

PRIMARY UMBILICAL ENDOMETRIOSIS: A CASE REPORT

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Abstract

Endometriosis is a common benign gynaecological disorder affecting 7-10% of women in the reproductive age group. Umbilical endometriosis represents 0.5-1% of all extragenital endometriosis and it is a rare presentation. A 33-year-old P₀₊₁ woman came at gynaec OPD with the complaints of pain and swelling of umbilicus which increased during menstruation since last two years. The nodule in the umbilicus bled during each menstrual period. Ultrasonography whole abdomen with special reference to periumbilical region revealed a hypoechoic lesion (9mm/9mm) communicating intraperitoneally. FNAC of the lesion showed cells in clusters and sheets with background of scant stromal fragments, hemosiderin laden macrophages and RBCs suggestive of umbilical endometriosis. Surgical excision of the lesion along with reconstruction of the umbilicus was done. Imaging modalities like USG, CT, MRI are nonspecific but are useful in determining the extent of the disease and planning of operative dissection specially, in large and recurrent lesions. FNAC is a fast and accurate method to make the diagnosis before surgery. Both FNAC and histopathological study with immunohistochemistry of CD10 are well known cornerstones in confirmation of diagnosis of umbilical endometriosis. Differential diagnosis of umbilical nodule should include pyogenic granuloma, hernia, residual embryonic tissue, primary or metastatic adenocarcinoma, nodular melanoma and cutaneous endosalpingosis. Surgical excision of the lesion should be done with wide and healthy margin to make accurate histopathological diagnosis and exclusion of malignancy. Hormonal therapy can be tried only for recurrent lesion, though recurrence is unusual.

Keywords: Endometriosis; Primary umbilical endometriosis; FNAC

Introduction

Endometriosis, a term used by Sampson, is the presence of endometrial glands and stroma outside the uterine cavity and musculature.¹ It is a common benign gynaecological disease affecting 7-10% of women in the reproductive age group.² Pelvis is the most common site of the disease. Umbilical endometriosis represents 0.5-1% of all extragenital endometriosis. Extragenital endometriosis usually occurs secondary to gynaecologically induced abdominal or pelvic scar including hysterectomy, episiotomy, caesarean section and laparoscopy.³ Deposits of endometriosis occurs less commonly (12%) in extrapelvic sites like intestinal tract, urinary tract, peritoneum, omentum, lung, thoracic cage, and umbilicus. Extrapelvic endometriosis is very difficult to diagnose due to extreme variability in presentation.

Case presentation

A 33-year-old P₀₊₁ woman came at gynaecology OPD with the complaint of pain and swelling of umbilicus (*Figure 1*) which increases during menstruation since last 2 years. The umbilical nodule also bleed during each menstrual period. The patient was married for 8 years and was treated previously for secondary infertility. On examination the umbilical nodule appeared to be endometriotic nodule. The patient was asked to review at Gynae OPD during her next menstrual period. The

patient was examined again during her menstruation and the nodule seemed to be more tender, presenting with signs of recent bleeding suggestive of a case of umbilical endometriosis without any history of prior pelvic or abdominal surgery (primary umbilical endometriosis). The patient was advised to do USG abdomen with special reference of the periumbilical region to rule out any soft tissue growth and she was referred to pathology department for FNAC (Fine Needle Aspiration Cytology). USG showed a hypoechoic lesion measuring 9mm x 9mm at the submucous plane in the umbilical region which was communicating intraperitoneally. FNAC of the lesions showed cells in clusters and sheets with background of scant stromal fragments, haemosiderin laden macrophages and RBCs, suggestive of umbilical endometriosis. Laparoscopy was done to exclude pelvic endometriosis. The patient was prepared for excision of umbilical endometriotic tissue under spinal anaesthesia. A 4.5cm infraumbilical transverse incision was made; dissection of the endometriotic tissue was done from the surrounding tissue along with tract upto the level of peritoneum (*Figure 2*). Endometriotic tissue was excised and peritoneum was opened, no endometriotic tissue was found within the peritoneal cavity. Haemostasis was checked and secured, abdominal wall was repaired in layers with reconstruction of umbilicus using non-absorbable suture. The specimen was

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sent for histopathological study. Her post operative period was uneventful. She was discharged from the hospital on 6th post-operative day after stitch removal. Her histopathology study (Figure 3) revealed it to be a case of umbilical endometriosis. During her follow up period there is no pain, swelling or bleeding from umbilicus during her menstrual period.

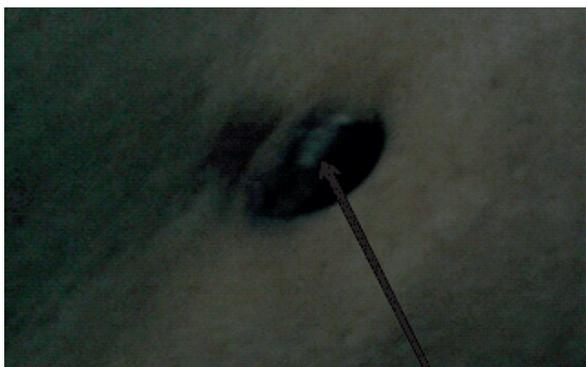


Figure 1. Umbilical Nodule



Figure 2. Subcutaneous part of umbilical nodule (during dissection)

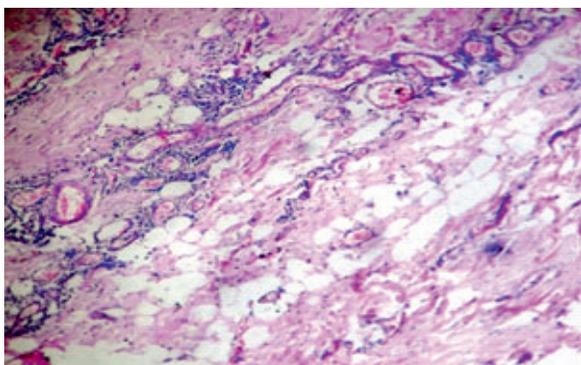


Figure 3. Clusters and sheets of cells with background of scant stromal fragments, haemosiderin laden macrophages and RBCs (400X).

Discussion

An umbilical endometriotic lesion without surgical history is a rare condition.^{4,5} Some case reports about umbilical endometriosis during pregnancy were described in literature.⁶ More than 100 cases of umbilical endometriosis have been described.⁴ It is very difficult to clarify origin of the primary umbilical endometriosis- different hypotheses have been proposed. Sampson hypothesized that endometriosis results from retrograde menstruation through the fallopian tube into the pelvis, however several other theories exist regarding the development of endometriosis including coelomic metaplasia, direct spread, iatrogenic dissemination and lymphatic or haematogenous spread. The disease might arise through metaplasia of urachus remnants in case of isolated umbilical endometriosis.⁷ The real mechanism still remains a mystery. Patients are usually in the reproductive age group and present commonly with swelling, pain, cyclical bleeding and discharge from the umbilicus. Umbilical endometriosis may range in size from 0.5-3.0 cm but can enlarge to even enormous sizes.⁸ While diagnosis is primarily clinical; USG, CT, MRI and laparoscopy are useful diagnostic modalities for endometriosis. MRI has an advantage over laparoscopy for evaluating pelvic and extraperitoneal disease and lesions concealed by adhesion.⁹ The imaging modalities are non-specific but are useful in determining the extent of the disease and planning of operative dissection, especially in large and recurrent lesions.^{3,9} As approximately 15% of the umbilical endometriosis is associated with co-existent pelvic endometriosis, subsequent gynaecological evaluation by laparoscopy for pelvic endometriosis is recommended.¹⁰

FNAC and histopathological study with immunohistochemistry of CD10 are well known cornerstones for confirmation of umbilical endometriosis. Actually FNAC is a fast and accurate method to make the diagnosis before surgery, avoiding errors in approach of the umbilical nodules. A fairly recent study suggested that, "red atolls (small red globular structure) are distinct features of endometriosis."¹¹

Differential diagnosis of the umbilical nodule should include pyogenic granuloma, hernia, residual embryonic tissue, primary or metastatic adenocarcinoma, nodular melanoma and cutaneous endosalpingosis.

Surgical excision of the lesion should be done with wide and healthy margin to make accurate histopathological diagnosis¹² and exclusion of malignancy.^{13,14} Sparing of the umbilicus/reconstruction of the umbilicus is recommended along with the surgical excision of the lesion. In severe cases or in presence of pelvic endometriosis, hormonal therapy with Danazol/GnRH analogs can be given to the patients.¹⁵ Although the local recurrence is uncommon, the patient should be warned of the risks of scar endometriosis and of recurrence. For prevention of the recurrence, post-operative treatment

with Dienogest and GnRH analogs is recommended.¹⁰

Conclusion

Although primary umbilical endometriosis is extremely rare, umbilical endometriosis should be suspected in every case of umbilical nodule even in the absence of pelvic endometriosis or previous abdominal surgery. Surgery is the main treatment. Hormonal therapy can be tried only for recurrent lesion though recurrence is unusual. Patient should be counseled before surgery about the risk of local recurrence. Clinical diagnosis of the umbilical endometriosis is very difficult as it is not associated with classical symptoms always.

Editor's comments:

Primary umbilical endometriosis is a rare under recognized disorder and is defined as the presence of ectopic endometrial tissue within the umbilicus. This disease must be considered in the differential diagnosis upon examining any umbilical lesions. Complete surgical excision with successive histology is recommended for obtaining a definitive diagnosis and optimal treatment.

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