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CASE REPORT

Primary Umbilical Endometriosis: A Rare Entity

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Abstract
Umbilicus is a rare site of endometrioma. Spontaneous or primary umbilical endometrioma are still rarer. A 41years old primipara presented with 1.5 years history of nodular umbilical swelling with cyclical pain and increase in size occurring more pronounced during menses with no prior history of any abdominal surgery or procedure or abdominal trauma. Clinical history gave a likely possibility of umbilical endometrioma. Surgical excision was done as she had not responded to prior hormonal treatment and histopathology confirmed the diagnosis.

Keywords: Endometriosis, Umbilical endometrioma, infertility

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Introduction

Endometrioses are functional endometrial glands and stroma outside the uterine cavity. It commonly occurs in pelvic organ presenting as secondary dysmenorrhoea, menorrhagia, pelvic pain and infertility [1]. Presence at site other than pelvic organ in abdomen or genital usually results because of any obstetrical or gynaecological procedure. However spontaneous primary endometriosis superficial to peritoneum are rare. Cutaneous or umbilical endometriomas are rare with estimated incidence of 0.5-1% but those without any prior history of laparoscopic or other surgical procedure are even rarer [1]. Primary cutaneous lesion is <30% of all cutaneous endometriosis. Umbilicus is most common site of primary cutaneous endometriosis accounting for 0.4-4% of all endometriosis [2]. Cutaneous endometriosis usually occurs on abdominal or pelvic scar of gynaecological surgeries such as hysterectomy, episiotomy, caesarean section or laparoscopy [3]. Umbilical endometriosis (UE) was first described by Villar hence also known as Villar's Nodule [2].

Case Presentation

A 41-year-old female para 1 living 1 abortion 1 (P_1 L_1 A_1) presented with history of swelling at umbilical region for 1 and half years associated with pain during menses. There was no history of associated discharge or bleeding from umbilicus during menses, no history of increase in size during coughing, rather the swelling increased in size during menses. She denied any other gynaecological complain. There was no preceding history of abdominal trauma or any abdominal surgical intervention, no history of contraceptive use. She had an ultrasound showing blood collection at umbilical region. She had taken symptomatic medical treatment for above symptoms but had no relief. On examination an indurated raised area 5*5 cm was present around umbilicus. In view of her clinical presentation, examination and sonographic findings a clinical diagnosis of primary umbilical endometrioma was made and planned for surgical excision as prior medical treatment had failed. Excision of umbilicus enbloc along with fat tissue and fibrous nodular swelling was done until normal tissue visible at margin and sent for histopathological examination followed by closure of umbilical defect. There was no communication with peritoneal cavity. On histopathology gross finding suggested cystic spaces filled with blood. On microscopy sub epithelial tissue showed cystically dilated endometrial glands with inflammatory infiltrate in glandular endometrial lumen along with haemorrhage with hemosiderin laden microphages in surrounding stroma, these finding were consistent with umbilical endometriosis (Figures 1 and 2).

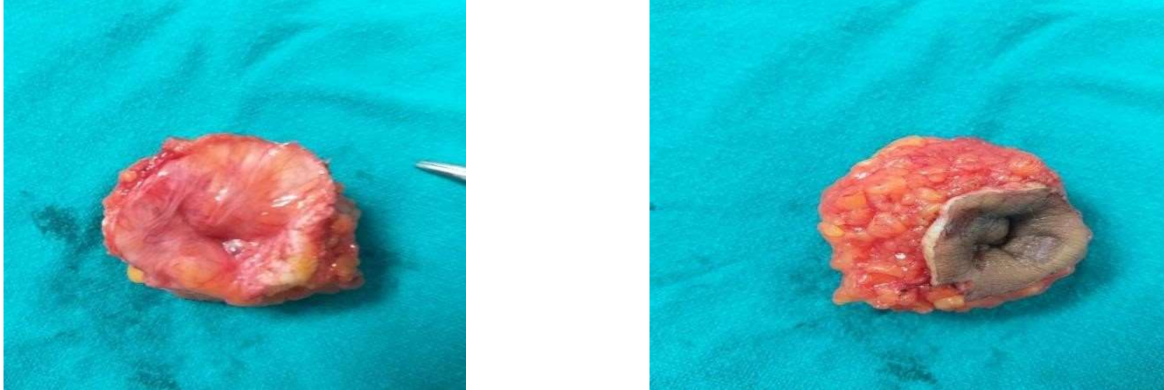

Figure 1. Excised Specimen

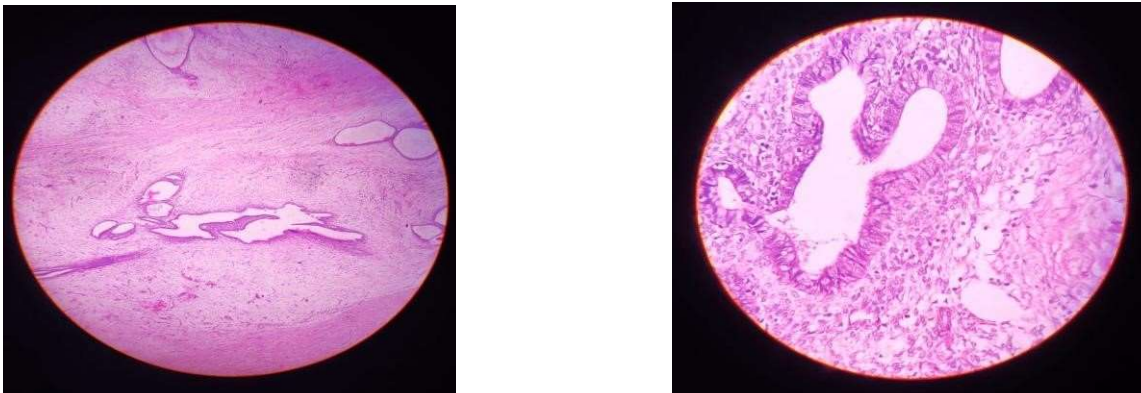

Figure 2. Histopathology images showing endometrial like glands and endometrial stroma

Discussion

Pelvic endometriosis commonly occurs in ovaries, uterosacral ligament, cul-de-sac, bladder as postulated by Sampson's retrograde theory of menstruation. Extra pelvic site like genital organs, skin, lungs or brain may occur due to lymphatic/ haematogenous dissemination postulated by Halban. Ectopic endometrium behaves in same way as when present in utero. Hormone level causes thickening, breakdown and bleeding leading to swelling, pain and or bleeding. Umbilical endometriosis may present similarly as pain, swelling or nodule, discolouration/ bleeding which aggravates during menstruation.

Pathogenesis of secondary UE is easy to explain as it follows procedure like laparoscopy or open surgery like caesarean section, amniocentesis etc. or may be associate with pelvic endometriosis but for primary UE there is unclear pathogenic mechanism. Hypothesis proposed are embryonal rest theory of Wolfian or Mullerian remnant as described by Latcher classification of all theories for endometriosis, the transplantation theory in which ectopic endometrial tissue harbours from retrograde menstruation or haematogenous lymphatic dissemination, or both [4,5]. Other hypothesis include the coelomic metaplasia or induction theory,

suggest that sloughed endometrium produces substance that form endometriosis induced by hormonal manipulation , inflammation or trauma or a recent theory of cellular immunity proposed cellular proliferation of ectopic endometrial cells wherein umbilicus acts as physiological scar with predilection for endometrial tissue [6].

Diagnosis of UE can be made by dermoscopy wherein cytologic smear reveals high cellularity with haemosiderin laden macrophages and sheets of stromal and epithelial cells on haemorrhagic background [1]. USG/ MRI usually helps to rule out another differential. FNAC is not conclusive. Histopathology where two of three findings i.e endometrial like glands, endometrial stroma or hemosiderin pigment confirms diagnosis. IHC confirm diagnosis by showing positivity for both oestrogen and progesterone receptors and for antigen CD 10, a marker for stromal cells in endometriosis [7].

Swelling at umbilical region must be differentiated from other possible differentials which either presents as swelling like lipoma, sarcoma, primary/ metastatic cancer, cyst, hernia or differentials which may presents as skin discolouration like pyogenic granuloma, nodular melanoma, primary/metastatic adenocarcinoma, residual embryonic tissue and cutaneous endosalpingiosis [1,3].

Management of UE is also not clear. Hormonal treatment with progesterone, danazol, GnRH analogous are usually not successful though provide a temporary relief in symptom and size providing a better assess to surgical treatment or they may be less responsive. Hence surgical treatment with either complete umbilical resection with/ without repair of underlying fascia or local resection of nodule are preferred approach[2]. It also helps in appropriate histopathology and exclude unusual cutaneous malignancies. Surgery should ideally be done with at least 1 cm normal margin to prevent local recurrence. Surgery usually done in post menstrual phase to avoid extensive excision.

Conclusion

UE should be considered in any women presenting with clinical pain, swelling, discharge or bleeding or discoloration of umbilicus even in absence of any prior abdominal surgery intervention. Total resection of umbilicus is preferred treatment and also avoid local recurrences.

Conflict of Interest -Nil

Funding received- Nil

Ethical Considerations: Addressed by the authors.

REFERENCES:

1. Efremidou E I, Kouklakis G, Mitrakas A, Liratzopoulos N, Polychronidis A Ch. Primary umbilical endometrioma: a rare case of spontaneous abdominal wall endometriosis Int J of General Medicine. 2012; 5:999-1002
2. Fancellu A, Pinna A, Manca A, Capbianco G, Porcu A. Primary umbilical endometriosis. Case report and discussion on management option.

Int J of Surgery Case Report. 2013; 4:1145-1148

3. Egin S, Pektas B A, Hot S, Mihnanli V. Primary umbilical endometriosis: A painful swelling in the umbilicus concomitantly with menstruation. Int J of Surgery Case Report. 2016; 8:78-80
4. Pallavi VB, Giurguis M M. Menstruating from the umbilicus as a rare case of primary umbilical endometriosis: a case report. J Med Case Rep. 2009; 3:9326
5. Kyamidis K, Lora V, Kanitakis J. Spontaneous cutaneous umbilical endometriosis; report of new case with immunohistochemical study and literature review. Dermatol Online J. 2011; 17:5
6. Chatzikokkinou P, Thorfinn J, Angelidis I K, Papa G, Trevisan G. Spontaneous endometriosis in an umbilical skin lesion. Acta Dermatovenerol Alp Panonica Adriat. 2009; 18:126-130
7. Capobianco G, Wenger J M, Marras V, Cosmi E, Ambrosini G, Dessole M, et al. Immunohistochemical evaluation of epithelial antigen BER-EP4 and CD 10: new marker for endometriosis. Eur J Gynaecol Oncol.2013;3:254-256