

## CASE REPORT

# Deciduosus of the Appendix Manifesting as Acute Abdomen in Pregnancy

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## SUMMARY

Acute appendicitis is one of the most common surgical diseases during pregnancy. In rare cases, ectopic decidual tissues (deciduosus) can develop in the appendix and cause inflammation. Here we report a well-documented case of appendicular deciduosus presenting with acute abdominal pain and resulting in gangrenous appendicitis.

**Key words:** appendicitis, pregnancy, deciduosus

## AIM OF THE DEMONSTRATION

In order to increase the awareness of rare appendicular diseases and the peculiar differential diagnosis of appendicitis in pregnancy, here we present a rare case of appendicular deciduosus causing acute appendicitis in a pregnant lady.

## CASE REPORT

A 33-year-old primigravida woman entered the hospital in 28/29 weeks of gestation. The patient reported right lower abdominal pain lasting for 11 hours. Initially, preterm delivery was suspected. The body temperature was 37.2 C°. The white blood cell count was 16.7x10<sup>9</sup>/L (laboratory reference interval (LRI) 4 – 10). The level of C-reactive protein (CRP) reached 27.4 mg/L (LRI 0 – 5). The abdominal ultrasonography showed picture of acute appendicitis as the appendix had thickened wall and was surrounded by a small amount of liquid. Gross intraoperative findings were suggestive of gangrenous appendicitis. Conventional appendectomy was performed. The postoperative period was uneventful. The patient received analgetic and antibiotic therapy. Fetal movements were monitored and sensed well. Uterus was normotonic. There was no abdominal pain except sensitivity around the surgical wound by palpation. By pathologic examination, the removed appendix grossly measured 6 x 1.2 x 0.8 cm and showed uneven surface. Microscopically, gangrenous appendicitis was revealed along with perforation and wide areas of deciduosus ectopic reaction s. deciduosus (Fig.1), characterised by nodules of discohesive large polygonal cells with widespread degenerative cytoplasmic vacuolisation resulting in signet ring cell like appearance. Acute inflammation with fibrinous and purulent component extended to the periappendicular and mesenteric tissues. By immunohistochemical investigation (IHC), the large polygonal and vacuolated cells in pathologic foci expressed progesterone receptors and vimentin but lacked pan-cytokeratin and calretinin. Thus, IHC confirmed deciduosus and ruled out malignant tumour. The differential diagnosis over the whole course of illness comprised acute appendicitis,

preterm delivery, cancer, acute pyelonephritis or rupture of ovarian cyst. However, considering the intraoperative surgical findings as well as morphological and IHC data, the final diagnosis was appendicular deciduosus, complicated by gangrenous appendicitis and phlegmonous periappendicitis and mesenteriolitis.

## DISCUSSION

Development of decidual cells outside the endometrium, first described by Walker in 1887, is named ectopic decidua or deciduosus (Walker, 1887; Bolat *et al.*, 2012). Such changes most commonly affect ovaries, uterine serosa (Kondoh *et al.*, 2012), fallopian tubes and cervix (Bolat *et al.*, 2012). Deciduosus is less frequently seen in the appendix (Adhikari and Shen, 2013), diaphragm, liver, spleen, paraaortic and pelvic lymph nodes or renal pelvis (Bolat *et al.*, 2012). The involvement of omentum is considered rare by some authors (Adhikari and Shen, 2013), while others suggest that it could be disclosed frequently, if careful sampling would be possible (Buttner *et al.*, 1993; Rodriguez *et al.*, 2006). Deciduosus can be found incidentally in tissues removed or biopsied during a caesarean section, treatment of tubal pregnancy, elective tubal ligation and appendectomy (Bolat *et al.*, 2012).

The pathogenesis of deciduosus is not yet fully understood (Kondoh *et al.*, 2012) and the physiologic nature of this reaction is considered controversial (Bolat *et al.*, 2012). The most frequently suggested explanations include *de novo* development from submesothelial stroma, or decidual transformation of pre-existing endometriosis (Bolat *et al.*, 2012; Kondoh *et al.*, 2012; Adhikari and Shen, 2013). The *de novo* pathway would involve progesterone-related subserosal stromal metaplasia. Confirming the role of hormonal influences, deciduosus regresses within 4 – 6 weeks after pregnancy along with decidual involution (Bolat *et al.*, 2012). High level of progesterone in twin gestations has been attributed to diffuse peritoneal deciduosus in such patients (Adhikari and Shen, 2013). Occasionally, deciduosus in non-pregnant ladies has been explained by adrenal progesterone secretion acting on submesothelial

stroma sensitized by inflammation or trauma (Buttner *et al.*, 1993; Rodriguez *et al.*, 2006). Although decidualis has been identified in patients lacking any history of endometriosis (Bolat *et al.*, 2012), increased awareness of this condition is recommended managing pregnancy in women with history of dysmenorrhea or endometriosis (Kondoh *et al.*, 2012).

Albeit decidualis frequently lacks clinical symptoms, it can manifest by abdominal pain and leukocytosis that has been related to the production of granulocyte colony-stimulating factor (Kondoh *et al.*, 2012). Occasionally, intestinal obstruction (Rodriguez *et al.*, 2006) or significant abdominal or gastrointestinal haemorrhage can develop (Bashir *et al.*, 1995; Kondoh *et al.*, 2012). Lethal outcome of decidualis has been previously reported underlining the necessity of correct diagnosis and timely surgical treatment (Theissig *et al.*, 1988).

In contrast to frequently mentioned asymptomatic course, our patient had pain and leukocytosis. In the presented case, these manifestations can be explained by severe acute inflammation related to rich presence of neutrophilic leukocytes. The gangrenous tissues were widely haemorrhagic. However, there was no evidence of prior primary haemorrhage within the tissues.

The clinical diagnosis of decidualis is difficult even in the presence of widespread intraabdominal involvement (Kondoh *et al.*, 2012). The intraoperative findings can yield broad differential diagnosis including primary or metastatic malignant tumours as decidual mesothelioma, abdominal carcinomatosis or metastatic abdominal spread of malignant melanoma (Ustun *et al.*, 2011; Adhikari and Shen, 2013). Regarding the differential diagnosis with malignant tumours, morphological examination of the removed tissues is the "gold standard". Decidualis is characterised by nodular architecture. The nodules are composed of large cells with well-defined cell borders and eosinophilic cytoplasm. Degenerative changes lead to cytoplasmic vacuolisation, physaliphorous, signet ring or lipoblastic appearance of cells and myxoid stroma. Mitotic activity and/ or nuclear atypia are absent (Rodriguez *et al.*, 2006; Bolat *et al.*, 2012). However, even by light microscopy decidual change may be mistaken for metastatic carcinoma or malignant mesothelioma (Adhikari and Shen, 2013). In such cases, IHC is advisable. Cytoplasmic positivity of vimentin and nuclear presence of progesterone receptors along with lack of pan-cytokeratin, HMB-45 and calretinin is characteristic for decidualis. Although the lesions typically are negative for cytokeratin, few positive cells can be present (Bolat *et al.*, 2012; Adhikari and Shen, 2013). Metastatic carcinoma is characterised by cytokeratin expression, while mesothelioma – by calretinin along with pan-cytokeratin, cytokeratin 5/6 and vimentin. HMB-45 expression is almost diagnostic of melanoma (Ustun *et al.*, 2011; Bolat *et al.*, 2012; Adhikari and Shen, 2013).

Appendectomy is the most common non-obstetric surgical intervention in pregnant patients. The incidence of acute appendicitis in pregnancy is estimated as 0.05 –

0.13% (Kirshtein *et al.*, 2009; Chung *et al.*, 2013). Thus, appendicitis must be considered as the cause of acute abdomen during pregnancy. Cases of decidualis manifesting by abdominal pain, leukocytosis and elevated CRP level necessitate even more careful consideration of appendicitis in the differential diagnosis (Kondoh *et al.*, 2012). The history of the described case shows an additional novel, clinically highly important fact – the appendicitis can also be true complication of decidualis.

In conclusion, decidualis is a rare, pregnancy-related process that in rare cases can affect appendix. In our patient, gangrenous inflammation supervened necessitating urgent appendectomy. Thus, decidualis should be considered in the differential diagnosis of acute abdomen in pregnancy.

**Conflict of interest:** None

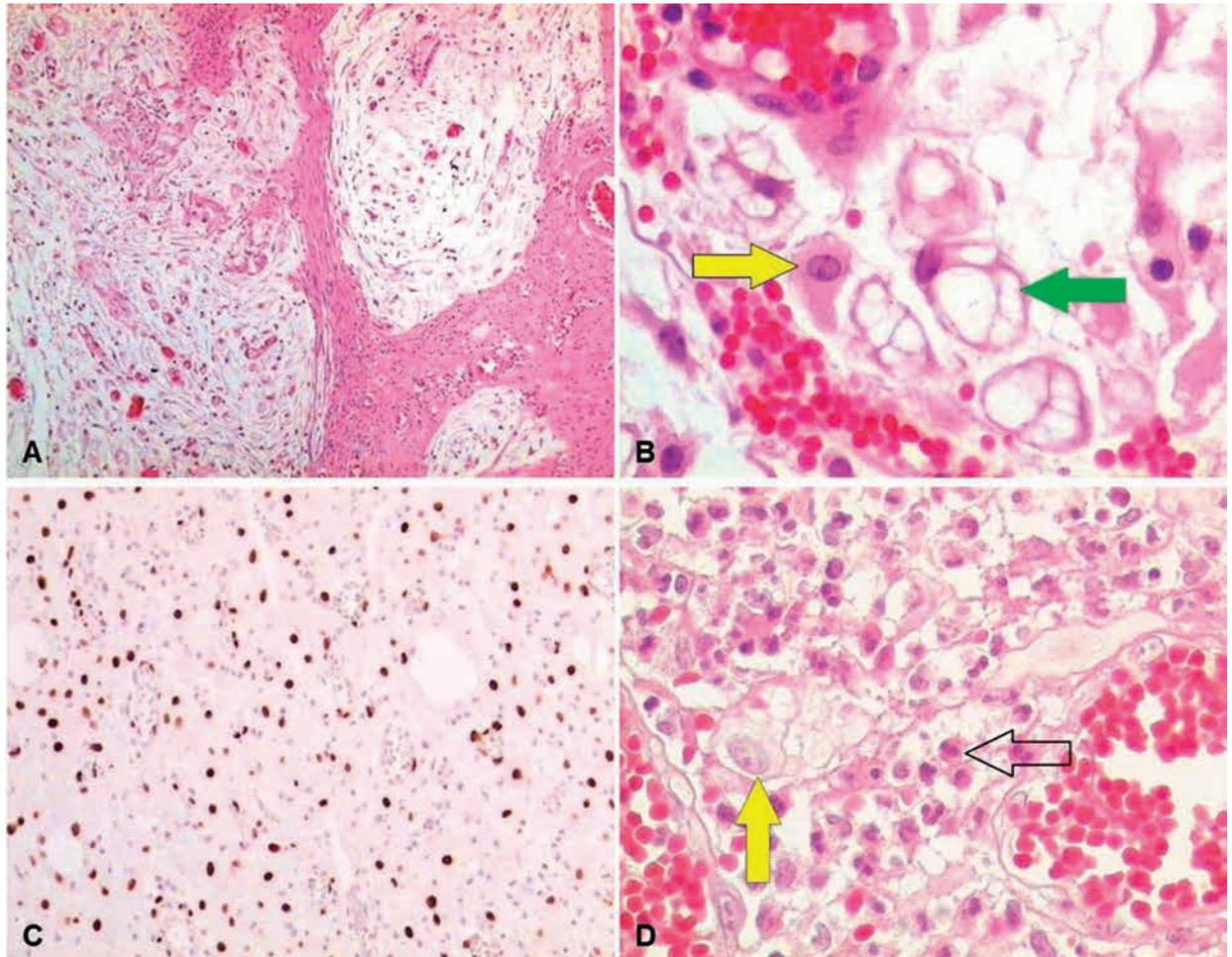
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**Fig. 1. Appendicular deciduosis. A, Overview of the affected tissue. Note the nodular foci of deciduosis. Haematoxylin-eosin (HE), original magnification (OM) 50x. B, Cell morphology of the decidual foci. Note the intact decidual cell (yellow arrow) and the degenerative changes resulting in signet ring cell like appearance (green arrow). HE, OM 400x. C, Expression of progesterone receptors (PR) in decidual cells. Immunoperoxidase, anti-PR, OM 100x. D, Purulent inflammation (arrow) surrounding a decidual cell (yellow arrow). HE, OM 400x.**