



**Case Report:**

**Villar's Nodule**

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**Abstract:**

Villar's Nodule or primary umbilical endometrioma is a rare condition, with an estimated incidence of 0.5 to 1% of all patients with endometrial ectopia. Several aetiological theories have been proposed for its pathogenesis with coelomic metaplasia being the favoured one and also, umbilicus may acts as a physiological scar with a predilection for endometrial tissue in the development of spontaneous umbilical endometriosis. Since endometriosis remains a diagnostic and therapeutic enigma even today, largely due to its variable presentations, we, hereby report a case of primary umbilical endometrioma due to its rarity and unusual presentation.

**Key Words:** Umbilical; Endometriosis

**Introduction:**

Primary umbilical endometrioma, also known as Villar's Nodule is a rare condition, with an estimated incidence of 0.5 to 1% of all patients with endometrial ectopia.(1) It was first discovered by Villar in 1886, hence the name (2) and is defined as presence of ectopic endometrial glands within umbilicus without prior history of pelvic endometriosis.(3)

Endometriosis remains a diagnostic and therapeutic enigma even today, largely due to its variable presentations and in spite of its classical presentation, misdiagnosis is not uncommon. We, hereby report a case of primary umbilical endometrioma due to its rarity and unusual presentation.

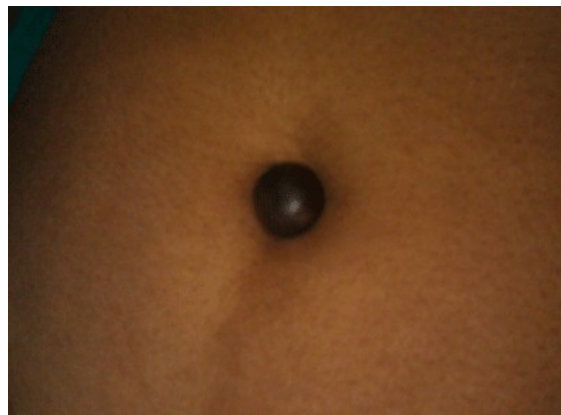
**Case Report:**

A 29 years old unmarried, nulliparous female presented in the outpatient department of Gynaecology, R G Kar Medical College & Hospital, Kolkata, with chief complaint of a painful nodule in the umbilical region since one year. The pain was dull in nature but tend to become severe during menstruation. Her menstrual cycles were regular with no history of dysmenorrhoea.

On local examination, a hyperpigmented, dark brown umbilical nodule was seen measuring 1.5 cm in its greatest dimensions. The nodule was firm in consistency and tender.(Figure 1) It was non-

reducible, non-pulsatile and with no impulse on coughing. The systemic examination was unremarkable.

Patient's routine haematological and biochemical profile was within normal limits apart from mild anaemia. Ultrasonography of whole abdomen was done which revealed a lesion of variable echogenicity, measuring 32x18mm, with mild adhesion to the underlying parites and no relation to the abdominal viscera.(Figure 2) The uterus along with its adnexa was normal. Excision biopsy of the umbilical nodule was done and the specimen was sent to the department of pathology for histopathological examination. During excision biopsy, the umbilical nodule was found to be densely adhered to the underlying rectus muscle.



**Figure 1: Hyperpigmented, dark brown umbilical nodule measuring 3x2 cm**

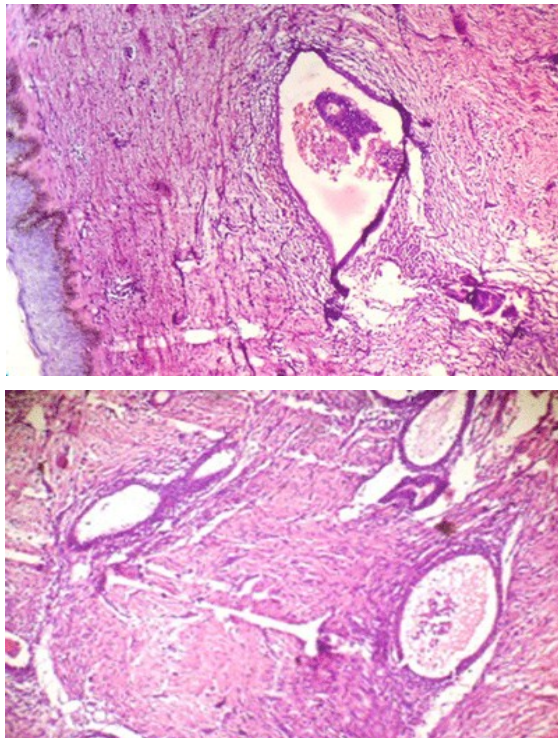


**Figure 2: Umbilical lesion of variable echogenicity, measuring 32x18mm**

#### **Pathological Examination:**

On gross, specimen was partly skin covered, nodular and measured 3.5x2.0 cm. Cut surface was brownish with areas of haemorrhages. Microscopic examination showed numerous endometrial glands surrounded by compact endometrial stroma embedded in the subepithelial connective tissue stroma. (figure 3 & 4) The overlying stratified squamous epithelium was unremarkable. A few haemosedrin containing macrophages were seen at places and a histopathological diagnosis of umbilical endometriosis was made.

After the diagnosis of endometriosis was established, no recurrence has been noted at the site till now and patient has no other complaints.



**Figure 3 & 4: Numerous endometrial glands surrounded by compact endometrial stroma embedded in the subepithelial connective tissue stroma (H&E, 400x)**

#### **Discussion:**

Umbilical endometriosis is rare, especially in patients who do not give a history of previous pelvic surgery or who do not have clinical evidence of preexisting pelvic endometriosis. Mean age of presentation of patient with umbilical endometriosis is 37.7 +/- 0.98 years with swelling (90.9%), pain (81.5%), and bleeding (49.2%) as the common mode of presentation. Most umbilical lesions are brown in color, followed by blue, purple, black and red. (4) This case also presented with brownish black swelling and prominent cyclical pain.

The pathogenesis of spontaneous cutaneous endometriosis is yet unknown. Several aetiological theories have been proposed. These include coelomic metaplasia, congenital presence of developmentally-displaced endometrial tissue, direct extension through the round ligament or the patent omphalomesenteric duct, or mechanical seeding of endometrial tissues via the lymphatic or venous system transfer via lymphatics or blood vessels.(5)

In the development of spontaneous umbilical endometriosis, as in the case presented, it is possible that the umbilicus acts as a physiological scar with a predilection for endometrial tissue.(6) However, in our opinion, it is still not clear how the endometrial cells has traveled to umbilical location and the other above mentioned theories might have played a role in the development of villar's nodule of present case.

Umbilical endometriosis can pose a diagnostic dilemma as it can simulate a malignant melanoma or the "sister Mary Joseph nodule"—a manifestation of intra-abdominal malignancy.(5) Any other condition that presents with a subcutaneous mass or discoloration of the umbilical skin, such as a benign nevus, a lipoma, an abscess, a cyst, or a hernia, as well as a metastatic deposit from a systemic malignancy should be considered in the differential diagnosis.(6)

Though the most important key feature of diagnosis is intermittent bleeding in accordance with the menstruation cycle,(4) magnetic resonance imaging, epiluminescence microscopy or, fine-needle aspiration cytology may help resolve the diagnostic dilemma.(5) However, histopathology is the main stay of diagnosis. Because of the classical symptoms and ultrasonography picture, and also the poor socioeconomic background of the patient, excision biopsy with histopathological examination was done in the present case to reach at a confirmatory diagnosis rather than undergoing all the above mentioned diagnostic investigations.

The treatment of choice of Villar's Nodule remains surgical excision with sparing of the umbilicus where possible, and recurrences, though reported, are rare.(8) Malignant transformation of umbilical endometriosis is extremely rare with only one case reported in the literature where umbilical endometriosis was present since age of thirty.(9)

To conclude, endometriosis is important to consider in cases of unclear skin lesions of the umbilicus, even in cases with no previous abdominal surgery. And also, excisional surgery is the treatment of choice so as to avoid lesion relapse and the risk of oncogenic transformation.

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