EJGO European Journal of Gynaecological Oncology

Magnetic resonance imaging of a vaginal paraganglioma and related literature review

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Summary

Purpose: To investigate the magnetic resonance imaging (MRI) findings of paraganglioma (PGL). *Materials and Methods:* A 17-year-old girl was admitted to the present hospital with a palpable painless mass in her right vulva that gradually grew. The preoperative contrast enhanced MRI revealed an irregular lump with heterogeneous hyperintensity on T2-weighted images in the lower pole of the right labia majora, with heterogeneous hyperintensity on diffusion weighted image (DWI), and significantly enhanced hyperintensity after administration of contrast agent. *Results:* The patient underwent surgery and postoperative pathology confirmed the diagnosis of vaginal PGL. *Discussion:* Vaginal PGLs are rare and lack specific symptoms. For round or irregular masses within the vagina, which manifest with hyperintensity or with mixed signal on T2WI and DWI with well-defined boundaries, and obvious heterogeneous enhancement in contrast-enhanced MR scanning, PGL should be considered.

Key words: Vaginal paraganglioma; Neuroendocrine tumor; Clinical features; Computed tomography (CT); Magnetic resonance imaging (MRI).

Introduction

Pheochromocytoma (PCC) is a neuroendocrine tumor originating from the chromaffin cells of the medulla of the adrenal glands or extra-adrenal chromaffin tissues and can secrete catecholamines [1]. Extra-adrenal PCC is termed paraganglioma (PGL), and accounts for about 10% of all PCCs [1]. PGL is a rare tumor derived from the parasympathetic or sympathetic nervous system that originates from the neural crest, which can occur at various sites between the neck to the pelvis at parasympathetic ganglions or the sympathetic chain [1]. PGLs of the female genital tract are exceptionally rare, but can originate from the ovaries, uterus, cervix, or vagina [2]. Vaginal PGLs are extremely rare and only a few cases of vaginal PGL have been previously reported in case studies.

Preoperative diagnosis of PGLs can be aided by a variety of anatomical and functional imaging techniques [1]. In contrast to ultrasound and computed tomography (CT), magnetic resonance imaging (MRI) is the morphological imaging modality of choice for localization of PCC and PGL, and can provide excellent anatomic details due to its high-contrast resolution, capacity to scan in multiple planes, and lack of ionizing radiation [3]. Herein the authors report a 17-year-old girl with a painless palpable mass in her right vulva. Preoperative contrast enhanced MRI clearly revealed the features of the tumor, and puncture biopsy and postoperative pathology confirmed the diagnosis of vaginal PGL.

Case Report

A 17-year-old girl was admitted to the present hospital with a vulval mass that had persisted for two years, and gradually enlarged for one year. Her menarche had occurred at the age of 13. The length of her menstrual cycle was 30 days, lasting for four to ten days and flow varied, but without dysmenorrhea. She discovered a mass with a diameter of two cm in her right vulva two years ago, but since she experienced no pain or discomfort, she did not see a doctor. However, the mass had noticeably grown within the last year, and was associated with pain after activity. The patient's blood pressure was 140/106 mmHg, body temperature, heart rate, and respiration rate were normal, and no substantial change in body weight was reported.

Serum level of free thyroxine 4 (FT4) was 10.95 pmol/L and cortisol was 8.0 ug/dl. The preoperative serum catecholamine test indicated that the noradrenaline (NAD) was 3,747.16 pg/ml and dopamine was 167.85 pg/ml, and the preoperative urine catecholamine test indicated that the NAD level exceeded 450 μg/day. The patient's blood pressure was elevated before surgery, even after antihypertensive treatment, and the abdominal CT indicated thickening of the left adrenal gland.

The gynecological examination revealed that an irregular lump of 5×3 cm was palpable in the lower pole of right labia majora and lateral to the hymen, without tenderness. Ultrasonography revealed an irregular solid mass of 47×33 mm in the subcutaneous tissue of the right labia majora, which extended into the muscular layer. The mass had an unclear boundary, heterogeneous echogenicity, and color Doppler flow imaging indicated rich vascularization. CT images made at another hospital indicated a mass in the right side of the vagina and anal region which involved surrounding tissues, and the uterus was displaced to the left and hydrometra existed.

The axial (A), sagittal (B), and coronary (C) FS-T2 weighted

Revised manuscript accepted for publication March 29, 2016

Eur. J. Gynaecol. Oncol. - ISSN: 0392-2936
XXXIX, n. 2, 2018
doi: 10.12892/ejgo3657.2018

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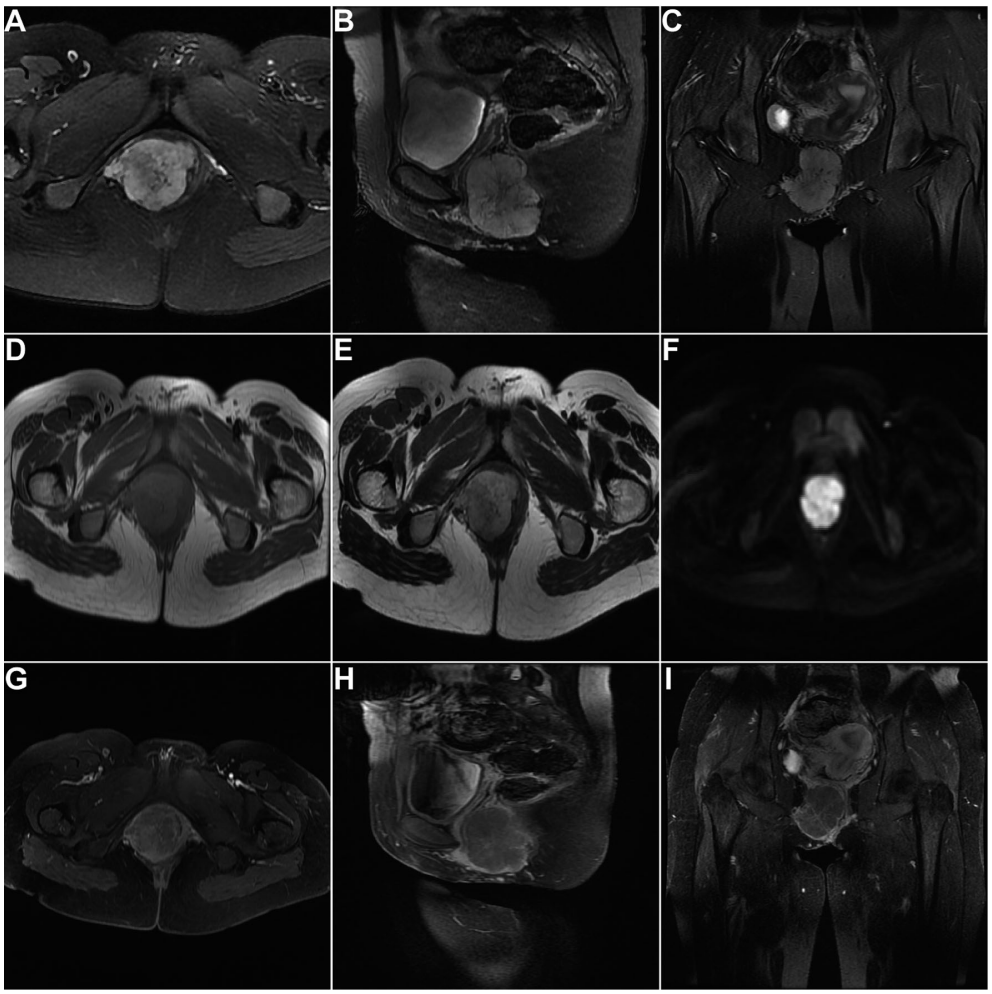

Figure 1. — The axial (A), sagittal (B), and coronary (C) FS-T2 weighted images displayed an irregular lump with heterogeneous hyperintensity in the lower pole of the right labia majora, which involved the anal sphincter. The in-phase (D) and out-phase (E) T1 weighted images showed there was no obvious signal decreasing. The mass revealed obvious inhomogeneous hyperintensity on diffusion weighted image (DWI) (F). The axial (G), sagittal (H), and coronary (I) contrast enhanced T1 weighted images showed that the mass was significantly enhanced.

images (T2WI) of MRI examination (Figure 1) indicated an irregular lump with heterogeneous hyperintensity in the lower pole of the right labia majora, which involved the anal sphincter. The in-phase (D) and out-phase (E) T1-weighted images (T1WI) indicated no obvious signal decrease. The mass displayed inhomogeneous hyperintensity on diffusion weighted image (DWI) (F). The axial (G), sagittal (H), and coronary (I) contrast enhanced T1WI improved visualization of the mass, and its relationship with surrounding tissues.

Cytological puncture examination, under ultrasound guidance revealed a clear cell carcinoma. Surgery was performed with the patient in lithotomy position after general anesthesia. Surgical exploration revealed that the vulva was normotrophic, and a soft mass of 8×6 cm was palpable in the middle and lower segment of the right labia majora. The mass was with rich blood supply at the surface, and with limited mobility. The mass bulged slightly outward, extruded within the vagina and the right side of the vaginal wall was thinned. Rectoabdominal examination revealed that the mass was closely related to the rectum and involved the anal sphincter. The mass was obliquely sliced to a depth of about six cm from the surface with an electric knife, separated from the surrounding tissue, and completely removed. Pathological diagnosis confirmed that the mass was vaginal PGL, with a total size of 7×4.5 cm. Immunohistochemical staining indicated that the mass was CgA (+), Syn (+) and S-100 (-).

The patient was followed up one week after surgery, the levels of noradrenaline and dopamine in the her urine were within normal limits, and her blood pressure was normal. No hypertension occurred prior to discharge.

Discussion

PGL is a rare neuroendocrine tumor derived from the paraganglia, accounting for only 0.012% of all neoplasms [1, 4]. Most PGLs are either asymptomatic or present as a painless mass. In this report the authors describe a patient in which PGL manifested as a painless mass in her right labia majora, which gradually grew. Only 1–3% of PGLs secrete clinically significant levels of hormones, and the symptoms of PGL resemble PCC [1]. The present patient had an endocrinologically active vaginal PGL, manifesting with refractory hypertension during her adolescence, associated with significantly increased catecholamine levels in both serum and urine. Catecholamine levels were reduced to normal after complete resection

Table 1. — *Cases of perineal paraganglioma reported in the literature.*

Year	Author	Position	Age	Symptoms	Size (cm)
1991	Colgan *et al.* [2]	Nympha	58	Vulvar pain	1
1998	Parkes *et al.* [6]	Vagina	11	Severe vaginal bleeding, hypertension	-
2003	Hassan *et al.* [5]	Vagina	24	A vaginal mass found during routine pelvic examination, hypertension (217/110 mmHg)	3
2007	Brustmann [7]	Vagina	33	Vaginal bleeding	1.9×1.4
2008	Shen *et al.* [8]	Vagina	38	Headaches, chest distress, and palpitations for 16 years	3 cm
2010	Akl *et al.* [9]	Vagina	65	A pelvic mass found during routine examination	3×2 cm
2013	Liu *et al.* [4]	Vulva	48	A lump found in the genitals	3.2×2.3×1.5
2014	Cai *et al.* [10]	Vagina	17	Vaginal bleeding - hypertension (140/110 mmHg)	4×3

of the tumor.

PGL of the female genital tract is extremely rare, but a few case reports describe PGL originating from the uterus, ovary, vagina, and cervix [5]. The present authors identified eight reports of female genital tract PGL published between 1991 and 2014, including one case of PGL in the nympha and others in the vagina (Table 1) [2, 4-10]. While most articles carefully reported immunohistochemical and pathological findings, one of these reports included imaging of PGL in the female genital tract. Thus, to the best of the authors' knowledge, this is the first case report presenting imaging of PGL in the female genital tract.

As CT and pelvic sonography provide limited information regarding abnormalities of the vagina, MRI is currently the most useful imaging method for assessment of female genital tract pathology, particularly for the uterus and vagina [1, 3]. The vagina and surrounding tissues can be clearly visualized by MRI [3]. Although the present patient received pelvic sonography in another hospital before admission to this hospital, and the pelvic sonography in this hospital was unavailable to the present authors. In the present patient vaginal PGL manifested as an irregular lump in the right labia majora with heterogeneous hyperintensity on T2WI, and obvious inhomogeneous hyperintensity on DWI, which was significantly enhanced after administration of contrast agent. The tumor involved the anal sphincter and its relationship with surrounding tissues was clearly visualized on multiplanar MRIs, allowing the development of an accurate surgical plan. Vaginal MRI is not only important for characterizing imaging features of vaginal masses and designing the therapeutic approach, but can also be used to assess post-treatment changes [3].

PGL that occurs in the female genital tract needs to be differentiated from Bartholin's gland cyst, vaginal carcinoma, and metastases [3]. Bartholin's cyst is the most common cystic lesion occurring in the vulva, mostly during the reproductive years. Although many patients are asymptomatic, cysts can be associated with significant discomfort and disruption of sexual function and daily activities. Bartholin's cysts usually manifest as well-defined hyper intense lesions with a thin wall on T2WI, with variable signals on T1WI and no rim enhancement [3]. Primary vaginal malignancies, which are rare and usually occur only in elderly patients, usually manifest as painless vaginal bleeding, vaginal discharge, urinary symptoms, pelvic pain or with a painless mass, although up to 30% of vaginal malignancies are asymptomatic. About 90% of primary vaginal malignancies are squamous cell carcinomas, and other tumors, e.g. adenocarcinomas, melanomas, sarcomas, and PGL are very rare [3]. MRI plays a prominent role in local staging of vaginal cancer, and is useful for the planning of therapeutic options. Most tumors affecting the vagina are secondary to primary cervical, vulval, bladder or rectal carcinomas. Metastases to vagina from tumors of extragenital organs are unusual, and mostly originate from adenocarcinomas of the gastrointestinal system [3].

Although vaginal PGLs are rare and lack specific symptoms, fortunately, we have characterized the MRI of these tumors. For round or irregular masses within the vagina, which manifest with hyperintensity or with mixed signal on T2WI and DWI with well-defined boundaries, and obvious heterogeneous enhancement in contrast-enhanced MRI scanning, PGL should be considered. MRI is particularly helpful for preoperative diagnosis of PGL in the female genital tract.

References

[1] Brink I., Hoegerle S., Klisch J., Bley T.A.: "Imaging of pheochromocytoma and paraganglioma". *Fam. Cancer*, 2005, *4*, 61.

[2] Colgan T.J., Dardick I., O'Connell G.: "Paraganglioma of the vulva". *Int. J. Gynecol. Pathol*., 1991, *10*, 203.

[3] Grant L.A., Sala E., Griffin N.: "Congenital and acquired conditions of the vulva and vagina on magnetic resonance imaging: a pictorial review". *Semin Ultrasound. CT MR*, 2010, *31*, 347.

[4] Liu Y.Q., Yue J.Q.: "Paraganglioma of the vulva: a case report and review of the literature". *Int. J. Clin. Exp. Pathol*., 2013, *6*, 2247.

[5] Hassan A., Bennet A., Bhalla S., Ylagan L.R., Mutch D., Dehner L.P.: "Paraganglioma of the vagina: report of a case, including immunohistochemical and ultrastructural findings". *Int. J. Gynecol. Pathol*., 2003, *22*, 404.

[6] Parkes S.E., Raafat F., Morland B.J.: "Paraganglioma of the vagina: the first report of a rare tumor in a child". *Pediatr. Hematol. Oncol*., 1998, *15*, 545.

[7] Brustmann H.: "Paraganglioma of the vagina: report of a case". *Pathol. Res. Pract*., 2007, *203*, 189.

[8] Shen J.G., Chen Y.X., Xu D.Y., Feng Y.F., Tong Z.H.: "Vaginal paraganglioma presenting as a gynecologic mass: case report". *Eur. J. Gynaecol. Oncol*., 2008, *29*, 184.
[9] Akl M.N., Naidu S.G., McCullough A.E., Magtibay P.M.: "Vaginal paraganglioma presenting as a pelvic mass". *Surgery*, 2010, *147*, 169.
[10] Cai T., Li Y., Jiang Q., Wang D., Huang Y.: "Paraganglioma of the vagina: a case report and review of the literature". *Onco. Targets Ther*., 2014, *7*, 965.

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