Primary Umbilical Endometriosis

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ABSTRACT

Endometriosis is presence of ectopic endometrial tissue outside uterine cavity. Although it is rare, foci of endometrial tissue can present in extra pelvic sites such as umbilicus, lungs, abdominal surgical scars and bowel. Umbilical endometriosis is a rare condition with an unclear pathophysiology. It is characterized by a discolored nodule in the umbilicus, which presents as cyclical menstruation symptoms like pain, swelling and bleeding. Imaging and histopathological examination can predict and confirm the diagnosis. In Nepalese literature, extra pelvic and pelvic site endometriosis have been explored, however there are no case reports of primary umbilical endometriosis. Therefore, we describe a case of a 38-year-old woman who presented with umbilical bleeding. The diagnosis of endometriosis was confirmed by clinical, radiological, and histopathological examination.

KEY WORDS

Endometriosis, Primary umbilical endometriosis, Umbilicus

INTRODUCTION

Endometriosis is a benign gynecologic disease that affects women of reproductive age. It is defined as the presence of endometrial tissue, glands and stroma located outside the uterine cavity. It is often associated with fibrosis and inflammatory reaction.[1] The most common sites of endometriosis are the ovaries and fallopian tubes but it can develop in any organ such as lung, liver, brain, skin, umbilicus and abdominal wall.[2] Umbilical endometriosis which is also known as Villar's Nodule is a rare entity. The prevalence of spontaneous umbilical endometriosis is between 0.5% and 1.0%.[3,4] Periodic bleeding from the umbilicus is the most common presenting symptoms and most of the patient have history for 1-2 years' duration.[4] Umbilical endometriosis secondary to surgical scars have been reported. However, primary umbilical endometriosis has not been reported yet in the context of Nepal. So we are reporting a rare case of primary umbilical endometriosis.

CASE REPORT

A 38 years old female, gravida 4 para 2 presented with a history of recurrent cramping pain and episodic bloody discharge from the umbilicus during menstrual cycle for one and half years. She had previously undergone two medical terminations for pregnancy. There was no history of any surgical procedure. Her menstrual cycles were regular. Other medical history was unremarkable except for dysmenorrhea. Physical examination revealed raised lesion in the center of her umbilicus measuring 0.5 cm. The lesion appeared dark bluish in color. There was no any scar, ulceration or discharge. Patient underwent Magnetic resonance imaging which revealed homogeneously enhancing altered signal intensity lesion measuring 38x19mm arising from umbilicus involving subcutaneous plane on the right side (Fig. 1).

Figure 1. Homogenously altered signal intensity lesion arising from the umbilicus (T1 weighted MRI)

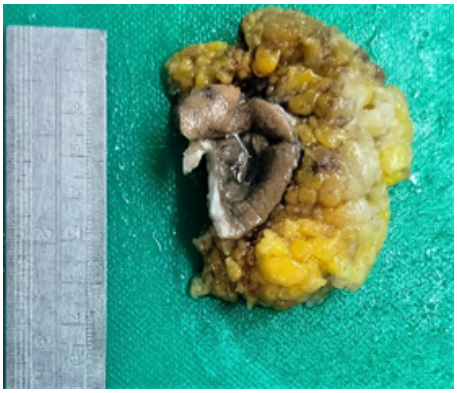

Figure 2a. Raised lesion in the center of the umbilicus on gross examination

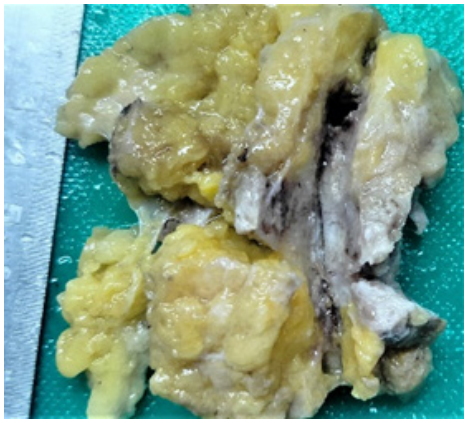

Figure 2b. Grey white areas with hemorrhagic specks on cut section

Wide local excision of the lesion was done and specimen was sent to department of pathology for histopathological examination. Gross examination showed skin covered fibro fatty tissue with nodular lesion measuring 2x2 cm in the center of umbilicus. Skin ulceration was not seen. Surrounding skin and fibro fatty tissue appeared unremarkable. Cut sections showed grey white areas with hemorrhagic specks (Fig. 2a and 2b). Microscopy revealed endometrial glands and stroma within the dermis of underlying skin along with normal adnexal structures (Fig. 3a and 3b). The lesion was diagnosed as primary umbilical endometriosis considering clinical, radiological and histopathological examination.

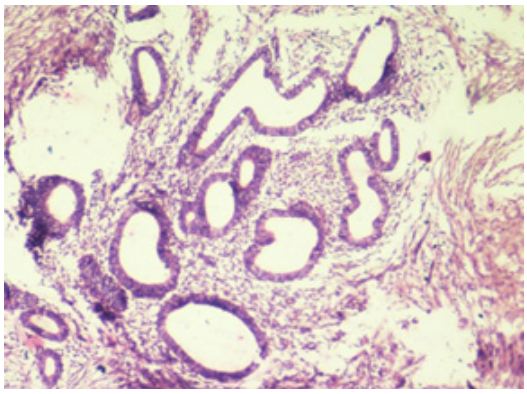

Figure 3a. Endometrial glands and stroma on H&E, 10X

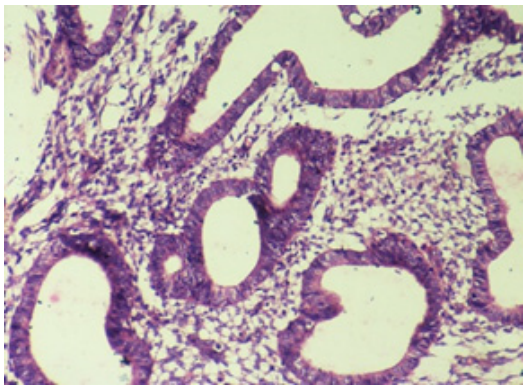

Figure 3b. Endometrial glands and stroma on H&E, 40X

DISCUSSION

Ectopic endometrial tissue indicates the existence of endometriosis. When it occurs inside the myometrium, it is referred to as adenomyosis, and when it occurs outside the uterus, it is referred to as endometriosis.[5] The prevalence of endometriosis is between 2-10% of general female population.[3] The most common site of endometriosis is ovaries followed by scars, cervix, fallopian tube, vagina and appendix in descending order of frequency in one study conducted in Nepal.[6]

It is unclear exactly how endometriosis develops. However, there are several theories explaining how endometriosis develops. According to the notion of embryonic rest, endometriosis arises from wolffian or mullerian duct remnants that serve as a forerunner to ectopic uterine tissue. According to the Coelomic Metaplasia Theory, certain inflammatory, hormonal, or traumatic stimuli might cause the embryonic coelomic mesothelium to undergo dedifferentiation into endometrial tissue. The most widely recognized hypothesis of migratory pathogenesis postulates that endometrial tissue can spread by vascular and lymphatic routes, direct extension, and surgical manipulation.[3,7] Endometriosis has been linked to changes in p53, PTEN, cyt P450 1A1, and PPARγ2. Additionally, implicated are prolonged radiation exposure and increasing estrogen exposure.[8]

Umbilical endometriosis is a distinctly rare entity. Umbilical endometriosis can be misdiagnosed as a cyst, abscess, lipoma, stitch granuloma, melanoma, or metastatic deposits. Umbilical endometriosis may be linked to an underlying hernia, making diagnosis more difficult.[7] When an adult patient has spontaneous umbilical bleeding or discharge, the cause may be an incarcerated umbilical hernia with omental varices brought on by portal hypertension, an umbilical hernia with fat necrosis, embryological remnants, omphalitis, or metastases.[9] Umbilical endometriosis has the potential to transform into an adenocarcinoma and become cancerous.[10]

Depending on whether it develops spontaneously or as a result of prior surgery, umbilical endometriosis can be primary or secondary.[11] In our case there was no history of previous caesarean section delivery or any other surgical manipulations which further confirmed primary umbilical endometriosis. According to Hirata et al. the median age at presentation was 39.2 years.[10] Before diagnosis, there had been symptoms for 1-2 years.[4] The previous findings are consistent with the present case in study. All premenopausal females who present with umbilical mass, discharge, discomfort, or cyclical bleeding from the umbilicus should be suspected of having umbilical endometriosis.[7] It often manifests as a single flesh-colored, brownish, or reddish nodule. The lesion may bleed or enlarge, which are common signs.[12] Menstruation is frequently linked to symptoms. But symptoms might not be associated with menstruation.[10] Our case also had similar clinical features associated with menstruation.

The formulation of a preliminary diagnosis is aided by the history and physical examination. However, the size of the nodule, the degree of subcutaneous tissue involvement, and the pathology of the surrounding tissues may all be determined by ultrasound. When endometriosis is suspected, MRI can be utilized as an imaging modality to assess the condition. MRI is helpful in distinguishing between endometriosis and sinister pathology such metastatic deposits, Sister Mary Joseph nodule, and granuloma. A homogenous enhancement in T1-weighted lesion with low signals on T2 weighting is one of the MRI characteristics of umbilical endometriosis.[11] Our case had an MRI scan with features of umbilical endometriosis. Histopathologic analysis of the excised specimen is the confirmatory diagnostic technique. The diagnosis of endometriosis is confirmed by the presence of endometrial glands and/or stroma, together with or without hemosiderin-laden macrophages.[13] To aid in confirming the diagnosis of endometriosis, immunohistochemical staining for ER+, PR+, CD10+, CK7+, and CK20- may be done.[14]

Primary umbilical endometriosis pathophysiology may be distinct from that of other kinds of abdominal wall endometriosis since those other types of endometriosis typically develop in relation to surgical history, notably caesarean scars.[10] Primary Umbilical endometriosis may occur as a result of the lymphatic or Hematogenous dissemination of endometrial cells. Another possibility is the direct extension of endometrial cells through the round ligament or the remains of the omphalomesenteric duct.[11] Umbilical endometriosis can be managed by variety of diagnostic and therapeutic strategies. Wide local excision is a curative modality of treatment.[15] The prognosis of primary umbilical endometriosis is good with low recurrence rate.[4]

Several studies and case reports regarding endometriosis in the setting of Nepal were found while searching the literature.[6,16,17] Regarding primary umbilical endometriosis, there was none. In the context of Nepal, our case is therefore unusual. In conclusion umbilical endometriosis should be suspected in the clinical settings of umbilical bleeding especially in the premenopausal women even in the absence of surgical history.

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