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Case Report

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Primary Umbilical Endometriosis. A Case Report and Literature Review

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ABSTRACT

Introduction: Endometriosis a disease condition characterized by the presence of endometrial glands and stroma outside the uterine cavity and musculature. It affects up to 7-10% of women in their reproductive age and usually involves pelvic organs. The common symptoms include dysmenorrhea, menorrhagia, pelvic pain, dyspareunia and infertility. Umbilical endometriosis is the rarest form of extra pelvic endometriosis. Secondary umbilical endometriosis occurs after abdominal procedures while primary umbilical endometriosis develops without previous procedures and the cause is barely known. Metaplasia of urachal remnant is possible explanation of primary umbilical endometriosis.

Case Presentation: A 32years old para two mother presented with umbilical swelling and pain which worsens during menses of 2 months. She had 2 cesarean scar both transverse pfannenstein incision. There was dark blue firm to hard 2x3cm umbilical mass on physical examination. CBC and OFTs results are normal. Ultrasound of the abdomen showed umbilical mass with homogeneous echotexture measuring 2 cm × 3 cm with irregular border and hypoechoic texture. FNAC suggested a diagnosis of umbilical endometriosis. The case was jointly discussed with obstetrics and gynecology side. After decision to excise the mas was made informed written consent was taken and the patient was taken to Operation Theater the mass was excised enblock with surrounding tissue and underlying fascia. The umbilicus was reconstructed. The specimen was sent for biopsy and the result confirmed the diagnosis.

Discussion: Primary umbilical endometriosis is a disorder first described by Villar in 1886 and among all cases of extra genital involvement of endometriosis primary umbilical endometriosis accounts for 0.5 t0 1%. The commonest site of extra-genital endometriosis is the pelvic organs which present with pelvic pain. The usual presentation of primary umbilical endometriosis is typically the presence of a discrete bluish-purple mass in the umbilicus which becomes swollen, painful, and bleed concomitantly with the menstrual cycle. Ultrasound and other imaging techniques help in making a diagnosis and the diagnosis is confirmed by cytological examination. The management of umbilical endometriosis is surgical excision and reconstruction of the umbilicus.

Conclusion: umbilical endometriosis is an uncommon form of extra pelvic endometriosis. There is a wide differential diagnosis for swellings of the abdominal wall, however, it should be suspected in any female patient of reproductive age with umbilical lesions, which become more painful and swollen during her menstrual period. Appropriate clinical examination and work up are helpful to make a diagnosis and do surgical excision. In addition, attention should be given to ruling out the coexistence of pelvic endometriosis and infertility.

Introduction

Endometriosis is a term first used by Sampson for a gynecologic complaint condition that is characterized by the presence of endometrial glands and stroma outside the uterine depression and musculature [1]. This problem occurs in 7 - 10 % of women of reproductive age and generally, it affects the pelvic organs [1,2]. The common symptoms of endometriosis include dysmenorrhea, menorrhagia, pelvic pain, and gravidity, dyspareunia [1,2]. Still, endometriosis may involve non-pelvic organs, and particularly in cases who had gynecologic procedures the circumstance of abdominal wall endometriosis is n0.03 –1.08 [2]. The common extrapelvic spots include the diaphragm, pulmonary, urinary tract, gastrointestinal tract, brain, and cutaneous endometriosis. Of all extrapelvic endometriosis, umbilical endometriosis comprises 0.5- 1. Umbilical endometriosis can be primary or secondary when it occurs following laparoscopic or open procedures. Primary umbilical endometriosis is also called Villar's bump because it was first described by Villar in 1886. The pathogenesis of endometriosis is slightly understood. One of the possible pathogenesis is supposed by Sampson, the so-called Sampson's proposition of retrograde period, others include coelomic metaplasia, induction proposition, embryonic Mullerian rests, bone gist stem cell proposition, and hematogenous and lymphatic spread [3]. In the case of umbilical endometriosis hematogenesis or lymphatic proposition is favored where there'sco-existing pelvic endometriosis. still, insulated umbilical endometriosis is explained by metaplasia of urachal remnants [3].

Case Presentation

A 32-year-old female patient presented with umbilical swelling and pain of 2 months duration. The swelling was gradually

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increasing in size and the pain worsens when she is on menses. Her menses are regular and come every 28 days. She has no pelvic pain or other compliant. She has two children, one is a female 11 years old and the other male child 9 years old, both delivered through the caesarian section. She did not conceive for 9 years despite not taking any contraceptives. On physical examination, she was well-looking. Her BP120 R 82 RR20 T 36.5 So2 95% on atmospheric oxygen.

Abdominal examination revealed a 3x2cm firm irregular dark blue umbilical mass. The mass is fixed to the underlying structures and the overlying skin. Laboratory investigations show a normal range of complete blood count and organ function tests. Her urine hcG is negative. The abdominopelvic ultrasound finding was described as an umbilical mass with homogeneous echotexture measuring 2 cm × 3 cm with irregular border and hypoechoic texture. FNA cytology smears show a clean background with flat sheets of uniform cells with oval to round nuclei, surrounding stromal tissue fragments, and pigment-laden macrophages consistent with endometriosis [Figure 2].

The case was discussed with the obstetrics and gynecology side. After the decision to excise the mass was made informed written consent was taken and the patient was taken to Operation Theater the mass was excised en-block with surrounding tissue and underlying fascia. The fascia was closed and the umbilicus was reconstructed. The specimen was sent for biopsy. The pathological analysis and microscopic sections of the specimen show skin covered tissue with underlying endometrial glands and stroma embedded in dense fibrous stroma (Figure 2). At the time this case report was written the patient had follow up visit, two months after the surgery. She has no complaints and on examination the wound is healed but there is dark discoloration of the skin around the scar [Figure 1].

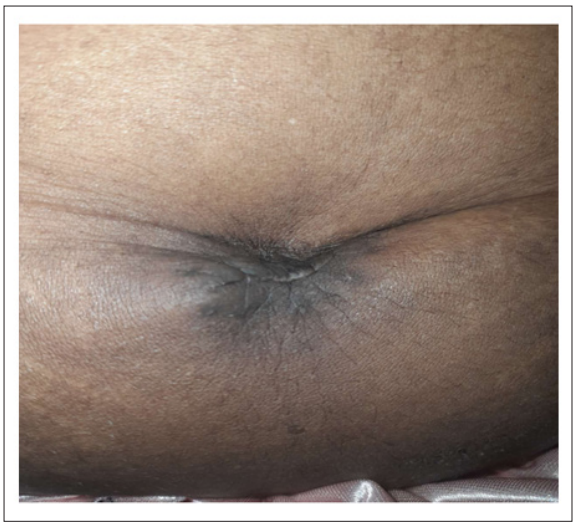

Figure 1: A post-excision and reconstruction picture showing a scar with dark discoloration

Discussion

Primary umbilical endometriosis is a complaint first described by Villar in 1886 and among all cases of redundant genital involvement of endometriosis primary umbilical endometriosis accounts 0.5 to 1. The condition affects 6% of premenopausal women [4-5]. The presence of extra-pelvic endometriosis can affect multiple organs and organ systems. The spots involved from utmost frequent point to least include the intestine, skin(including the umbilical and abdominal scars), inguinal region and ham, lungs and pleura, pancreas, meninges and chines. Overall, cutaneous and subcutaneous endometriosis is believed to be caused by a cicatricial process following abdominal and/ or pelvic surgical procedures, similar as laparoscopy/ laparotomy, cesarean section, hysterectomy, myomectomy, episiotomy, and appendectomy, junking of Bartholin's gland tubercle, amniocentesis and intrauterine injections for revocation [6].

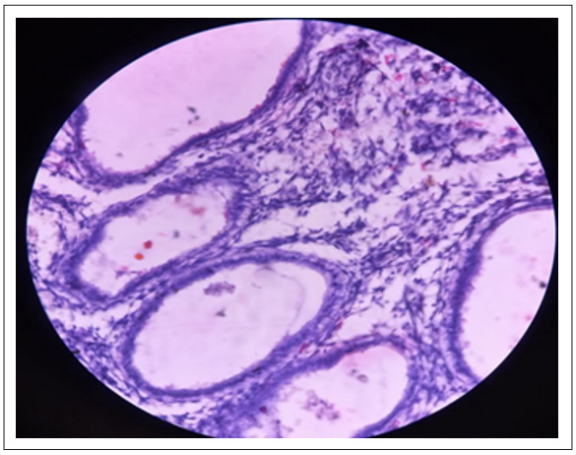

Figure 2: A picture of the biopsy slide demonstrating endometrial glands and stroma

In 1920, Sampson proposed a proposition that endometriosis is caused by a retrograde period passing through the fallopian tube in the pelvis. Other proposed mechanisms include [6]. The ectopic presence of endometrial stem cells or vulnerable system blights. Examinations into the inheritable threat factors for endometriosis revealed that differences to p53, PTEN, Cytochrome P450 1A1, and Peroxisome proliferator- actuated receptor γ2Pro-12-Ala have been intertwined in endometriosis [7]. Implantation of an endometrial towel into a surgical gash has been proposed as an explanation of the condition's pathophysiology. It's allowed during a cesarean delivery, endometrial towel is invested into the gash [8]. Umbilical endometriosis may be incorrect for discrimination opinions like abdominal wall abscess, lipoma, hematoma, granuloma, neuroma, sebaceous tubercle, incisional hernia, carcinoma, sarcoma, etc [8]. Ectopic endometriosis is a rare event, it manifests as a painful subcutaneous mass that worsens during monthlies and generally bothers the case several times. In addition, endometriosis associated with scars may have nasty changes [9]. Our case had caesarian section 11 times and 9 times back both indicated due to her pelvic periphery. In both cases, the gash was a transverse Pfannenstiel gash c- C-section and there was no surgical procedure involving the umbilicus during both procedures. Her umbilical swelling and symptoms appeared after 9 years of operation but in a non-scared umbilicus. She had a 3x2cm establishment irregular dark blue umbilical mass. In another analogous case reported by Yahaya and associates typical symptoms of umbilical endometriosis were described as the presence of a separate bluish-grandiloquent mass in the umbilicus which becomes blown, painful, and bleeds concomitantly with the menstrual cycle. Opinion of umbilical endometriosis involves FNAC, vivisection, and immunohistochemistry [10]. In the case we present the opinion was made by clinical examination, ultrasound, FNAC, and vivisection. The treatment of umbilical

endometriosis according to several former reports is that en-bloc excision of umbilical endometriosis lesions, including the peritoneum, and the posterior reconstruction of the umbilicus, guarantees the radicality of the procedure [11]. In most of these cases with umbilical endometriosis that have been reported in the literature, surgical treatment was performed successfully with a veritably low rate of rush. Still, there is little experience concerning medical treatments. Some suggest long-term treatment with progesterone, danazol, and norethisterone all of which showed disagreeing results in veritably many cases, gonadotropin-releasing hormone analogs and nonstop treatment with a monophasic OC have noway been applied to cases with umbilical endometriosis [12]. In the case of the present case, surgical excision and reconstruction was done and until the time this case was written, she only had dark skin abrasion. She was not able to conceive for 9 years. This may strengthen the substantiation that umbilical endometriosis may contribute to infertility [13].

Conclusion

umbilical endometriosis is an uncommon form of extra pelvic endometriosis. There is a wide differential diagnosis for swellings of the abdominal wall, however, it should be suspected in any female patient with a lesion, which becomes more painful and swollen during her menstrual period. Appropriate clinical examination and investigation help make a diagnosis and do surgical excision. In addition, attention should be given to ruling out the coexistence of pelvic endometriosis and infertility.

Informed consent

Ethical approval

For publication of a case report at our institution, Jigjiga University, Shek Hassen Yabare Referral Hospital, ethical approval is not necessary. This is because the patient is not involved directly in the study with interview or any other intervention. The identity of the patient is also protected and not mentioned in the case report.

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Author Contribution

Sintayehu Asrat, MD, general surgeon,assistant professor of surgery, department of surgery, jigjiga university, Ethiopia. Involved in conception and design of, drafting and revising of the article and final approval of the version to be submitted and involved in the the management and follow up of the patient.

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Conflict of Interest

The authors have no conflict of interest to declare.

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