

Deciduos of the appendix: a case report

Deciduose do apêndice: relato de caso

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Abstract

Introduction: Ectopic deciduosis refers to decidual tissue outside the uterus. Ectopic decidua appears on the surface of the female reproductive organs and peritoneum; however, rare cases can be found in the appendix, lymph nodes, lungs, kidneys, and skin. Deciduosis is a benign condition that usually does not cause any symptoms and resolves spontaneously 4-6 weeks after delivery. However, an ectopic decidua involving the appendix wall often causes appendicitis and presents with acute abdominal signs. **Methodology:** The descriptive study of the case report type and the data obtained from the patient's medical record. **Case presentation:** This paper reports a woman aged 29 years with complaints of abdominal pain, like being stabbed in the pit of the stomach and then to the lower right abdomen for two days. A physical examination revealed tenderness in the lower abdomen with a positive McBurney sign. At that time, the patient was pregnant with a gestational age of 20-21 weeks. The patient underwent an appendectomy with the tissue sent to the anatomic pathology laboratory with the presence of decidual cells in the subserous layer and serous fat, and it was concluded as deciduosis of the appendix. **Conclusion:** Based on clinical and laboratory findings, the patient was diagnosed with deciduosis of the appendix.

Keywords: Appendix; Deciduosis; Pregnancy.

Resumo

Introdução: A deciduose ectópica refere-se ao tecido decidual fora do útero. A decídua ectópica aparece na superfície dos órgãos reprodutivos femininos e do peritônio; no entanto, casos raros podem ser encontrados no apêndice, linfonodos, pulmões, rins e pele. A deciduose é uma condição benigna que geralmente não causa sintomas e se resolve espontaneamente 4-6 semanas após o parto. No entanto, uma decídua ectópica envolvendo a parede do apêndice geralmente causa apendicite e apresenta sinais abdominais agudos. **Metodologia:** O estudo descritivo do tipo relato de caso e os dados obtidos do prontuário do paciente. **Apresentação do caso:** Este trabalho relata uma mulher de 29 anos com queixas de dor abdominal, como ser esfaqueada na boca do estômago e depois no abdome inferior direito por dois dias. O exame físico revelou sensibilidade no abdome inferior com sinal de McBurney positivo. Naquela época, a paciente estava grávida com idade gestacional de 20-21 semanas. A paciente foi submetida a apendicectomia com o tecido enviado ao laboratório de anatomopatologia com presença de células deciduais na camada subserosa e gordura serosa, concluindo-se como deciduose do apêndice. **Conclusão:** Com base nos achados clínicos e laboratoriais, o paciente foi diagnosticado com deciduose do apêndice.

Palavras-chave: Apêndice; Deciduose; Gravidez.

Resumen

Introducción: La deciduosis ectópica se refiere al tejido decidual fuera del útero. La decidua ectópica aparece en la superficie de los órganos reproductores femininos y el peritoneo; sin embargo, se pueden encontrar casos raros en el

apéndice, los ganglios linfáticos, los pulmones, los riñones y la piel. La deciduosis es una condición benigna que generalmente no causa ningún síntoma y se resuelve espontáneamente de 4 a 6 semanas después del parto. Sin embargo, una decidua ectópica que afecta la pared del apéndice a menudo causa apendicitis y se presenta con signos abdominales agudos. *Metodología:* Estudio descriptivo del tipo caso clínico y de los datos obtenidos de la historia clínica del paciente. Presentación de caso: Este artículo reporta una mujer de 29 años de edad con quejas de dolor abdominal, como ser apuñalado en la boca del estómago y luego en el abdomen inferior derecho por dos días. Un examen físico reveló dolor a la palpación en la parte inferior del abdomen con un signo de McBurney positivo. En ese momento, la paciente estaba embarazada con una edad gestacional de 20-21 semanas. Al paciente se le realizó apendicectomía con tejido enviado al laboratorio de anatomía patológica con presencia de células deciduales en la capa subserosa y grasa serosa, y se concluyó como deciduosis del apéndice. *Conclusión:* Con base en los hallazgos clínicos y de laboratorio, la paciente fue diagnosticada con deciduosis del apéndice.

Palabras clave: Apéndice; Deciduosis; El embarazo.

1. Introduction

Ectopic deciduosis refers to the presence of decidua tissue outside the uterus. Although its pathogenesis is not fully elucidated, it is thought to originate from subserous stromal cells subjected to progesterone stimulation. Ectopic decidua appears on the surface of the female reproductive organs and peritoneum; however, in rare cases, it can be found in the appendix, lymph nodes, lungs, kidneys, and skin (Sorokin et al., 2020). The hormone progesterone circulating during pregnancy stimulates decidualization of the endometrium. It is characterized by stromal tissue hypertrophy, increased glandular secretion, and vascular proliferation. This cell transformation can occur outside the endometrium, one of which can be found in the appendix, although it is rare (Ghannouchi et al., 2021).

Deciduosis is a benign condition that usually does not cause any symptoms and resolves spontaneously 4-6 weeks after delivery. However, ectopic decidua involving the appendix wall often causes appendicitis and presents with acute abdominal signs. Omental and peritoneal involvement are other manifestations that may mimic peritoneal carcinomatosis (Sorokin et al., 2020).

Only a few cases of deciduosis of the appendix have been reported (Ghannouchi et al., 2021). Although deciduosis often has no clinical symptoms, it can manifest as abdominal pain and leukocytosis (Balta et al., 2014). The histomorphological examination is still the gold standard in diagnosing deciduosis of the appendix (Ghannouchi et al., 2021).

2. Methodology

This is a descriptive study of the case report type. In general, case study research is centered on a phenomenon, which is described in as much depth as possible (Yin, 2017; Pereira et al., 2018). Analysis of the medical record for description of the clinical case, being presented in chronological order, and literature review using scientific databases. Our research ethics committee did not conduct any ethical clearance for the case report due to its already discussed at the clinicopathological conference and approved by the clinician, patient, and pathologist. Following ethical principles, the patient consented to disseminating the data and displaying images of his case for academic purposes through the signing of a Free and Informed Consent Form.

3. Case Report

A 29-year-old woman came to the ER with a referral from Bali Jimbaran Hospital on April 16, 2022, with the main complaint of abdominal pain such as being stabbed in the pit of the stomach and then into the lower right abdomen two days ago, clinical diagnosis at the time of admission for abdominal pain observation suspected acute appendicitis. A physical examination revealed tenderness in the lower abdomen with a positive McBurney sign. At that time, the patient was pregnant with a gestational age of 20-21 weeks.

An examination was carried out at the Clinical Pathology Laboratory of the Udayana University Hospital on April 16, 2022, with a complete blood count of leukocytosis (WBC 13.070/mm³) and Hb 10 gr/dL. The urinalysis found a clear yellow color, +1 erythrocytes, +2 leukocytes, positive amorphous crystals, and positive bacteria.

The appendectomy was performed on April 16, 2022. due to abdominal pain observation, et causa suspected perforated appendicitis during pregnancy G1P0A0 21-22 weeks. The Anatomic Pathology Laboratory of Udayana University Hospital received the tissue on April 18, 2022. The container contained one piece of appendiceal tissue measuring length of 5.5 cm and diameter of 0.9 cm, with periappendiceal fat and hyperemia of the appendix. In the obliterative lumen, slices are blackish (Figure 1).

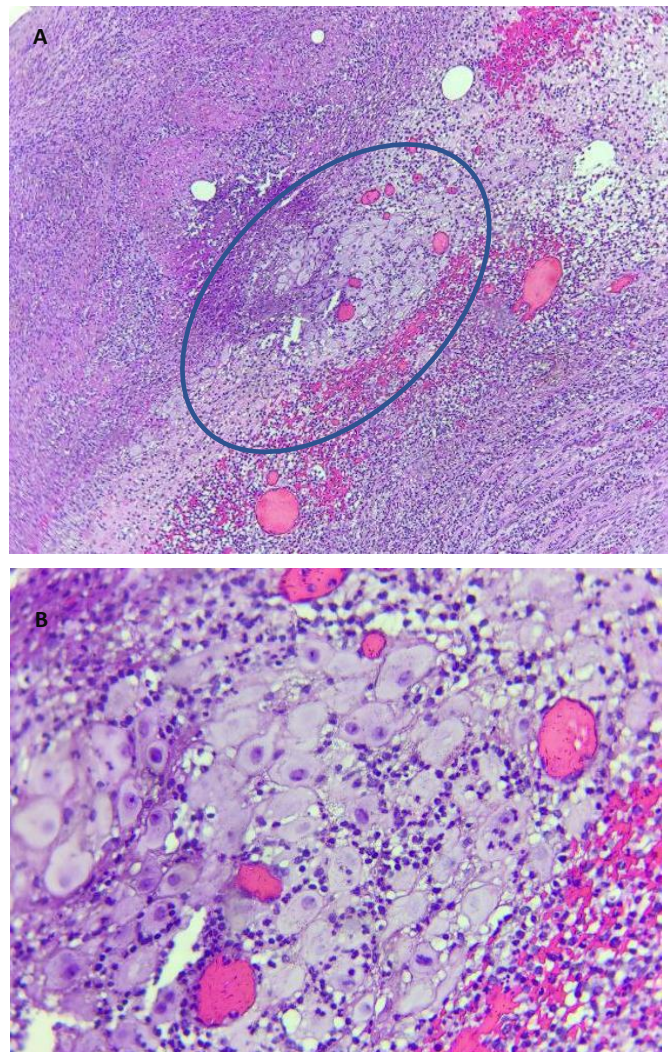
Figure 1 - Macroscopic view of the appendix tissue.



Source: Authors.

On microscopic examination of the proximal and middle tissue sections, it appears that the appendix tissue section contains PMN inflammatory cells, neutrophils, and lymphocytes in the mucosal layer that extends to the serous layer. Some of these inflammatory cells infiltrate the mucosal surface epithelium. Fibrin deposits and dense intraluminal inflammatory cells were also seen. The serous layer appears edematous with the widening of the capillaries. There were also clusters of decidua cells in the subserosa and serous fat layers.

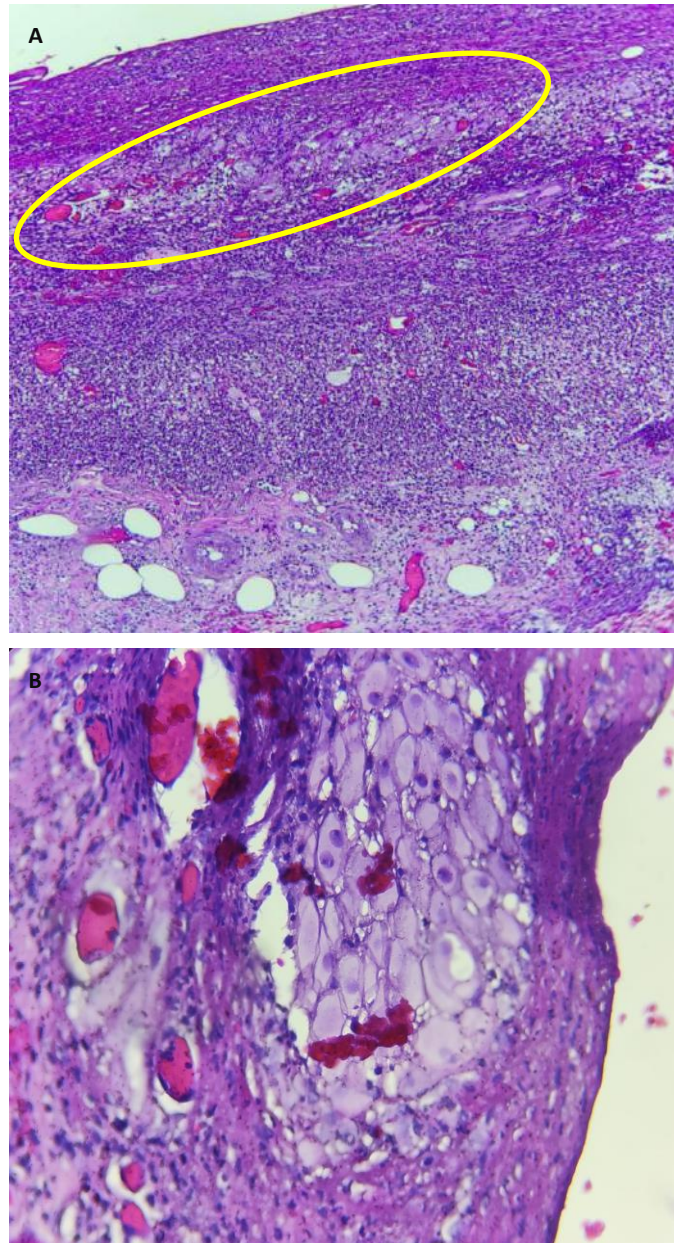
Figure 2 - Microscopic appearance of the proximal and middle sections of the appendix. A. The serous layer appears edematous with dilated capillaries (blue circle mark) (H&E, 100x). B. Groups of decidual cells were also seen in the subserosa layer (H&E, 400x).



Source: Authors.

On microscopic examination of the distal appendix tissue sections, it was seen that the appendix tissue was prepared with a mucous layer containing inflammatory cells of PMN, neutrophils, and intraluminal fibrin. In the muscularis propria and serosa layers, there were scattered and clusters of decidual cells with round to polygonal cell morphology and extensive basophilic cytoplasm, centrally spherical nucleus, hyperchromatic, regular nuclear membrane, mild nuclear pleomorphism.

Figure 3 - Microscopic appearance of the distal tissue section of the appendix. A. In the muscularis propria and serosa layers, scattered and clusters of decidua cells are seen (yellow circle mark) (H&E, 100x). B. The morphology of the decidual cells is round to polygonal, with broad basophilic cytoplasm. Centrally spherical nucleus, hyperchromatic, regular nuclear membrane, mild nuclear pleomorphism (H&E, 400x).



Source: Authors.

Based on the histopathological description above, it is concluded that this case is suitable for decidualosis of the appendix.

4. Discussion

In this case, we report a 29-year-old woman with inflammation of the appendix during pregnancy 20-21 weeks and diagnosis as decidualosis of the appendix after histopathological examination. The literature states that decidualosis, extrauterine decidua, or ectopic decidua is decidua tissue found outside the uterus. Decidualosis outside the uterus is a rare case and most

commonly found on the ovaries, cervix, uterine serosa, and the lamina propria of the fallopian tubes. But it is rarely found on the serous surfaces of the abdominal and pelvic organs such as the peritoneum, omentum, liver, diaphragm, appendix, urinary bladder, small intestine, large intestine, mesentery, paraaortic-pelvic and renal-pelvic lymph nodes (Balta et al., 2014; Chai & Wijesuriya, 2016; Jadhav & Doshetty, 2022; Kaneko et al., 2021; Smits et al., 2020).

The patient presented with a gestational age of 20-21 weeks at the diagnosis. The patient was initially referred from Bali Jimbaran Hospital with a clinical diagnosis of observation of abdominal pain et causa suspected of acute appendicitis. Acute appendicitis is an important differential diagnosis in patients with acute abdomen during pregnancy (Balta et al., 2014; Tsunemitsu et al., 2021). Appendicitis in pregnancy is a rare surgical case, with an incidence ranging from 1: 1250 to 1: 1500 pregnancies (Chai & Wijesuriya, 2016), with the prevalence of acute appendicitis during pregnancy ranging from 0.05-0.13% (Kaneko et al., 2021). According to the literature, decidualis is often associated with pregnancy. The incidence of decidualis generally occurs in the second and third trimesters of pregnancy, with the earliest gestational age at 16 weeks (gestational age range 18-40 weeks), with an age range of 18-40 years (Kaneko et al., 2021; Lier et al., 2017; Meserve, 2019). However, there is no relationship between decidualis and indications for cesarean section (Markou et al., 2016). Regression can be found after 4-6 weeks postpartum (Akçay et al., 2020; Kaneko et al., 2021) but can reappear in the subsequent pregnancy (Luna Aufroy et al., 2018). In addition to pregnancy, decidualis has also been associated with progestin use (Malpica et al., 2020; Nucci, 2019). Ectopic decidualis can also be seen in nonpregnant women and postmenopausal conditions involving the corpus luteum, ovarian stromal cells, and adrenal-derived progesterone (Kaneko et al., 2021).

There are several pathogeneses of the emergence of ectopic decidualis. Ectopic decidua transformation results from "de novo" development of the submesothelial stroma to form nodule-like aggregations or decidua transformation of pre-existing endometriosis during pregnancy (Chai & Wijesuriya, 2016; Daya et al., 2014). In addition, the entrapment or failure of coelomic remnants migration during the embryonic development leads to progesteron-induced subcoelomic mesenchymal pluripotent cell metaplasia during pregnancy also plays a role in the pathogenesis of ectopic decidualis (Adhikari & Shen, 2013; Pigac & Masic, 2016; Smits et al., 2020).

In decidualis of the appendix, there is an increase in pressure due to the narrowing of the lumen of the appendix and inflammation caused by occlusion of the appendix by the decidua tissue or the formation of intraluminal decidual polyps (Ghannouchi et al., 2021; Smits et al., 2020). Patients with decidualis are generally asymptomatic but can also appear to resemble appendicitis (Chai & Wijesuriya, 2016; Kaneko et al., 2021). These symptoms arise due to peritoneal irritation, where endometrial tissue expands in response to increased levels of sex steroid hormones, which causes mechanical compression and irritation of the affected tissue. In addition, it is caused by excessive secretion of microsomal acid phosphatase by decidual cells may increase the synthesis of prostaglandins, causing prostaglandin-mediated contraction of the appendix muscle wall (Chai & Wijesuriya, 2016; Kaneko et al., 2021; Luna Aufroy et al., 2018).

The difficulty in diagnosing acute appendicitis during pregnancy includes the shifting position of the appendix, leukocytosis, and digestive symptoms such as nausea, vomiting, and abdominal pain, which can be found in pregnancy (Chai & Wijesuriya, 2016).

On macroscopic examination, the appendix tissue appeared with periappendiceal fat and hyperemia, with black obliterative lumen slices. In the literature, decidualis tissue appears as pale brown or greyish-white tissue with nodules with a soft or gelatinous consistency (Meserve, 2019).

In this case, the diagnosis of decidualis of the appendix was established after a histopathological examination of the tissue. A histomorphological examination is a gold standard in diagnosing decidualis of the appendix (Ghannouchi et al., 2021). The normal decidua appears as endometrial stromal cells that transform during pregnancy in response to ovarian and placental hormones, particularly progesterone (Chai & Wijesuriya, 2016), characterized by stromal tissue hypertrophy,

vascular proliferation, and glandular secretion (Smits et al., 2020).

On microscopic examination of the appendix tissue in this patient, clusters of decidual cells in the subserosa layer and serous fat were seen in the proximal and middle sections of the appendix, and in the distal section of the appendix, scattered and clusters of decidual cells were seen in the muscularis propria and serosa layers. Inflammatory cells of PMN neutrophils and lymphocytes were also seen in the mucosal layer, which extended to the serous layer, some infiltrating the mucosal surface epithelium. The microscopic examination of the appendix was concluded as decidualization of the appendix. Ectopic decidual tissue found without glands are known as decidualization, while found with glands are known as decidualized endometriosis (Kaneko et al., 2021). In decidualization of the appendix, decidual islands can cover the mucous, muscular, and serous layers of the appendix, which consist of sheets of large polyhedral decidual cells (Luna Aufroy et al., 2018; Noffsinger, 2017). Decidualization is characterized by a nodular architecture consisting of large polygonal cells with eosinophilic cytoplasm and round nuclei with prominent nucleoli. Decidual cells are usually perivascular (Ghannouchi et al., 2021). No mitotic activity and/or nuclear atypia was found. Decidual cells are commonly found beneath the mesothelium in the subcoelomic mesenchyma or adipose tissue (Smits et al., 2020).

In this case, immunohistochemistry was not required due to the characteristic histopathological findings. In patients with atypical histopathology, immunohistochemical examination for decidualization can be performed. Decidual cells may express vimentin, desmin, CD10, and/or smooth muscle actin, as well as progesterone and estrogen receptors. Immunohistochemical examination for keratin, calretinin, S100, CEA, and EMA is usually negative (Ghannouchi et al., 2021; Malpica, 2021; Noffsinger, 2017; Smits et al., 2020). The immunohistochemical examination confirms the nature of the decidual tissue, in which the decidualization exhibits pseudotumoral and infiltrative characteristics. The examination is to rule out other differential diagnoses in the form of primary malignant tumors or metastatic malignant tumors such as decidual mesothelioma, abdominal carcinomatosis, metastatic melanoma, benign peritoneal decidualization, signet ring cell adenocarcinoma, epithelioid leiomyosarcoma, rhabdomyosarcoma, and placental site trophoblastic tumor (Adhikari & Shen, 2013; Smits et al., 2020).

Appendectomy surgery was performed due to abdominal pain observation et causa suspected perforated appendicitis on April 16, 2022. In decidualization of the appendix, the condition with pain and intraperitoneal bleeding is a condition that requires surgery (Kaneko et al., 2021). There are several complications due to delaying surgery on decidualization of the appendix, namely perforation of the appendix, with a percentage of 14-43% in pregnant women and fetal death reaching 3-36% of cases (Kaneko et al., 2021). In addition, decidual bleeding is a potential risk factor for preterm labor (Lier et al., 2017).

5. Conclusion

We report a case of decidualization of the appendix in a woman in the second trimester of pregnancy. Decidualization of the appendix is a rare case and difficult to distinguish from acute appendicitis clinically, despite the use of various available modalities. It should be noted that decidualization of the appendix is a differential diagnosis that needs to be considered in the suspicion of acute appendicitis in pregnant women. Histopathological examination is the gold standard in diagnosing decidualization. For those reasons, clinicopathological correlation with a highly experienced pathology field will provide an accurate diagnosis of decidualization of the appendix.

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