Villar's Nodule: A Rare Presentation of External Endometriosis

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Introduction

Umbilical endometriosis, also called Villar's Nodule, was first described by Villar in 1886. It is extremely rare, the incidence being only 0.5-0.1% of all women with extra gonadal or external endometriosis [1]. This condition should not be mistaken for scar endometriosis occurring in a subumbilical incision scar.

In the late 19th century, the term endometriosis was coined by Sampson to characterize ectopic tissue, possessing histologic architecture and function of the uterine endometrium. Endometriosis involving the subcutaneous tissues of the umbilicus must be suspected in a woman presenting with localised cyclical pain and swelling, in association with menstruation [2].

The most common sites for extra gonadal endometriosis are the gastrointestinal tract, especially the rectum, colon, and the peritoneal surfaces. Ureteral or urinary bladder involvement is also known to occur and could result in cyclical pain and hematuria during menstruation. Pulmonary involvement could manifest as pneumothorax, hemothorax or hemoptysis during menstruation. External endometriosis, involving the subcutaneous tissues, has also been reported in the vicinity of a surgical scar following surgeries such as hysterectomy, hysterotomy, cesarean section, episiotomy and laparoscopy [3].

Case Report

A 44 year old lady presented with complaints of a painful lump in the umbilicus for the last two years. The pain was a dull ache throughout the month but became severe and acute during her periods. Her menstrual cycles were regular and flow was normal. She did give a history of pelvic discomfort during her cycles though there was no specific history of significant dysmenorrhoea. There was no history of vicarious

menstruation or cyclical pain anywhere else in the body.

She had two living issues both delivered vaginally and her last childbirth was fifteen years ago. She did not give any history of subfertility or any treatment for infertility in the past. There was no past history of pulmonary or extra pulmonary tuberculosis. She did not give history of a hysterotomy or any other pelvic surgery, including laparoscopy, in the past.

General physical examination was essentially unremarkable. Local examination of the umbilicus revealed that there was a firm tender nodular mass measuring 4 X 3 cm located inferior and posterior to the umbilical ring. The margins were well defined and the surface was irregular. On abdominal tensing the nodule appeared superficial to, but attached to the underlying muscles. Pelvic examination revealed that uterus was normal in size, anteverted, mobile and non tender. There was some nodularity in the fornices though no definite mass could be palpated.

On investigation her biochemical and hematological parameters were normal. Ultrasound of the abdomen and pelvis revealed an echogenic nodular space occupying lesion located in the abdominal parietes just below the umbilicus. The uterus and left ovary appeared normal; however there was a 3 X 3 cm thick walled, partly cystic lesion, with internal echoes in the right adnexa separate from the ovary.

Keeping in mind the umbilical nodule with cyclical pain and the ultrasound findings, the possibility of external endometriosis with pelvic endometriosis was kept in mind and patient was taken up for surgical excision of the nodule. In the same sitting the patient was also subjected to a pelvic endoscopy. First laparoscopy was performed and the incision for passing the 10 mm port, through which the telescope was introduced, was made below the lower margin of the umbilical nodule. This incision was later on extended upwards in order to excise the nodule. The umbilical nodule itself was densely adherent to the rectus muscle and had to be dissected free using sharp dissection and diathermy. Pelvic laparoscopy

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Fig. 1 : Microphotograph of Villar's Nodule showing endometrial tissue composed of endometrial glands and surrounding stroma. Inset – Endometrial tissue under high power.

revealed that the patient had advanced pelvic endometriosis with a chocolate cyst in the right ovary measuring 3 X 4 cms. A laparoscopic chocolate cystectomy with fulguration of the active endometriotic deposits in the pelvis was done in the same sitting. As the patient did not have any symptoms of pelvic endometriosis it was decided not to undertake any radical surgery.

Histopathological examination of the umbilical nodule revealed plenty of endometrial glands surrounded by compact stroma (Fig. 1), confirming the diagnosis of umbilical endometriosis.

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. In a review of literature, Victory et al [4] found only 122 reported cases of umbilical endometriosis worldwide from 1966 till 2007.

The exact aetiopathogenesis of endometriosis is not known. However there are two major theories which can explain the formation of this condition: one is the metastasis theory and the other is the metaplasia theory. The metastatic theory suggests that ectopic implantation of endometriotic tissue to extrauterine tissue occurs by either lymphatic or hematogenous metastasis. In the case being reported upon, the spontaneous umbilical endometriosis could have arisen due to endometrial tissue that is transported via lymphatics or vascular channels. The metaplasia theory states that embryonic coelomic

epithelium under certain stimuli differentiates into endometrial tissue.

The association of endometriosis and sub fertility remains controversial and according to the American Society of Reproductive Medicine, endometriosis, if moderate or severe, is seen to be associated with infertility [5]. Having said that, it is also seen that 44% of asymptomatic women with normal fertility are detected to have moderate to severe endometriosis, during surgery for tubal ligation [6], and this could explain why our patient did not manifest with infertility despite having severe endometriosis.

The treatment of choice of Villar's Nodule remains surgical excision with sparing of the umbilicus where possible, and recurrences, though reported, are rare [7]. Malignant transformation of the umbilical nodule into endometrial carcinoma has also been reported [8].

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Conflicts of Interest

None identified

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