

Endometriosis of Canal of Nuck

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Endometriosis is a common benign gynecological condition defined as the presence of endometrial tissue in tissues outside the uterine cavity. Apart from the common sites of endometriosis, rare sites other have also been reported including the liver, the thoracic cavity, the muscles, nerves, and more rarely in a patent Nuck canal. We aim to evaluate the clinical presentation, diagnostic features, and management of the Nuck endometriosis. A meticulous search of three electronic databases was performed until May 2020 for articles reporting cases of Nuck endometriosis. A total of 36 patients from 20 studies were analyzed. Median age of patients was 36 years with 33 women being of reproductive age. A right-sided lesion was identified in 30 cases (83.3%), while all patients suffer from a groin mass with cyclic pain in a proportion of 22%. All the patients finally underwent surgery for investigation of the lesion and fixation of the defect. Five cases of malignancy were detected at final pathology. All of them were alive with a median reported overall survival of 37 months. Nuck endometriosis should be included in the differential diagnosis of female patients with groin swelling. An evaluation by a gynecologist is important when endometriosis is suspected.

Keywords: nuck cyst ; nuck endometriosis ; canal of nuck

1. Introduction

Endometriosis is a common benign gynecological condition, which is characterized as the presence of endometrial tissue, containing glands and stroma, in tissues outside the uterine cavity ^[1]. The disease affects approximately 10–15% of women of reproductive age ^[1]. Although endometriosis can be asymptomatic, in many cases, it presents with pelvic pain, infertility, or incidentally at histological examination of an ovarian cyst ^[2]. Due to the close relationship of endometriosis with estrogen secretion, postmenopausal women are rarely affected ^[3]. The diagnosis of endometriosis can be suspected by evaluation of cyclic pain and symptoms and can be enhanced with imaging findings ^[4]. In recent years, the role of ultrasonography has gained significant popularity as diagnostic tool in the detection of pelvic endometriosis as well of less common extragenital lesions ^[4]. However, the definite diagnosis is only made by histologic confirmation of the excised specimens. The management of endometriosis is dictated by the severity of the symptoms as well as the wish of infertility correction ^[5]. Treatment options include medical, surgical, or combination of both therapeutic modalities ^[5]. To date, there is not enough evidence available comparing surgical with medical treatment and indicating the potential superiority of one of them. However, initial treatment with pharmaceutical regimens including analgesics (nonsteroidal anti-inflammatory drugs), GnRH analogs, and danazol seems effective in pain relief of a significant proportion of cases ^[6]. Surgical treatment with lesion resection and/or electrocauterization, nerve excision, or hysterectomy and oophorectomy through laparoscopy or laparotomy could be applied in cases of failure of the conservative treatment modalities ^[6]. The incidence of malignant transformation is low, but it is supported that if ovarian endometriomas is left untreated, there is a high probability of malignant transformation into endometrioid and clear cell carcinomas, the incidence of which reportedly ranges 0.6–1% ^{[7][8]}. There are also a few cases of malignant transformation of endometriosis into endometrial stromal sarcoma ^[9]. The most common locations of endometriotic lesions are usually the ovaries, the utero-sacral ligaments, the pouch of Douglas, and the bladder ^[10]. Nevertheless, there are also uncommon intra- and extrapelvic sites of detection of such as in the liver, in the thoracic cavity, in the muscles, and nerves ^{[11][12]}.

The canal of Nuck (NC) was named after Anton Nuck in 1691 and represents the anatomical defect when a parietal peritoneal pouch follows the gubernaculum during its development ^[13]. The lower part of the gubernaculum becomes the round uterine ligament, which inserts the abdominal wall and runs through the inguinal canal along with the processus vaginalis ^[14]. The NC in the female system corresponds to the processus vaginalis in males. Normally the protrusion of parietal peritoneum obliterates during the ninth month of fetal life ^[15], but in cases of incomplete or failure to obliterate, it may manifest as a hydrocele, an indirect hernia or entrapment of various pelvic organs ^[16]. The pathology of the inguinal region includes a variety of diseases and arises from the different groin structures, complicating the differential diagnosis. Despite the fact that the pathology of the NC is not so widespread among either general surgeons or gynecologists due to the rarity of the NC diseases, they have been associated with severe morbidity and they need further investigation ^[16]. For

the investigation of the pathology in the NC, the ultrasonography (US) has been claimed as a fast, inexpensive initial modality with significant diagnostic accuracy, while magnetic resonance imaging is performed in case of controversial US analysis [16]. Other diseases of the inguinal and vulva region such as Bartholin pathology, lymph node enlargement, abscesses, and soft tissue tumors are included in the differential diagnosis of the pathology in the canal of Nuck [16]. Endometriosis in the canal of Nuck is a quite rare surgical entity, which can be a diagnostic challenge in many cases. In worst scenario, the incarcerated endometriotic tissue can be transformed into a malignancy.

2. Discussion

The present study analyzed 36 cases of Nuck endometriosis from a total of 20 studies. Median patient age was 36 years while almost all of them were of reproductive age (92%). A right-side predominance was detected in more than 80% of the lesions of the NC, while all patients underwent surgical exploration of the mass with subsequent excisional procedures. Five cases of malignant tumors arising from the NC endometriosis were recorded. All of the aforementioned patients were alive in a median follow-up of 37 months.

The differential diagnosis of a groin lump or a subcutaneous mass of the inguinal region is challenging and may include a wide variety of entities such as hernia, lymph node enlargement, malignancy, endometriosis, Nuck hydrocele, lipomas, and abscess [17]. The presence of endometriosis in the inguinal canal is rare with an estimated prevalence of 0.3–0.6% of all endometriosis cases [18][19]. The age range of patients presented with groin lump compatible with endometriosis is 22–46 years [19]. If endometriosis in the inguinal region is suspected, it could be endometriosis within the inguinal hernia sac or endometriosis of the NC, the round ligament and the subcutaneous tissue [20]. A limited number of approximately 130 cases of groin endometriosis have been reported in the literature [21]. A painful lump with cyclical pain and enlargement is the most common presentation of endometriosis of the groin region [19]. The clinical presentation is similar irrespective of the tissue from which the endometriosis originates, disabling the differential diagnosis. To that end, preoperative imaging with US and/or MRI could be valuable, despite the fact that surgical exploration and histopathology could clearly set the origin and the final diagnosis of the disease [22].

Diagnosis of endometriosis in the NC is even more uncommon in patients with patent NC due to failure of the canal to close within the first year of life in females [23]. The aforementioned defect can result in hydrocele formation or herniation of intraabdominal surrounding tissues through the canal [23]. Under certain circumstances, which, however, remain unknown, the NC can be infiltrated by endometrial cells and results in the formation of endometriosis. A plethora of theories have been proposed to explain the presence of endometrial-like tissue outside the uterine cavity. One of them suggests that during retrograde menstruation, endometrial cells elude to the pelvic cavity via the fallopian tubes resulting in the formation of endometriotic implants throughout the pelvic structures [24]. This theory can also explain the local spread of endometrial tissue through the inguinal canal and the patent NC. Another theory advocates for the systemic lymphatic or hematogenous spread and aggregation of endometrial cells in distant organs as well as the deposition through the structures of the inguinal canal [19]. Finally, the third and most prevalent theory is that of coelomic metaplasia, which can construe the presence of endometriosis in patients with congenital (Mayer-Rokitansky-Kuester-Hauser syndrome) or iatrogenic lack of uterus (hysterectomized patients) [25]. This theory could be considered the potential mechanism of endometriosis formation in 3 of the included patients who underwent hysterectomy and had no previously known history of endometriosis.

As proved in our study, endometriosis in the canal of Nuck is largely characterized by groin mass enlargement potentially associated with localized pain. It can be misdiagnosed as a typical inguinal hernia or Nuck hydrocele. In some cases, the gradual enlargement of the inguinal mass during menstruation and/or catamenial inguinal pain could be suggestive of the presence of endometriosis. Nonetheless, this was recognized only in a proportion of 8 patients in our series. A meticulous medical history from the patient with the groin mass suspected for endometriosis is critical for the preoperative diagnosis of the disease. In that setting, the patient should be thoroughly questioned about family history of endometriosis, history of any gynecological surgery with special consideration to previous caesarean section or myomectomy, and history of endometriosis and the respective treatment [26]. Patients with history of endometriosis are more likely to have concomitant disease of the inguinal canal through the round ligament. Furthermore, the characteristics of pain including frequency, duration, association with menstruation and location should be also thoroughly asked [26]. Similarly, a proper preoperative imaging is also of critical importance. Ultrasound has been widely utilized for the investigation of the pathology of the Nuck canal [16]. In the majority of the cases presented in this study, the U/S of the affected groin revealed a hypoechoic cystic lesion with internal echoic structures. In some cases, a CT, MRI, or even a PET-CT supplemented the U/S findings. High-resolution U/S is an inexpensive and reliable modality to set the suspicion of diagnosis of the Nuck endometriosis [23]. Nonetheless, the final diagnosis is based on the histology and immunohistochemistry.

We noted a significant predominance of the right-sided endometriotic lesions of the Nuck canal. This finding is in accordance with the findings of previous studies in the pathology of the inguinal canal and the patent NC [27]. The most prevalent reason for this is the clockwise flow of the intraperitoneal fluid, which under the effect of gravity, promotes the prolonged stay of endometriotic cells in the right side and is further enhanced by the protective role of the sigmoid colon on the left side [19][20]. Another less common theory identifies the presence of a lymphatic pathway of atypical lymph nodes from the peritoneal cavity to the right groin which could potentially transfers endometriotic cells to the right-sided groin [19]. However, this does not apply in the case of right sided predominance of pelvic endometriosis [19].

As shown in our study, surgical resection with simultaneous repair of the defect of the canal in the majority of cases is the optimal treatment option for patients with endometriosis of the NC. Similarly to the case of Nuck hydrocele, the gold standard treatment remains excision of the cystic structure (laparoscopic or open) and closure of the internal inguinal defect with or without mesh [23]. Additionally, in case of extension of the disease to the labia majora, a concomitant repair of the vulva may be necessary including vulvectomy or clitorrectomy procedures depending on the extent of the disease [28]. The type of surgical procedure varies from transabdominal laparoscopic or open approach to excisional biopsy of the tumor through an inguinal approach. The approach is largely determined by the clinician's estimation of the patient's medical history, physical examination, symptomatology, laboratory testing, and preoperative imaging. The surgeon should also be aware so as to avoid dissemination of endometriotic cells to the surrounding tissues during surgical manipulations in order to reduce recurrences [19]. Additionally, the postoperative administration of hormonal suppression therapy is a matter of controversy. A variety of hormone suppression regimens including combined oral contraceptives, androgens, progestins, GnRH analogs, and levonorgestrel intrauterine device have been proposed for the decrease in disease recurrence after surgery for endometriosis [29]. According to a recent systematic review and meta-analysis, the risk of endometriosis recurrence was significantly reduced in patients who postoperatively received any kind of hormonal suppression compared to those who received placebo or no therapy (1766 patients, data from 14 studies RR 0.41, 95% CI 0.26 to 0.65, $p < 0.01$) [29]. The same was also observed in case of postoperative pain, which was significantly lower in the hormonal suppression group (652 patients, data from 7 studies, SMD -0.49, 95% CI: -0.91 to -0.07, $p < 0.01$) [29]. Nonetheless, due to the limited cases of endometriosis related to groin, there is no accurate data on the proportion of patients who received postoperative hormone therapy as well as on the respective recurrence rates.

Given the rarity of the presence of a patent NC, the detection of malignancy is an even rarer finding. Only a limited number of cases with malignant tumors arising from Nuck endometriosis have been reported in the literature. We herein analyzed a total of 5 cases with malignant disease. To date, no consensus has been reached with regards to the optimal management of Nuck endometriosis-related malignancy. Based on the available cases, complete surgical resection is the preferred treatment of the primary lesion. Among the 3 patients who developed recurrence, one had local recurrence, one had distant lung metastasis, while the remaining patient had initially local recurrence and developed lung metastasis 2 years after the first surgery.

The present review is, to the best of our knowledge, the first in the literature, which presents a cumulative report of cases of endometriosis developed in the NC. A meticulous search of the literature eliminated the risk of potential loss of articles. However, there are some limitations that need to be addressed before reaching to safe conclusions. First of all, the estimation of the exact prevalence of Nuck endometriosis could not be precisely achieved, while robust conclusions concerning the pathophysiologic pathway of Nuck endometriosis formation, the clinical appearance of the disease, as well as the optimal treatment could not be reached. The currently available outcomes are limited to case reports and small cases series of moderate quality, which precludes further research due to the rare entity. Furthermore, there is significant heterogeneity among the included studies and some parameters were omitted by some of them which consists another critical factor to aggregate the currently available knowledge in the field. Finally, no sufficient information was available by the recruited studies with regards to the perioperative use of hormonal replacement therapy. More specifically, in the study by Uno et al., the preoperative hormonal therapy was prescribed aimed to reduce the size of the lesion without, however, any significant impact, whereas Wang et al. postoperatively prescribed a 3-month gonadotropin-releasing hormone agonist [30][31].

3. Conclusions

Despite its rarity, the Nuck endometriosis is an existing entity, which should not be neglected when investigating the pathology of the NC. The presence of endometriosis in the NC should be considered in the differential diagnosis of groin lump in female patients. Due to the fact that the pathology of the groin is mostly investigated by general surgeons, they should thoroughly ask for patients' previous medical history and complaints with regards to the disease and consult a

gynecologist when they suspect the presence of endometriosis. Additionally, the role of imaging and preoperative designation of the diagnosis should be highlighted so as for the cases of the presence of malignancy to appropriately design the management of the disease.

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