VOLUME - 12, ISSUE - 01, JANUARY - 2023 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper | Obstetrics & Gynaecology

PRIMARY UMBILICAL ENDOMETRIOSIS (VILLARS NODULE): A RARE PRESENTATION

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ABSTRACT Umbilical endometriosis aka Villars nodule is the commonest type of cutaneous endometriosis.1 It is usually seen in reproductive age group. The patients with this condition present with cyclical bleeding from umbilicus and pain/swelling over the umbilicus. The diagnosis of umbilical endometriosis is made only on histopathological examination. A 44 years old female presented in OPD with complain of pain and swelling at umbilicus for 2 months with episodes of bleeding from umbilical area during menstrual cycles. A detailed history, general physical examination, systemic examination and local examination was performed. Routine investigations, ultrasound of whole abdomen and pelvis and FNAC from swelling was advised. Histopathological examination of the tissue received on FNAC revealed glandular component in the cells suggestive of endometriosis. Then patient was diagnosed with umbilical endometriosis and was put on medical management for the same.

KEYWORDS :

INTRODUCTION

Endometriosis is the presence of endometrial tissue outside the uterine cavity. It is a benign condition affecting 10-15% of women, classically affecting reproductive age group.[2] The lesion occur mostly in the pelvis sites involving the cul-de-sac, ovaries, uterosacral ligaments, ovarian fossa and bladder.[3] Extrapelvic endometriosis occur less commonly and includes diaphragm, pulmonary, urinary tract and cutaneous endometriosis. Umbilical endometriosis aka Villars nodule is the commonest type of endometriosis.[1] Umbilical endometriosis can be categorized as Primary when it occurs spontaneously or secondary when it occurs following laparoscopic or open surgical procedures, the latter being more common.[4]

Primary umbilical endometriosis is a rare condition seen in 0.5-1% of patients of endometriosis. The condition is called Primary cutaneous endometriosis when there is no prior history of endometriosis or surgeries. Primary umbilical endometriosis was first described by Villars in 1886 and hence it is called Villars nodule.[5] .

The pathogenesis is not well understood. The most common postulated theories are Sampson's theory of retrograde menstruation followed by coelomic metaplasia, induction theory, stem cell theory, hematogenous/lymphatic spread and embryonic theory. The theory favoured in the case of umbilical endometriosis is hematogenous/lymphatic spread where there is coexisting pelvic endometriosis. Isolated umbilical endometriosis could develop from metaplasia of urachal remnants.[6]

Due to its rarity, there are no clear guidelines on its treatment modalities. Main options in the management are medical, surgical or conservative after ruling out umbilical endometriosis.

We hereby report an interesting case of 44 years old female with umbilical endometriosis.

Case Report

A 44 years old P_2L_2presented to OPD with complain of cyclical pain over the umbilicus along with gradual onset of swelling from umbilicus for past 2 months associated with scanty bleeding from umbilicus which coincided with her menstrual cycle. The bleeding and pain would start few days before her menstrual cycle and seized by day 2 of her menstrual cycle. Her menstrual cycles were regular with normal flow and no dysmenorrhea. She also noted blackish discolouration of umbilicus along with swelling over it. There was no history of any previous umbilical/pelvic surgeries, or any type of hormonal treatment.

On examination, there was an umbilical nodule of around 3*3 cm with dusky blue discolouration (Figure1) with no visible bleeding/discharge present during examination and no other palpable abdominal mass was found. Per speculum and per vaginal examination was done and was found to be unremarkable. Ultrasound of abdomen and pelvis were advised which was found to be normal with no signs of pelvic endometriosis. FNAC from the umbilical swelling was advised. Histopathological examination showed endometrial glands and stroma confirming the diagnosis (Figure 2 & 3).

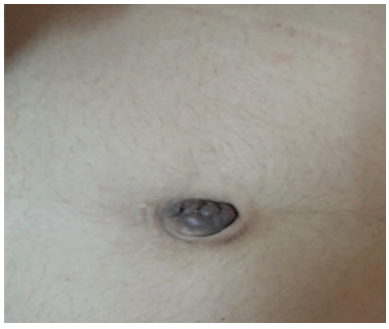

Figure 1: clinical appearance of umbilical nodule with blackish discolouration.

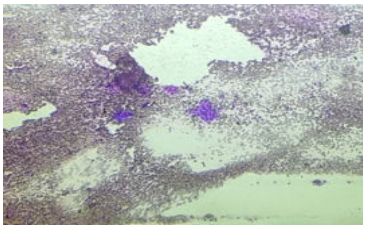

Figure 2: Micrograph of the histological examination of the tissue showing endometrial glands and stroma on low power 10x

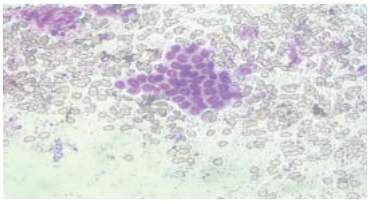

Figure 3: Micrograph of the histological examination of the

tissue showing endometrial glands and stroma on low power 40x

Following histopathological report, patient was given options for conservative or medical/surgical management and the patient opted for medical management. Patient was put on Injection Depot medroxy progesterone acetate (DMPA) (150mg single dose intramuscularly followed by 2nd dose after 3 months and then patient was advised follow up. Patient had a follow up 3 months after 2nd dose, the swelling over umbilicus was reduced along with relief from symptoms.

DISCUSSION

Endometriosis is the presence of endometrial tissue outside the uterine cavity. It is a benign condition affecting 10-15% of women, classically affecting reproductive age group.[2] The lesion occur mostly in the pelvis sites involving the cul-de-sac, ovaries, uterosacral ligaments, ovarian fossa and bladder.[3] Extrapelvic endometriosis occur less commonly and includes diaphragm, pulmonary, urinary tract and cutaneous endometriosis. Umbilical endometriosis aka Villars nodule is the commonest type of endometriosis.[1] Umbilical endometriosis can be categorized as Primary when it occurs spontaneously or secondary when it occurs following laparoscopic or open surgical procedures, the latter being more common.[4]

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As the presentation is very rare, there is no definite treatment plan available for this condition. Medical management with hormonal therapy, using estrogen, progesterone and danazol has shown recurrence after stoppage of treatment which eventually needed surgical management. Hence, the treatment of choice is surgical, with en bloc excision of nodule with adequate margins to prevent recurrence. However, if patient does not consent to complete removal of umbilicus, local excision of lesion preserving the umbilicus may be done. The absence of umbilicus may be a cause of psychological distress, hence, the best result may be obtained by ensuring a wide excision and recreation of new umbilicus to give youthful appearance to abdomen. There are multiple methods of neoumbilicoplasty like the purse string method, the double VY procedure and the cone flap.[8,9,10,11]

CONCLUSION

Umbilical endometriosis is a rare entity, especially, primary endometriosis. It is both a diagnostic and treatment dilemma with clinical presentation ranging from umbilical swelling, cyclical pain and sometimes bleeding from lesion highly suggesting the diagnosis. Once the diagnosis is confirmed after histopathological examination medical and surgical treatment can be offered but surgical excision remains the treatment of choice.

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