

Extra-Gonadal Endometriosis: Unusual Presentation

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Background: Extra-gonadal Endometriosis is a rare entity. It is rarely reported from this region.

Objective: To present six cases of unusual extra-gonadal endometriosis.

Design: Retrospective review

Setting: Surgical Department, Salmaniya Medical Complex, Kingdom of Bahrain.

Method: Six patients with extra-gonadal endometriosis who presented to the surgical department during a period of three years (between 2002 –2005) were reviewed.

Result: The patients were females with age ranging between 29 and 48 years. Three patients presented with painful umbilical mass and bleeding during menses. One patient had a painful nodule in her lower segment caesarian section scar; another had endometrioma of the appendix and one presented with acute small bowel obstruction.

All had regular menstrual cycle. The majority had associated menorrhagia and dysmenorrhea but did not have any significant past gynecological history.

CT scan in three patients confirmed the presence of umbilical nodule or mass but was not helpful in reaching the diagnosis pre-operatively. In the other three cases, pre-operative investigations were not helpful in reaching the definitive diagnosis.

The management in all the cases was complete resection. Histopathology confirmed the diagnosis.

Conclusion: Six cases of extra-gonadal endometriosis with unusual presentation had been reviewed and presented in this study. Extra-gonadal endometriosis is a rare condition presenting to the general surgeon. Clinical awareness is more important in diagnosis than radiological findings. Complete surgical resection is the rule in all cases.

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Endometriosis is a painful, chronic disease that affects more than five million women and girls in the USA and Canada, and millions more worldwide. It has been well described since early 1900's.

Endometriosis is the abnormal growth or ectopic functional endometrial tissue outside the uterine cavity¹.

This misplaced tissue or ectopic implants develop into growths or lesions which respond to the cyclic hormonal stimulation in the same way the uterine endometrial tissue responds². The response includes intrauterine proliferation, secretory activity, and cyclical sloughing of menstrual material².

It usually occurs in the abdomen: on the ovaries, fallopian tubes, and ligaments that support the uterus; the area between the vagina and rectum; the outer surface of the uterus; and the lining of the pelvic cavity.

The cause is unknown, but theories have suggested different explanations. In umbilical endometriosis the theory of retrograde menstruation could explain its development³. Other leading theories include metaplastic conversion of coelomic epithelium and hematogenous or lymphatic transport of endometrial cells. Other theories such as impaired immune system and genetic factors were found to predispose some women to endometriosis. Surgical transplantation has been blamed in many cases where endometriosis was found in abdominal scars, such as lower segment cesarion section scar.

There is no specific blood test to diagnose endometriosis. Surgical resection is the standard management.

Rare cases have undergone malignant transformation and given rise to endometrial carcinoma⁴.

The aim of this study is to present six cases of extra-gonadal endometriosis and to increase the awareness of surgeons for this disease entity and recognition at an early stage to improve management.

CASE 1

Forty-eight years old female patient, married with three children, presented with history of para-umbilical swelling of twenty years duration. Recently, over the last six months she started to have pain and bleeding per umbilicus during menstruation. The patient has normal menstrual history. There was no past history of any gynecological, medical or surgical treatment. She is known to have hypertension and on regular treatment. She has family history of polycystic disease of the kidney, hypertension and renal failure in her mother and eldest brother.

On examination; the patient was healthy. The blood pressure was 140/80. There was a palpable right kidney. Local examination showed a para-umbilical hernia with fleshy dark red

nodule protruding out. The nodule was 3x4x4 cm³ and showed signs of recent bleeding with clots.

Investigations showed normal hemoglobin (12.5 gm %), normal liver and renal functions. CT scan abdomen and pelvis showed multiple cysts involving liver and both kidneys. The uterus was bulky suggestive of the presence of fibroid, but the ovaries were normal. In the umbilicus there was cystic soft tissue complex swelling (2x2 cms), not communicating with bladder or bowel (fig. I). The provisional diagnosis was adult polycystic disease with involvement of the umbilicus (fig.1).

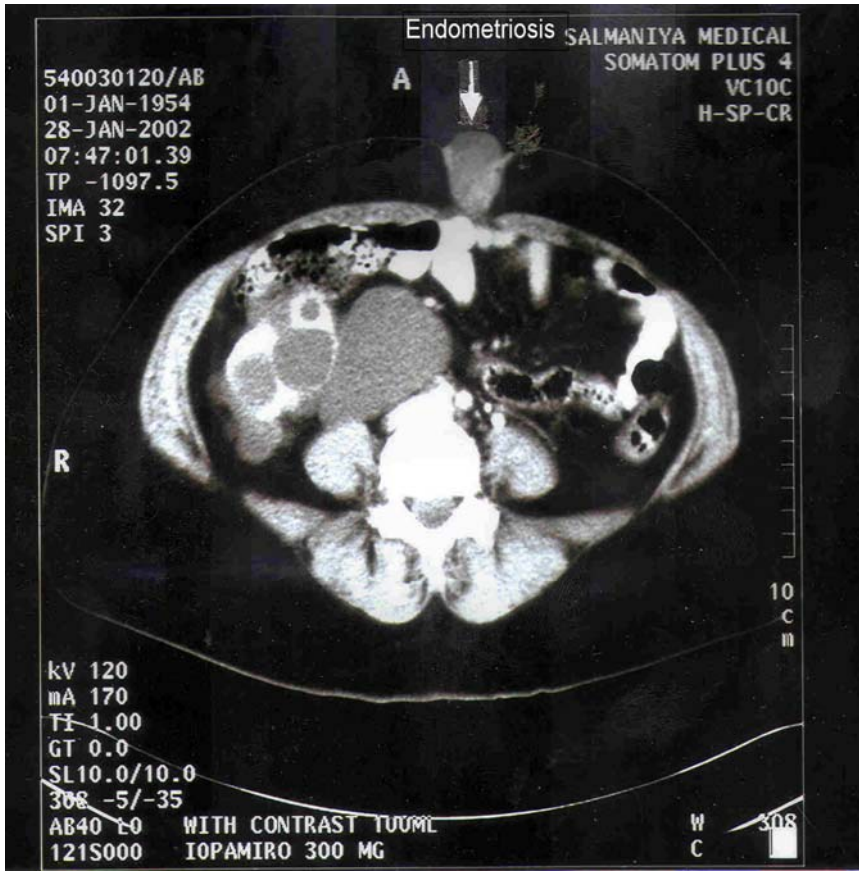


Figure 1: Post contrast and enhanced CT scan of the lower abdomen shows abnormal polycystic right kidney. The isodense nodule at the umbilicus was confirmed to be an endometrioma deposit (case 1)

The patient underwent excision of the umbilical lesion, repair of the para-umbilical hernia and reconstruction of the umbilicus. The final diagnosis confirmed completely excised endometrioma of the umbilicus.

One year later the patient presented again with recurrence of the hernia but not endometriosis. She underwent repair of the incisional hernia and reconstruction of the umbilicus. Follow-up

visits for three years showed no recurrence of the umbilical endometrioma but was still on regular visits to nephrology clinic for renal polycystic disease.

CASE II

Thirty-eight years old unmarried female patient presented to the surgical clinic with painful umbilical nodule of two months duration. The nodule was increasing in size, and bleeding during menstruation. The patient was having regular heavy periods with dysmenorrhea. There was no positive medical, surgical or family history.

On general examination, the patient was healthy. The abdominal examination was normal except for about one cm bluish colored lesion deep in the umbilicus.

All blood investigations were normal. CT scan of abdomen and pelvis were normal, except for an umbilical nodule of less than 2 cm in diameter, but there was no communication to underlying structures or associated hernia (fig. 2). Pelvic organs were normal.

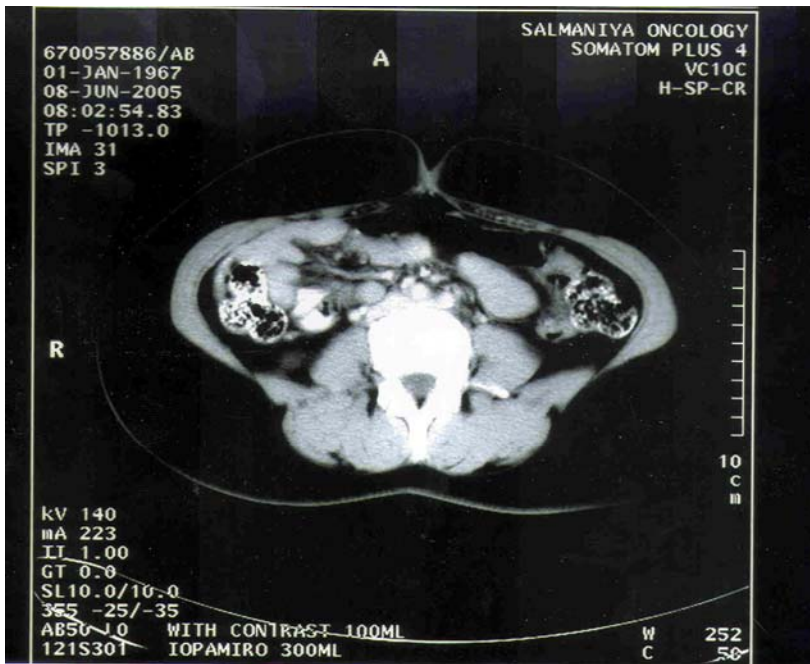


Figure 2: Post contrast enhanced CT scan of the lower abdomen shows isodense lesion at the umbilical stump, later confirmed to be endometrioma (case V)

The nodule was excised. There was no associated hernia. The histopathology confirmed the diagnosis of endometrioma which was completely excised. Patient had an uneventful recovery. No recurrence on regular follow-up visits for two years.

CASE III

Forty-one years old female, married and having five children, presented to the surgical clinic complaining of umbilical swelling of 4 years duration. The swelling increased in size, became painful, and was bleeding during menstruation. The patient had regular but heavy menstrual cycle. There was no significant past medical or surgical history.

On general examination, the patient was over-weight. Abdominal examination revealed bulging out bleeding umbilical nodule of about 5 cm in diameter. There was no associated umbilical hernia.

Blood investigations were normal. CT scans abdomen and pelvis were normal except for the umbilical nodule which was extending to the fascia. Pelvic organs were normal (fig. 3). She underwent complete excision of the mass and the umbilicus down to the fascia with repair of the defect and reconstruction of the umbilicus (Fig 4, 5). Histopathology confirmed the diagnosis of endometrioma.

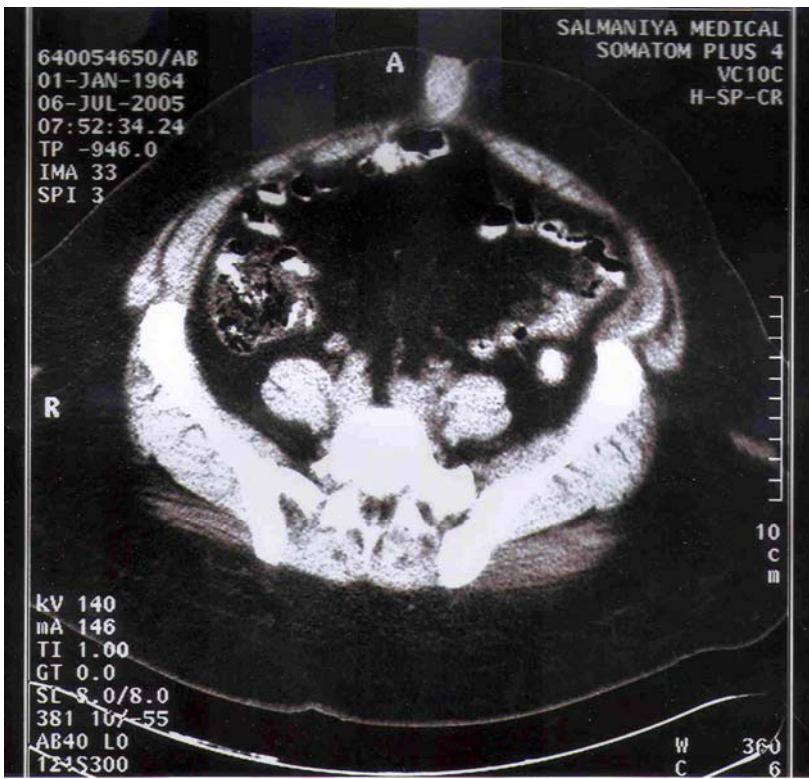


Figure 3: Post contrast CT scan of lower abdomen shows soft tissue nodule at umbilicus; later confirmed to be endometrioma (case VI)



Figure 4: The endometrioma is bulging through the umbilicus; the incision is being marked for excision of the nodule completely down to the fascia and reconstruction of the umbilicus



Figure 5: The umbilical nodule after excision

The patient had an uneventful recovery. Gynecological evaluation was normal. Follow-up visits for two years showed no recurrence and no sign of pelvic endometriosis.

CASE IV

Twenty-nine years old, single and healthy patient was admitted through surgical emergency with diagnosis of acute appendicitis. No significant past history was revealed.

The operative findings were congested appendix, right ovarian cyst (7x5 cm) with partial torsion, normal tubes, and large amount of peritoneal serous fluid. She had appendectomy and right ovarian cystectomy.

Histopathology report showed focal endometriosis of the appendix, and a right ovarian mature cystic teratoma. The patient had uneventful recovery postoperatively. She was seen only once during follow-up.

CASE V

Thirty-nine years old, female patient, known to have primary infertility of 18 years duration presented to surgical emergency with intermittent colicky central abdominal pain radiating to the back of one day duration. She was also having repeated vomiting of three weeks duration. The patient had past history of laparoscopy several times for investigation and release of adhesions.

On examination, there was distension and tenderness over the epigastric region associated with hyperactive bowel sounds. Rectal examination showed empty rectum.

Abdominal x-rays, plain and erect have showed signs of small bowel obstruction. Trial of conservative treatment failed; therefore the patient was taken for diagnostic laparoscopy followed by laparotomy. The findings were extensive omental adhesions, dilated proximal small bowel down to terminal ileum, and a tumor like lesion was seen 10 cm from ileo-ceccal junction. No abnormalities were seen in her pelvic organs. The adhesions were released and the ileal lesion was resected with end-to-end anastomosis.

The histopathology showed endometriosis (tumor like lesion) in the ileum (Fig.6) involving all layers. The patient was kept on Danazol, 200mg four times daily for one year. No recurrence after three years of follow-up.

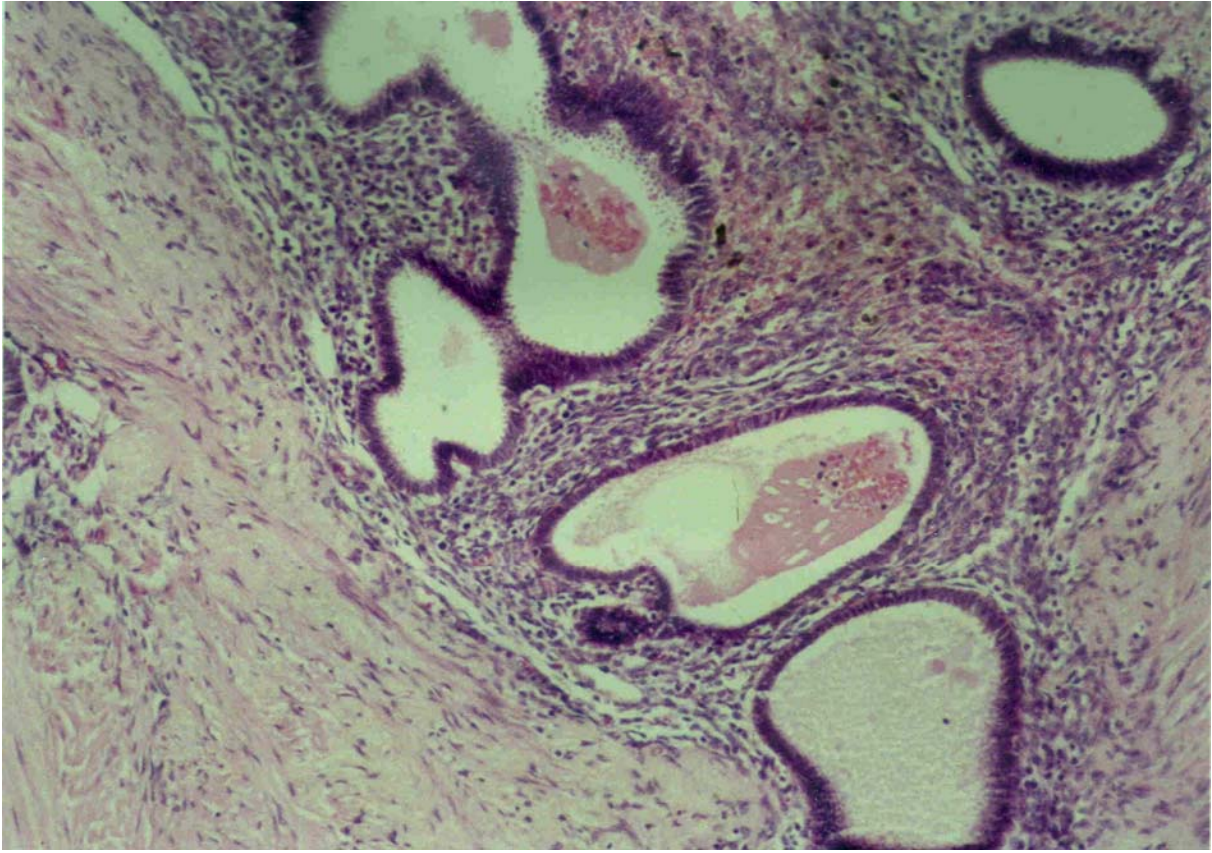


Figure 6: Microphotograph shows mildly dilated endometrial glands with characteristic stroma around and fibrosis towards the periphery. (haematoxylin + Eosin x100) (Case V)

CASE VI

Forty-two years old divorced lady, who was a known case of diabetes, hypertension, and iron deficiency anemia and has seven children, was admitted with a painful mass in the hypogastric region of one year duration, at the site of previous lower segment caesarian section scar. The mass used to increase in size and become tender during each pre-menstrual period.

On examination, the nodule was tender, bluish, and 3x3 cm in size. The patient underwent exploration and complete excision of the mass. Provisional diagnosis of irreducible hernia was thought, but histopathology report confirmed endometrioma. Three years follow-up visits showed no recurrence.

DISCUSSION

Extra-gonadal endometriosis is a disease that is confined to the female population. Interestingly, scattered lesions were found in men exposed to exogenous estrogens. It has no racial predilection. Its exact incidence is not known. Extra-pelvic endometriosis may occur in up to 12% (8-15%) of all menstruating women^{2,4}.

Extra-gonadal endometriosis is rarely diagnosed pre-operatively. Presentations to general surgeons may be atypical and pose diagnostic difficulty, mimicking other acute diseases^{5,6}. More unusual implantation sites can be responsible for bizarre symptoms such as the presentation in our patients. The skin or subcutaneous planes lesion is often slightly tender and painful. At the time of menstruation, the pain becomes more pronounced and may be associated with swelling and slight bleeding of the lesion⁵.

In this study, three patients presented with primary isolated spontaneous umbilical endometriosis. One of them was having co-incidently autosomal dominant polycystic disease of liver and kidneys. Another one was never pregnant and never had pelvic surgery which is very rare^{7,8}. This patient had typical presentation and underwent successful limited resection with no recurrence. Isolated umbilical endometriosis is rare with an estimated incidence of 0.5 to 1% of all patients with endometrial ectopia^{3,9,5,14}. It can occur after surgery but is generally spontaneous¹¹⁻¹⁴. In a retrospective study of 10 patients who underwent laparoscopic assisted subtotal hysterectomy with unprotected removal of uterine segments through the umbilical incision showed that two patients developed umbilical endometriosis¹⁷. This data support the concept that implantation and the potential development of menstrual endometrium is increased after surgery.

One patient in this study had endometrioma in the scar of her lower segment cesarean scar. Although the symptoms were pathognomonic, the pre-operative diagnosis was difficult due to surgeon unawareness. The first case of cesarean section endometrioma in the general surgical literature was reported in 1975¹. Cutaneous endometriosis commonly occurs in a surgical scar from abdominal or pelvic procedures, which include hysterectomy, cesarean section, episiotomy, and laparoscopy⁴. In some studies it was estimated to be 0.1% in surgical scars post cesarean section^{2,10,15,16}. One study stressed that the true incidence of cesarean section scar endometrioma is difficult to determine, but ranges from 0.03% to 0.15%¹.

This study described a case of acute appendicitis which was associated with an ovarian cyst. Histopathology was primary endometriosis of the appendix and the ovarian cyst was a teratoma. Appendiceal endometriosis, while relatively uncommon in patients with endometriosis, it is rare in the general population. In patients with right lower quadrant or pelvic pain, the appendix should be inspected for endometriosis and any evidence of non-gynecologic disease¹⁷.

In this study, there was one patient who presented with acute small bowel obstruction secondary to primary endometriosis. A case was reported of a young Nigerian female diagnosed with chronic intestinal obstruction due to rectosigmoid endometriosis causing stricture. She was successfully treated¹⁸.

Another case report described a pre-menopausal woman with severe constipation causing intermittent obstruction. Colonoscopy revealed a tight rectal stricture; however, mucosal biopsies were normal. Exploratory surgery revealed an intense fibrotic reaction involving the rectum and uterus, necessitating a simultaneous low anterior resection and hysterectomy. Pathology established a diagnosis of endometriosis¹⁹.

The pre-operative diagnosis of rectal endometriosis can be difficult to establish. Endometrial deposits do not invade the mucosa; therefore, colonoscopy with biopsies is non-diagnostic. Surgery may be the only definitive way to obtain the diagnosis. Rectal endometriosis must be included in the differential diagnosis of rectal stricture¹⁹.

In general, diagnosis is usually difficult. This is mainly due to lack of awareness¹. Pathognomonic but not always present is the presence of tumor like palpable lesion in abdominal wall near a preceding surgical scar, the cyclic character of painful symptomatology, and the augmentation of size and bleeding pre and during menstruation.

Ultra-sonography or CT scan with fine needle aspiration can be used for cutaneous and subcutaneous lesions. The definitive diagnosis is usually achieved by surgery and histological examination of the resected lesion. MRI could be the best investigation as it has been shown to be useful for delineating the size and location of extra-pelvic endometriosis and excluding intra-abdominal extension of the disease⁶.

Medical as well as surgical therapies have been used in the treatment of this condition. Medical therapy with Danazol, an anti-gonadotropin, has been used in the treatment of endometriosis¹. When used for cesarean scar endometrioma it has provided temporary relief of symptoms, but does not ablate the lesion¹. Simple surgical excision remains the treatment of choice³. Concurrent pelvic endometriosis needs to be excluded and if present needs to be treated to prevent re-seedling of endometrial tissue from the pelvis⁶. Local recurrence after adequate surgical excision is uncommon.

CONCLUSION

Six cases of extra-gonadal endometriosis with unusual presentation had been reviewed and presented in this study. General surgeons are infrequently involved in the management of extra-pelvic extra-gonadal endometriosis, so the pre-operative diagnosis is not often entertained, predominantly because of lack of awareness. The presence of cyclic symptoms of pain with or without bleeding is almost pathognomonic of the condition. When the diagnosis is made on clinical grounds, although difficult, no further studies are necessary before wide surgical excision is performed.

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