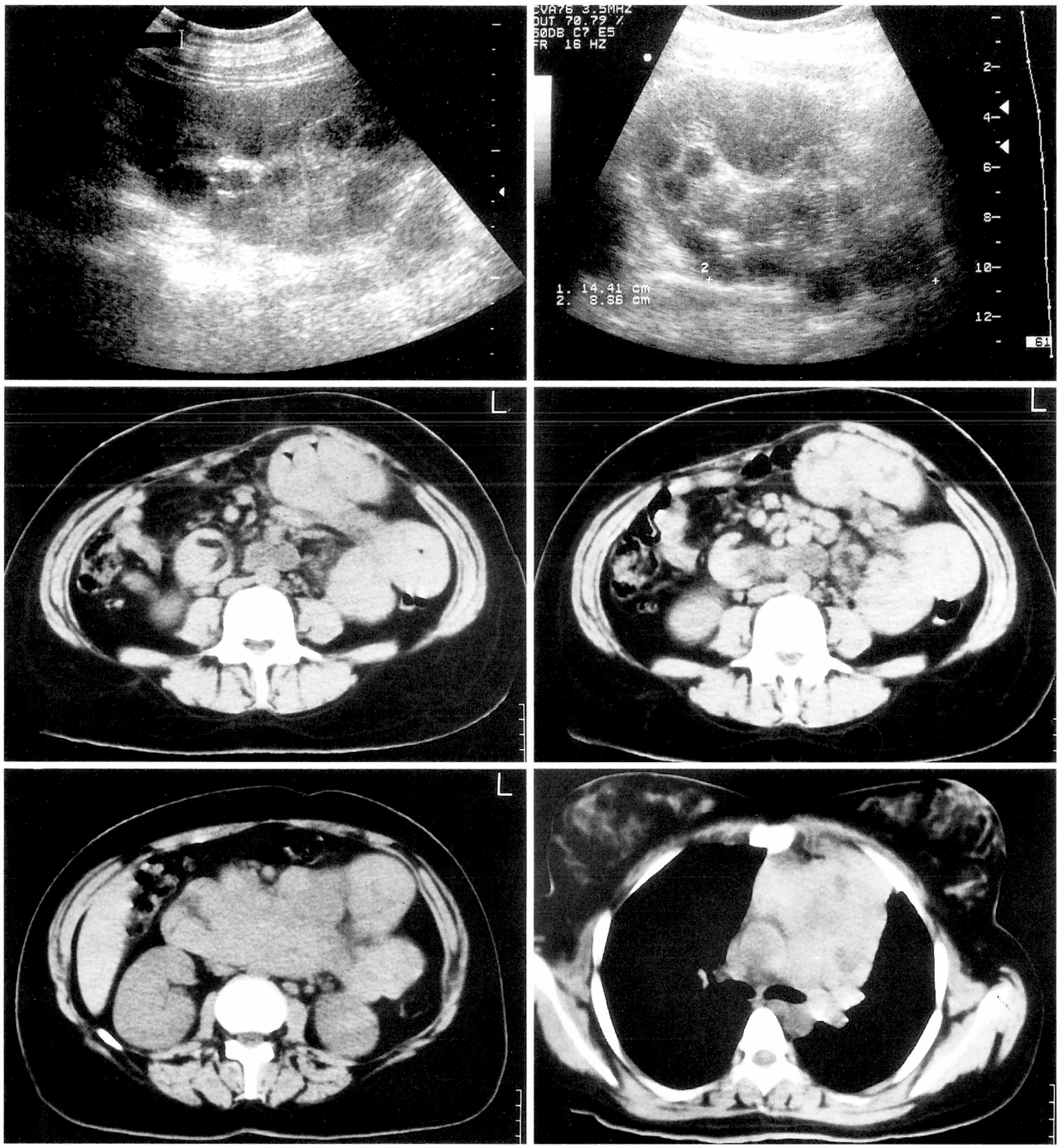

Figure 1. — Longitudinal scan at midline showing multiple paraortic nodes.
Figure 2. — Longitudinal scan showing a large mass (14.4 x 8.9 cm) with pseudo-kidney pattern, which indicates bowel pathology. Note the small hypoechoic areas representing small nodes.
Figure 3. — Computed tomography of the abdomen showing oedematous appearance of the small bowel at the junction of the jejunum with the ileum with halo; this picture resembles intussusception.
Figures 4. and 5. — Computed tomography of the abdomen showing multiple paraortic-mesenteric lymph nodes with a diameter larger than 1 cm and giving the appearance of a soft tissue mass (block of lymph nodes).
Figure 6. — Computed tomography scan of the chest showing a block of lymph nodes, which occupied the anterior mediastinum and encapsulated the ascending aorta and the pulmonary cone. Note the presence of central necrotic areas in the lymph nodes.

until the day of labor onset. During that period fetal well-being assessment with cardiotocography, ultrasound and umbilical vessel Doppler fluxometry was normal. She had a normal spontaneous delivery at the 40th week of gestation of a healthy boy weighing 3500 g and she was discharged on the fourth postpartum day.

On the seventh day after delivery she was admitted to our Emergency Gynaecologic Clinic. The patient's pulse rate was 95 beats per minute, blood pressure 120/75 mmHg and her temperature 37.2°C. The initial haematocrit was 32%, the white blood count 11,000 cells/μl with 80% polymorphonuclears and the platelet count was 350,000/μl. The urine pregnancy test was positive. On abdominal palpation a large tender mass of firm consistency was found in the midline and the left hypochondrium of the abdomen. Bimanual examination showed an enlarged uterus and the patient complained of mild tenderness during cervical movement. In order to determine the nature of the palpable abdominal mass ultrasonographic examination of the upper and lower abdomen was performed.

Abdominal ultrasound scan demonstrated a slightly enlarged uterus measuring 9 x 6.5 x 5 cm with an endometrial thickness of 16 mm. Both adnexa were regular. At midline a block of paraortic lymph nodes was found (Figure 1), while at the left hypochondrium a large mass (14 x 9 cm) with the pattern of pseudokidney in longitudinal scan and multiple small hypoechoic areas in the periphery strongly suggested bowel pathology (Figure 2). The left kidney and spleen were normal.

A furher CT scan of the chest, upper and lower abdomen was obtained. Oral or intravenous contrast medium was not administrated since the patient did not cooperate. Examination of the lesser-pelvis revealed remarkable enlargement of the uterus with inhomogeneous appearance because of the presence of haemorrhagic elements as a result of recent labour. Also, a thickened small bowel was found, specifically at the junction of the jejunum with the ileum, resembling imaging of intussusception (Figure 3). In addition, blocks of mesenteric and paraortic lymph nodes were observed (Figures 4 and 5). The diameter of most lymph nodes ranged from 1 to 3 cm, although we could not exclude the presence of larger lymph nodes since no contrast medium was administered. The abdominal blocks of lymph nodes led to pressure being applied to the intestine, to the inferior vena cava and aorta (by the paraortic block), to the portal vein (by the paraortic block) and to the pancreas (by the retrosternal block). The mediastinum block of lymph nodes led to pressure being applied to the cone of the pulmonary artery, to the pulmonary veins, to the bronchial trunk and to the pulmonary parenchyma (Figure 6). Pleural effusion or pneumothorax was absent. The above findings rendered a picture of lymphoma in Stage at least III.

Subsequent exploratory laparotomy confirmed our preoperative diagnosis of intussusception at the junction of the jejunum with ileum. Histopathological examination of the lymph node biopsies established the diagnosis of a primary non-Hodgkin's lymphoma. The patient was referred to the Haematology Department of our Hospital for combined chemotherapy treatment.

Discussion

Lymphomas are common cancers in humans. Except for Hodgkin's disease, the non-Hodgkin's lymphoma is the most common type. It generally occurs in an older age group (average age 42 years) than Hodgkin's disease [1], and for this reason the occurrence of non-Hodgkin's lymphoma during pregnancy is rarely observed. Subsequently, patients suffering from non-Hodgkin's lymphoma tend to seek treatment at an advanced stage, and therefore have to deal with a more aggressive disease [1].

Whether pregnancy affects the course of non-Hodgkin's lymphoma has yet to be determined [2]. Banks (1985) suggests that relative maternal immunosuppression to circumvent "rejection" of the fetus may allow an opportunity for neoplasia to progress, which may be only partly offset by any "lympholytic" properties of elevated serum glycocorticoids during pregnancy [7]. Based on a few case reports, it seems that most lymphomas that complicate pregnancy are not only of the aggressive type, but also of disseminated nature [4, 8]. Silva et al wrote that from 57 reported cases with non-Hodgkin's lymphoma during pregnancy only 20 women (32%) survived without evidence of the disease at the time of the study and 37 died (60%) [8]. Considering the diagnosis of the disease at the second trimester the prognosis was much poorer. Ward and Weiss reviewed 42 cases, in which 16 were diagnosed during the second trimester. Only two patients (13%) had no evidence of the disease, while all the others died (87%) [5]. In a review by Moore and Taslimi the rarity of non-Hodgkin's lymphoma during pregnancy and the wide variation of symptomatic complaints, many of which are common to pregnancy, delayed diagnosis of non-Hodgkin's lymphoma during pregnancy by more than three weeks in 40% of cases and three months in 20% [6]. Judging from the extent of the non-Hodgkin's lymphoma in our patient seven days after delivery the disease probably already existed during the last trimester of pregnancy. The tendency to make a late diagnosis of non-Hodgkin's lymphoma during pregnancy may be because attention is focused on the pregnancy itself, or because the symptoms of the disease may be masked or discounted by the pregnancy. Failure to diagnose the disease early can jeopardize the patient's health and life [9].

Intussusception in adults is rare compared to children. In 75 to 85% of the cases, there is an underlying identifiable bowel lesion as a leading point [10]. The sonographic pictures of intussusception are not specific and vary with the length and segment of bowel involved, and direction of the scanning plane [10, 11]. The most frequent sonographic feature in intussusception is a "target-like" lesion in the transverse scan and a pseudo-kidney pattern in the longitudinal section [10]. The thickened hypoechoic rim represents an oedematous change of the wall of the intussuceptum while the hyperechoid centre is due to multiple interfaces of compressed mucosal and serosal surfaces of the inner loop [10]. In our case, ultrasonography demonstrated a large mass (14 x 9 cm) with a hypoechoic peripheral rim and a hyperechoic center with multiple hypoechoic areas resembling lymph nodes. A subsequent CT scan of the upper-lower abdomen and thorax revealed the presence of blocks of paraortic, mesenteric, and mediastinal lymph nodes. Most of the lymph nodes had a central necrotic area. Moreover, at the junction of the jejunum with the ileum

intussusception was depicted. These findings were representative of lymphoma causing intussusception. In our patient the diagnosis of non-Hodgkin's disease was made in puerperium and therefore no concern existed about the potential harm of a tomographic scan on the fetus. It has been suggested that staging of non-Hodgkin's during pregnancy include personal medical history, physical examination, routine blood tests, bone marrow biopsies, chest X-ray with abdominal shielding and abdominal ultrasound [4, 5].

In conclusion, we have reported a rare case of advanced Stage non-Hodgkin's disease in a young puerpera complaining of colicky abdominal pain due to intussusception, who initially visited gynaecologists. Judging from the extent of the non-Hodgkin's lymphoma in our patient seven days after delivery, the disease probably existed during the last trimester of pregnancy. Therefore, our study demonstrated a remarkable delay in diagnosis of non-Hodgkin's lymphoma in pregnancy. It seems that attention was focused on the pregnancy and the symptoms of the non-Hodgkin's disease were underestimated.

Acknowledgements

The authors would like to thank colleagues in the Departments of Internal Medicine, General Surgery and Haematology for the care and further management of the patient. Also, the authors would like to thank our colleagues in the Department of Pathology for the microscopic examination of the surgical specimens.

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