

Primary umbilical endometriosis: cytological diagnosis of a rare presentationGajender Singh¹, Komal Brar^{2*}, Padam Parmar³, Gurupriya J⁴, Sant Prakash Kataria⁵, Neha Yadav⁶, Rajeev Sen⁷¹Professor, ²Junior Resident, ³Senior resident, ⁴Junior Resident, ⁵Professor, ⁶Junior Resident, ⁷Senior Professor & Head
Department of Pathology, PGIMS, Rohtak, Haryana***Corresponding author**

Dr. Komal Brar

Email: komalbrar99@gmail.com

Abstract: Primary umbilical endometriosis is a rare entity with an incidence of 0.5-1% of all patients with endometriosis. Pelvis is the most common site of the disease. Extrapelvic site is not common and difficult to diagnose due to variable presentation. A 36-year-old female presented with a umbilical swelling for 2 weeks. Fine needle aspiration cytology of the lesion showed groups and clusters of cells along with few groups of stromal cells in the background of occasional hemosiderin laden macrophages admixed with blood, leading to a diagnosis of umbilical endometriosis, which was confirmed on histopathology.**Keywords:** Fine needle aspiration, cytology, umbilicus, endometriosis, primary.

INTRODUCTION

Endometriosis term is used to describe the presence of endometrial glands and stroma outside the endometrial cavity. The most common site of endometriosis is the pelvic cavity [1]. Endometriosis is rare in the extra pelvic sites and umbilical endometriosis is responsible for only 0.5 to 1 % of all cases of extra pelvic endometriosis [2]. It can be a consequence of abdominal or scars induced in surgeries such as hysterectomy, caesarean section, laparoscopy etc [3]. Few cases of primary spontaneous endometriosis of the umbilicus have also been reported [4-6].

CASE REPORT

A 36-year old female presented with umbilical swelling for a period of 2 weeks. There was no history of bleeding from the site of complaint. On examination there was a tender, soft ill-defined swelling measuring 2.5 X 1 cm in the umbilical region. Ultrasonography of the local site showed a cystic lesion measuring 5 X 5 mm with an echogenic nidus seen at the site of complaint. A fine needle aspiration was performed which yielded a blood mixed aspirate. The smears prepared were stained with May-Grunwald-Giemsa (MGG) and showed endometrial epithelial cells in groups, clusters, sheets and dispersed singly having round to oval nuclei, inconspicuous nucleoli and scant to moderate amount of cytoplasm in some along with few groups of stromal cells (Figure 1a, 200x; 1b, 200x; 1c, 100x), in the background of occasional macrophages admixed with blood (Figure 1d, 100x). Cytological features were suggestive of endometriosis. The patient subsequently underwent wide excision of

the umbilical swelling, diagnosis was confirmed on histopathology.

DISCUSSION

Spontaneous or primary umbilical endometriosis was first defined by Villar in 1886; therefore it is also called Villars nodule [3]. The pathogenesis of endometriosis is not understood very well, there are several theories proposed as reason of development of endometriosis. They include coelomic metaplasia, lymphatic, haematogenous or direct spread etc. [4]. Latcher classified these theories into three main categories, the embryonic rest theory, the coelomic metaplasia theory and the implantation theory. In cases of spontaneous endometriosis at distant site implantation by haematogenous or lymphatic spread may be the cause [7].

Extra pelvic endometriosis presents in a variety of ways which makes its diagnosis a difficult task. Clinical diagnosis is usually difficult in cases of umbilical endometriosis. Patients may present with a mass in the umbilical region associated with pain and cyclical bleeding [1]. The diagnosis is usually performed clinically, but diagnostic procedures such as ultrasonography, computed tomography can be used often [7].

The cytology smears are cellular and comprise of epithelial and stromal fragments admixed with haemorrhage and hemosiderin laden macrophages. The cytologic features of endometriosis are related to cyclic hormonal changes. Proliferative phase is characterized by epithelial cells forming cohesive sheets of uniform, small cells with scant cytoplasm, round to oval nuclei

with bland chromatin. In secretory phase, the cells show microvacuolation. The background is sanguinous and contains a mixture of lymphocytes, neutrophils and histiocytes [3, 8]. The risk of malignant transformation from umbilical endometriosis is very low. Only two

cases of umbilical endometriosis with malignant transformation have been reported [9]. The preferred treatment is wide local excision of the mass; several physicians also prefer the use of additional hormonal therapy [10].

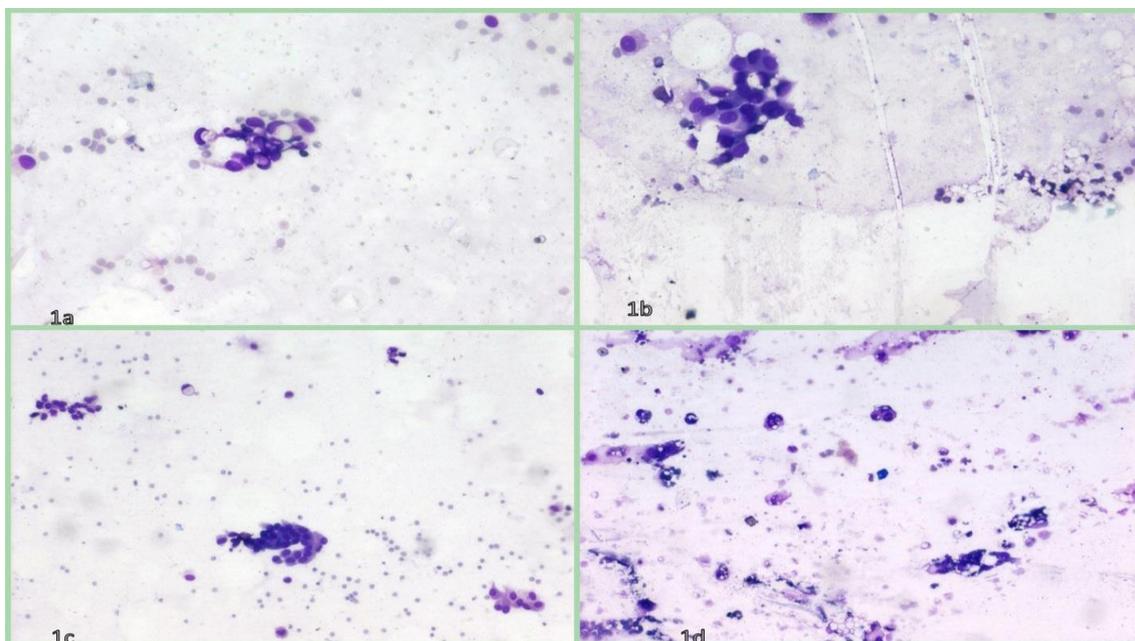


Fig 1a & b: Microphotograph showing groups and sheets of endometrial epithelial cells and few stromal cells in the background (MGG, 200x).

Fig 1c: Microphotograph showing groups and sheets of endometrial cells with crowded nuclei (MGG, 100x)

Fig 1 d: Microphotograph showing few scattered endometrial cells and macrophages admixed with blood (arrow) (MGG, x100)

CONCLUSION

FNAC is an accurate method to diagnose endometriosis before surgery, and eases the approach of the umbilical nodules.

REFERENCES

1. Bagade PV, Guirguis MM. Menstruating from the umbilicus as a rare case of primary umbilical endometriosis: a case report. *Journal of medical case reports*. 2009 Dec 10; 3(1):9326.
2. Papavramidis TH, Sapalidis K, Michalopoulos N, Karayanopoulou G, Raptou G, Tzioufa V, Kesisoglou I, Papavramidis ST. Spontaneous abdominal wall endometriosis: a case report. *Acta Chirurgica Belgica*. 2009 Jan 1; 109(6):778-81.
3. Fernandes H, Marla NJ, Pailoor K, Kini R. Primary umbilical endometriosis-Diagnosis by fine needle aspiration. *Journal of cytology*. 2011 Oct 1; 28(4):214.
4. Catalina-Fernandez I, Lopez-Presa D, Saenz-Santamaria J. Fine needle aspiration cytology in cutaneous and subcutaneous endometriosis. *Acta cytologica*. 2007 Jul 1; 51(3):380-4.
5. Khaled A, Hammami H, Fazaa B, Zermani R, Ben Jilani S, Kamoun MR. Primary umbilical endometriosis: a rare variant of extragenital endometriosis. *Pathologica*. 2008 Dec; 100(6):473-5.
6. Teh WT, Vollenhoven B, Harris PI. Umbilical endometriosis, a pathology that a gynecologist may encounter when inserting the Veres needle. *Fertility and sterility*. 2006 Dec 31; 86(6):1764-e1.
7. Efremidou EI, Kouklakis G, Mitrakas A, Liratzopoulos N, Polychronidis AC. Primary umbilical endometrioma: a rare case of spontaneous abdominal wall endometriosis. *Int J Gen Med*. 2012; 5:999-1002.
8. Medeiros FD, Cavalcante DI, Medeiros MA, Eleutério J. Fine-needle aspiration cytology of scar endometriosis: Study of seven cases and literature review. *Diagnostic cytopathology*. 2011 Jan 1; 39(1):18-21.
9. Lauslahti K. Malignant external endometriosis. A case of adenocarcinoma of umbilical endometriosis. *Acta pathologica et microbiologica Scandinavica. Supplement*. 1971 Dec; 233:98-102.
10. Bektaş H, Bilsel Y, Sarı YS, Ersöz F, Koç O, Deniz M, Boran B, Huq GE. Abdominal wall endometrioma; a 10-year experience and brief review of the literature. *Journal of Surgical Research*. 2010 Nov 30; 164(1):e77-81.