The occurrence of neuroma in the pelvicarea in women – a review of the literature and case report

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Original article

DOBROSŁAWA L. SIKORA-SZCZĘŚNIAK[1], GRZEGORZ SZCZĘŚNIAK[2], WACŁAW SIKORA[1]

[1] Oddział Ginekologiczno-Położniczy, Radomski Szpital Specjalistyczny w Radomiu
Ordynator Oddziału: lek. Mieczysław Szatanek

[2] Oddział Chirurgii Ogólnej, Radomski Szpital Specjalistyczny w Radomiu
Ordynator Oddziału: dr n. med. Robert Rojewski

Address for correspondence:
Dobrosława L. Sikora-Szczęśniak
Oddział Ginekologiczno-Położniczy, Radomski Szpital Specjalistyczny w Radomiu
ul. Tochtermana 1, 26-600 Radom
tel. (48) 361 51 23, fax (48) 361 52 33, e-mail: dosiass@wp.pl

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Summary

Introduction. Neuroma (neurilemmoma, schwannoma) is derived from neurolemmocytes of peripheral nerves, usually sensory but also motor. Histologically, therearetwotypes of neuroma: Antoni A, with a solid structure and the presence of nuclearpalisades (Verocaybodies), and Antoni B, with loosecellularsystems and the presence of pseudocysticchanges with myxoidelements. It isbenign and extremelyrarelyundergoesmalignanttransformation. Degeneratedneurilemmoma (ancientschwannoma) is a rarevariant of neuroma, characterized by a reduction in cellnumber, degenerativechanges (cysts, calcification), bleeding and hyalinization. Thesechangesareattributed to "aging" as a result of tumor growth, leading to vascularinsufficiency. Histologically, ancientschwannomaischaracterized by a significantdecrease in the size of AntoniAarea (rich in cells) and expansion of Antoni B type.

Purpose. A review ofliteratureconcerningthe occurrence of neuroma in the retroperitoneal space of the pelvis minor and in the female internal genital organs, including the period of pregnancy.

Materials and methods. A review of therelevantliterature. It includedacase of neuroma treatedsurgically in the Department-a variant of ancientschwannoma.

*Results.*Onlyfewreports on the occurrence of neuroma in the pelvis minor werefound. The treatment of choicein the case of neuroma, includingancientschwannoma, is a completesurgicalexcision, whichisoftenassociated with numerous intra- and postoperativecomplications. In the presentedcase, ancientschwannomalocated in the retroperitonealspace of the pelvis minor (perianal-presacral) was anobstacle for parturition. After performinganelectivecaesareansection (cc), the tumor was removed and peripartumhysterectomy was performed. In the literature,therewere no publicationsconcerning neuroma in thislocation, treatedsurgicallyduring cc in combination with peripartumhysterectomy.

Conclusions. Neuromaslocated in thepelvisminoraredifficulttodiagnose; prior to surgery,theyareusuallydiagnosedincidentallyorwhentheirsizecauses the "mass effect". Surgicalprocedures, due to theircomplexity, oftenrequire the involvement of multidisciplinaryteams of doctors and securinglargeamounts of blood and blood products.

Keywords: retroperitoneal ancient schwannoma; caesareansection; peripartumhysterectomy

INTRODUCTION

Neuromas (neurilemmomas, schwannomas) arise from neurolemmocytes (of neuroectoderm origin) of peripheral nerves, except for cranial nerve I and II. They usually derive from sensory nerves but motor nerve origin has also been reported [1, 2]. Histologically, there are two types of neuromas. The Antoni A type has a solid structure with nuclear palisades (Verocay bodies). The Antoni B type is characterized by loose cellular systems with pseudocystic changes with myxoid elements. They are benign lesions and extremely rarely undergo malignant transformation.

Neuromas in the retroperitoneal space are rare. According to various authors, they account for 0.3–5.0% of all neuromas [1, 3, 4]. Tumors in such localization are usually found in patients between the age of 40–60, more often in women [5]. Hooja et al. and Wong et al. quote opinions of other authors who claim that these are the most uncommon tumors found in this region [6, 7]. Malignant neuromas constitute approximately 1% of all cases. They are very aggressive and insensitive to chemo- and radiotherapy [7]. The location in the retroperitoneal space of the pelvis minor is extremely rare and accounts for < 1% of all benign neuromas [1, 8]. Due to their slow growth and anatomical localization in the retroperitoneal space, these tumors are asymptomatic until they start compressing (the mass effect) the surrounding organs thus producing symptoms [1, 9]. If a tumor of this type is located in the pelvis, the symptoms can include abdominal pain, discomfort as well as tightness in this region and in the lower back. Urinary and gastrointestinal symptoms caused by the compression on the urinary bladder and intestine include: pollakiuria, urinary incontinence or acute urinary retention, hydronephrosis, bloating, nausea and vomiting [7, 9, 10, 11. 12].

Degenerated neurilemmomas (ancient schwannomas) are a rare variant of neuromas, accounting for 0.8% of soft tissue tumors [3, 13]. Histologically, they are characterized by a considerable reduction in the Antoni A area (hypercellular area) and expansion of the Antoni B area with a reduction in cells with large, hyperchromatic and segmented nuclei without mitosis. Degenerative changes (cysts, calcification and sometimes ossification), bleeding and hyalinization can be observed. These changes are attributed to "aging" resulting from tumor growth that leads to vascular insufficiency and "spontaneous degeneration" [12, 14, 15]. Ancient schwannomas in women usually develop at the age of 20–50 [16]. Up to 20% of these tumors are associated with neurofibromatosis type 1 [3]. These neoplasms in the retroperitoneal space are usually larger and behave as neuromas. Their malignant transformation is uncommon [1, 17].

The removal of this type of a tumor from the presacral retroperitoneal space is associated with certain surgical difficulties resulting from limited access and poor visualization in the pelvis [9]. Complications after the removal of this type of a neoplasm located in the pelvis minor include hemorrhage due to the difficulty in obtaining hemostasis when the presacral venous plexus is damaged, and injury to the nerves of the pelvic and sacral plexuses, which occurs during the removal of neuromas [1, 9, 12]. Intraoperative hemorrhage is the most serious complication of such procedures. Fatal hemorrhages during surgeries or withdrawing from performing the procedure due to the risk of uncontrollable bleeding have been reported [5, 12]. Currently, such procedures can be conducted laparoscopically, and recently, also with the use of a robot (da Vinci system). Authors emphasize advantages of laparoscopic surgery, such as: minimal invasiveness, better visualization of the surgical field, minimal intraoperative blood loss and short hospitalization after the procedure [13, 18, 19, 20, 21, 22].

Due to excessive vascularization and topographic changes concerning pelvic organs surrounding the uterus, peripartum hysterectomy is associated with a risk of their injury, considerable blood loss and coagulation disorders resulting in the necessity of multiple blood and blood product transfusions [23]. The final diagnosis is established on the basis of histological and immunohistochemical examinations (positive staining for S-100 protein as well as negative staining for CD117 and smooth muscle actin) [5, 6, 20].

AIM

The aim of the paper was to review the literature on the cases of schwannomas in the retroperitoneal space of the pelvis minor in women.

MATERIAL AND METHODS

The authors analyzed the individual literature reports on the cases of schwannomas in women diagnosed due to various complaints concerning the reproductive organs. The second group contained publications on this condition during pregnancy, including a presentation of a case treated in our center. Attention was paid to diagnostic methods and difficulties in establishing a correct pre- and intraoperative diagnosis.

RESULTS

A case of neuroma in the uterus of a 67-year-old patient was reported by Theodosopoulos et al. [5]. Machairiotis et al. presented a case of right ovarian schwannoma localized in the region of the right parametrium of a 58-year-old woman [10]. Durian et al. noted the occurrence of fallopian tube schwannoma localized in the region of the parametrium [25]. Moreover, cases diagnosed preoperatively as internal reproductive organ pathologies (uterine fibroid, ovarian tumors) that ultimately turned out to be pelvic retroperitoneal neuromas were presented by, among others, Aran et al. [11], Ninshu et al. [18], Chen et al. [24], Tageuchi et al. [26] and Korkontzelos et al. [27].

There have been only a few case reports of retroperitoneal neuromas detected in pregnant patients, and diagnosed preoperatively as dermoid cysts or ovarian tumors. Bardeguez et al. were the first to present such a case [28]. Andrews S. et al. stated in their publication that their case presentation concerning neuroma in the pelvis minor during pregnancy was the third case of this type reported in English literature [29]. Van Wijngaarden et al. described a patient with pelvic neurilemmoma diagnosed intraoperatively during laparotomy in a primipara operated in the 16th week of gestation, who had been preoperatively diagnosed with an ovarian dermoid cyst. Subsequently, it was an indication for an elective cesarean section [30]. Insegno et al. noted a case of a patient in the 35th week of gestation, in whom pelvic neuroma caused small intestine intussusception and obstruction. Early surgical treatment helped avoid complications [31]. Ibraheim et al. described a case of a woman operated ineffectively in the second trimester of gestation and effectively during the subsequent (the third) laparotomy following an elective cesarean section [32]. Parveen et al. also presented a description of a procedure conducted to remove neuroma performed in the second trimester of gestation [33]. Dahiya et al. reported a case of a primipara in the 22nd week of gestation in whom the surgery due to abdominal retroperitoneal tumor (neuroma) necessitated the resection of the bowel and performance of a colostomy. Six weeks after labor, which took place in the 35th week of gestation, surgical anastomosis of the bowel was performed [34].

CASE PRESENTATION

The patient (K.W.), aged 37, in the 39th week of her fifth pregnancy, who had had four natural labors, was admitted to the Department on March 21, 2005. The obstetric examination revealed a tumor in the region of the pouch of Douglas that constituted a birth obstruction. An obstetric ultrasound scan demonstrated the presence of a solid tumor with the size of 100x80 mm. During the pregnancy, ultrasound scans conducted in week 25, 31 and 37 revealed a tumor that was referred to as fibroid on the posterior uterine wall with the size of 80x66, 81x67 and 91x75 mm in the respective scans. The patient reported symptoms that she had not experienced during her previous pregnancies. These were: sacrolumbar pain, constipation, more frequent urination and, in the third trimester, vaginal and perineal varices causing pain in this region. The surgery was planned on the following day, and the necessary amounts of packed RBCs and blood products were secured. The abdominal cavity was opened with a lower midline incision. The cesarean section was performed in a typical way. The female neonate was in a good condition (3970 g/56 cm; Apgar 10). After wound debridement and uterus exposure, a solid tumor with the size of 110x80x60 mm was detected in the pelvic retroperitoneal space (in the perirectal, presacral region). The tumor was enucleated. Because of excessive bleeding from the tumor bed that was difficult to control, the uterus was removed without the adnexa in the typical manner to enable better access and facilitate the management of hemorrhage. It was found that the left internal iliac vein was injured. The injury was repaired, the site of bleeding was managed and hemostasis was obtained. Setons were inserted to the tumor bed through the vagina, next to a drain. Following three hours of surgery and four hours of observation in the ICU, hemorrhagic shock developed. The patient was re-operated. Approximately 2,500 ml of blood with clots was detected in the peritoneal cavity. Again, bleeding originated from the left internal iliac vein and tumor bed. The injured vein was managed with a hemostatic suture. Hemostasis of bleeding sites was obtained by applying sutures, using electrocoagulation, surgicel and spongostan. The tumor bed was filled with gauze setons introduced by a separate incision in the abdominal wall. A drain was inserted to the pouch of Douglas. During the surgery and in the postoperative period, the patient received 16 units of packed RBCs and 5 units of FFP. Histological diagnosis established in the Department of Pathomorphology (No 693809): *Degenerated neurilemmoma /ancient schwannoma/*. There are no features of malignancy. The patient was discharged on day 10 after the surgery.

DISCUSSION

Imaging examinations, such as US, CT, MRI or PET-CT, play a significant role in the preoperative diagnosis of retroperitoneal neoplasms in the pelvis minor, but not all of them can be conducted during pregnancy. CT can reveal degenerated areas in neuromas. MRI presents neuromas as masses of low signal intensity in T1-weighed images and high signal intensity in T2-weighted images. In the latter, signal intensity depends on the "cellular density" in neuroma. Hypercellular regions are characterized by intermediate signals whereas type B tissues give a bright signal [16]. MRI (particularly with gadolinum) is more specific than CT. These examinations deliver information about the localization, size of a tumor and possible invasion into the surrounding structures, all of which are important in the planning of surgery [17].

Ultrasound or CT-guided fine-needle aspiration biopsy (EUS-FNA or CT-FNA) can also be applied. However, opinions concerning such procedures are controversial. This is because cellular pleomorphism detected in degenerated areas can be misinterpreted as a feature of malignancy. Moreover, there is a risk of bleeding, infection and neoplastic spread [5, 14, 35]. Scintigraphy is also listed among useful examinations in the preoperative diagnosis of ancient schwannomas [15].

Hijoka et al. have described the usefulness of EUS-FNA in the diagnosis of retroperitoneal neuromas, particularly the small ones that are difficult to detect in US or CT [35].

CT angiography enables to determine the blood supply to the tumor thus facilitating hemostasis during the surgery, or embolization (in order to reduce the tumor mass) and, subsequently, its radical removal. Such management is not, however, commonly applied [5]. The imaging modalities listed above can be helpful in obtaining information about the tumor size, localization and relation to the adjacent tissues, but they do not provide the final diagnosis [5, 24]. They enable precise planning and safe performance of a surgical procedure.

Despite accurate imaging, the final diagnosis of retroperitoneal neuroma, particularly in the pelvis minor, is uncertain. That is why, neuromas located in the internal reproductive organs and retroperitoneal space of the pelvis minor are usually diagnosed in histological tests of surgical specimens.

The radical removal of the mass in laparotomy is sufficient treatment of choice of neuromas, also in the case of ancient schwannoma [1]. Some authors believe that that simple enucleation of neuroma, as in ancient schwannoma-type tumors, can also be effective [3]. However, most authors do not recommend such management [16].

Cases of hysterectomy due to neuromas in the region of the parametria, in the pelvic retroperitoneal space, or in the uterus, ovary or ovarian tube, which are diagnosed only in a histological analysis of a surgical specimen, are not presented frequently because of the rare occurrence of such lesions in the pelvis minor [5, 10, 11, 24, 25].

Tumors in the pelvic retroperitoneal space are rarely listed among indications for elective peripartum hysterectomy.

In the case presented above, non-malignant ancient schwannoma in the perirectal and presacral region of the pelvic retroperitoneal space was a birth obstacle. Its localization necessitated elective term cesarean delivery. Following its removal, peripartum hysterectomy was necessary to obtain conditions for hemostasis.

The authors have not found any similar reports concerning this type of a tumor in the pelvic retroperitoneal space removed during a cesarean delivery with simultaneous peripartum hysterectomy.

In the case discussed, uterine fibroid was the only tumor taken into account and therefore imaging was limited to ultrasound only. The simultaneous performance of three major surgical procedures and the necessity of reoperation required efficient cooperation of doctors of various fields and other health care professionals, particularly in terms of securing blood and blood products. Moreover, there were no neurological complications, which are possible during such surgeries due to the difficulty in identifying nerves [12].

During a ten-year observation period, no complications of the procedure have been observed, and the tumor has not recurred. In the latest US examinations (on June 6, 2011 and January 1, 2013), no pathological lesions were detected in the pelvis minor, abdominal cavity and retroperitoneal space.

CONCLUSIONS

1. Neuromas located in the pelvis minor are difficult to diagnose. Prior to surgery, they are usually diagnosed incidentally or when their size causes the „mass effect."
2. Due to their complexity, such surgical procedures often require the involvement of multidisciplinary teams of doctors and securing large amounts of blood and blood products.

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