Case Report

Appendicular endometriosis as a cause of chronic abdominal pain alone in the right iliac fossa: case report and literature review

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ABSTRACT: Endometriosis is a disease characterized by the presence of ectopic endometrial glands and stroma. Although its etiology is undefined, it is suggested to be a result of coelomic metaplasia, retrograde menstruation, to provide a genetic component, or to be one that occurs due to blood or lymphatic spread. The involvement of the gastrointestinal tract is common. However, appendicular endometriosis is a rare condition. It is usually asymptomatic. Recurrent pain in the right iliac fossa is an unusual clinical manifestation. There are no non-invasive complementary tests to confirm the diagnosis. Laparoscopy is the main option for research, due to its diagnostic and therapeutic features. A histopathological examination is necessary for the diagnosis. Although surgical and drug therapies have special indications, the combination therapy showed lower symptom recurrence. This study reports a case of appendicular endometriosis that was diagnosed and treated in the service of Coloproctology of the Base Hospital at ***Faculdade de Medicina*** of São Jose do Rio Preto. There is also a literature review about this situation.

Keywords: endometriosis; gastrointestinal tract; appendix; abdominal pain.

RESUMO: Endometriose é uma doença caracterizada pela presença de estroma e glândulas endometriais ectópicas. Apesar de sua etiologia não definida, sugere-se que seja decorrente de metaplasia celômica, menstruação retrógada, apresente componente genético, ou ocorra devido à disseminação linfática ou sanguínea. O acometimento do trato gastrointestinal é comum; no entanto, a endometriose apendicular é condição rara e se apresenta com maior frequência de forma assintomática. Dor recorrente em fossa ilíaca direita é uma manifestação clínica incomum. Não há exames complementares não invasivos que confirmem o diagnóstico. A laparoscopia é a principal opção durante a investigação, por sua característica diagnóstica e terapêutica. O diagnóstico pode ser feito apenas após um exame histopatológico. Embora as terapias medicamentosa e cirúrgica apresentem indicações particulares, a terapêutica combinada mostra menor recorrência dos sintomas. O objetivo do trabalho é relatar um caso de endometriose apendicular diagnosticado e tratado na Disciplina de Coloproctologia do Hospital de Base da Faculdade de Medicina de São José do Rio Preto, além de revisar a literatura acerca dessa situação.

Palavras-chave: endometriose; trato gastrointestinal; apêndice cecal, dor abdominal.

INTRODUCTION

Endometriosis is characterized by the presence of stroma and endometrial glands outside the uterine cavity. It can be called internal endometriosis or adenomyosis, when the endometrial tissue is inside the uterine muscle, or external endometriosis, if there is extrauterine ectopic implant[1]. The latter divides into intraperitoneal (blatter, Douglas' Pouch, ovaries, sigmoide, appendix) and extraperitoneal (umbilicus, surgical scars, vagina, vulva, lungs)[2].

It affects females aged 25 to 45 years, with estimated incidence rates of 4 to 17% among women at

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reproductive age[3,4]. One third of the infertility cases among females are associated with endometriosis[5].

The etiology of the disease is not defined. Among the most accepted theories, coelomic metaplasia is mentioned[6], as well as retrograde menstruation, proposed by Sampson[7], and the genetic theory, besides blood and lymphatic spread.

Ectopic endometrial implants are usually present in the pelvic region, especially the pelvic peritoneum, ovaries, fallopian tubes, and rectovaginal septum[3]. The involvement of the gastrointestinal tract is common, ranging from 3 to 37% of the cases[1,3,8]. Bowel endometriosis affects the rectum and the rectosigmoid junction in 70 to 85% of the cases[1,3,9]. The small intestine involvement is around 7% more likely restricted to the terminal ileum. The less affected sites are the cecum (3.6%) and the appendix (3%)[3,10].

CASE REPORT

SMASR, 44 years old, mother of five, presented with abdominal pain in the right iliac fossa, which lasted for two years. The pain was continuous, mild and worsened in the last four months. There was no gynecological or gastrointestinal complaint, nor any relation with the menstrual cycle. At first, a total abdominal ultrasound was performed (18th day of the menstrual cycle), which revealed a hypoechoic oval image, with echogenic center and regular forms, measuring 2.0x1.4 cm, located between the cecum and the right attachment.

Due to the lack of definition in this case, a magnetic resonance imaging test was requested and revealed an oval and heterogeneous image, with intense central signal, intense signals in the right iliac fossa, with intimate contact with the cecum and small intestine loops, measuring around 2.4x2.1x2.2 cm. After the infusion of the contrast, there was intense pervading (Figure 1).

Tumor markers/antigens associated with cancer 125 (CA 125) and carcinoembryogenic antigens (CEA) were normal, as well as the thoracic x-ray. Colonoscopy revealed invaginated appendix (Figure 2).

Because it was impossible to exclude the hypothesis of neoplasm, a right colectomy was performed, associated with ipsilateral salpingo-oophorectomy. The mass had approximately 4 cm in diameter. The biopsy of the surgical specimen confirmed the diagnosis of endometriosis (Figure 3).

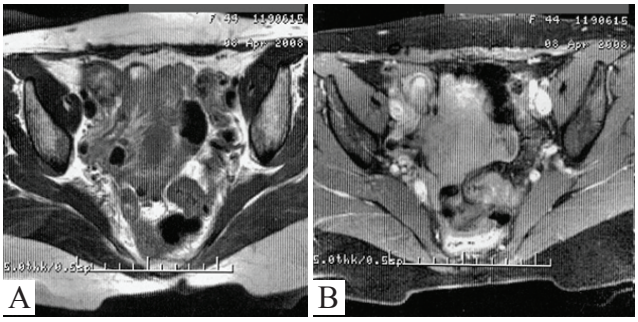

Figure 1. *(A) and (B) Mass in the right iliac fossa at magnetic nuclear resonance.*

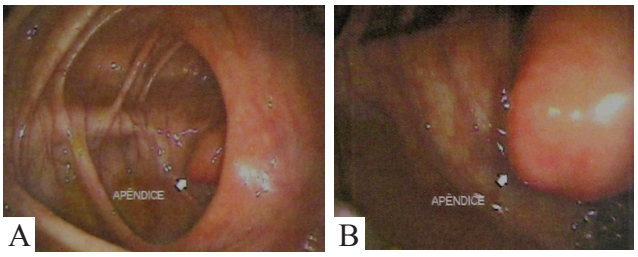

Figure 2. *(A) and (B) Invaginated appendix at colonoscopy.*

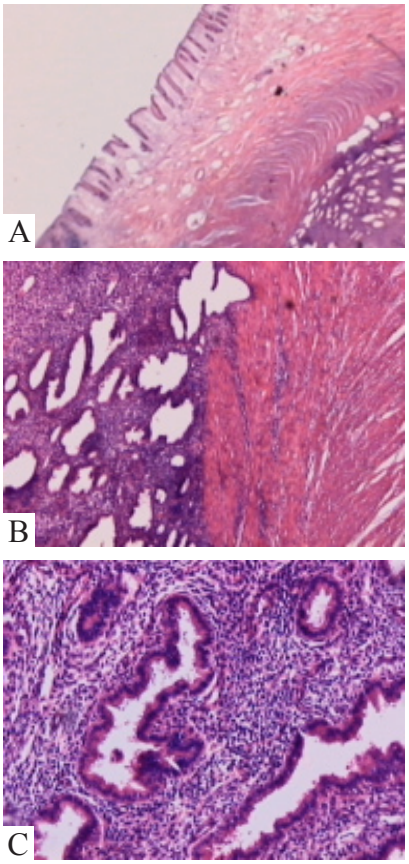

Figure 3. *(A) Aspect of endometriosis. (B) Relation between endometrial tissue and the muscular layer of the appendix. (C) Stroma and endometrial glands.*

The patient recovered well after surgery. Follow-up had no signs of recurrence or complications.

DISCUSSION

Endometriosis affecting the ileocecal appendix is rare. The clinical picture is complex, and may be categorized into groups according to the symptoms: asymptomatic patients, those with acute appendicitis, patients presenting invagination of the appendix, those with appendicular perforation and also the ones with atypical symptoms (abdominal pain, nausea and melena)[1,11]. Also, the chronic obstruction of intestinal lumen may lead to the formation of mucocele or inflammatory periappendicular mass, which is hard to distinguish from neoplasm[12].

The laboratory evaluation for CA 125 dosage presents low sensitivity and specificity due to high serum concentrations in other affections, thus, it is not a possible diagnosis[12].

No radiological examination can diagnose ileocecal appendicular endometriosis[13]. At ultrasound, it is possible to see cystic, solid or complex images due to the hormonal response of the ectopic tissue in the appendix, while computed tomography shows distended appendix with no inflammation. However, they help to rule out pathologies that manifest the same way[14].

Laparoscopy is the diagnostic test of choice in cases of endometriosis because it enables the direct visualization of the affected site, revealing macroscopic characteristics, determining the extension of the lesions and providing material for the histopathological investigation. Besides, it is also a therapeutic method[11].

The diagnosis is confirmed by a histopathological evaluation. Lesions are usually superficial and small, located on the third distal and, sometimes, with weak adherence, similar to what happens in the tubes affected by endometriosis[11]. The mucosa of the organ is rarely affected. The endometrial tissue in the muscle and the seromuscular layer is present in 2/3 of the cases, and the affection of the serum layer is present in 1/3 of these cases[1].

The differential diagnosis of appendicular endometriosis should include diverticular disease, colorectal carcinoma, inflammatory bowel disease, carcinoid tumors, benign intramural neoplasm, occult intra-abdominal metastases, mesenteric neoplasm and pelvic abscess[13].

Since the ectopic endometrial tissue responds to ovarian hormones, estrogen and progesterone, patients with few implants clinically respond to hormonal therapy. In cases of advanced lesions, such as the ones with nodular proliferation, hormonal therapy leads to partial regression within several months of treatment. However, none of the drug therapies completely eradicates endometrial implants, that is, after the discontinuation of medicines, symptoms may return. Also, the resulting collateral effects may inhibit the adhesion to the treatment[13]. Since the CA 125 dosage is correlated with the severity of the disease, it has proved to be useful to assess the response to clinical treatment[12].

Patients with obstructive bowel symptoms who are refractory to clinical treatment, or those cases in which it is not possible to exclude the neoplastic etiology, present surgical indication. Cases of appendicular endometriosis should be treated as appendicectomy[12]. Also, surgical resection prevents the recurrence of symptoms and the late development of local endometrial carcinoma, which is a possible but not common complication[13]. However, surgical treatment does not remove the microscopic disease. So, CA 125 dosage after resection in women who presented high levels prior to the procedure is important for the follow-up, in case of recurrence of the symptoms[12].

The recommendation for olingosymptomatic patients who are younger than 40 years old and manifest the wish to get pregnant is conservative surgical therapy, which consists of partial oophorectomy, besides the resection of the appendix[2]. Surgical removal of the appendix is confirmed by the correlation reference between appendicular endometriosis and infertility[11]. The radical surgical treatment is recommended for patients aged 40 years or more, or for those who are younger but do not respond to clinical therapy or conservative surgery, presenting with severe symptoms. It consists of bilateral oophorectomy and hysterectomy, with improvement of the symptoms in 90% of the cases[2].

It is important to observe that, although clinical and surgical treatments have very specific indications, in most cases the combined therapy presents best results to eradicate the disease and minimize recurrences[12]. Preoperative drug therapy for a period of three to six months enables the reduction of endometrial implants, their vascularization and also of the inflammatory process, besides reducing the formation of adhesions[12].

CONCLUSION

Endometriosis affecting the ileocecal appendix is a rare condition, being mostly asymptomatic. Due to the need for histopathological confirmation, because of the variety of clinical presentations and the absence of complementary non-invasive tests that suggest this affection, preoperative diagnosis is extremely difficult. In this context, laparoscopy is important because of its diagnostic and therapeutic features. Preoperative clinical treatment and the continuation of drugs for a long postoperative period are related to the lower recurrence of symptoms.

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