Vaginal paraganglioma presenting as a gynecologic mass: case report

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Summary

Paragangliomas in the vagina are extremely rare. Unwitting surgical excision of a functional paraganglioma may precipitate life-threatening complications. We present a case of a 38-year-old woman with a vaginal mass 3.0 cm in diameter who experienced a hypertensive crisis during an unwitting attempted surgical excision of the vaginal mass. The diagnosis of a vaginal functional paraganglioma was then made based on to her 16-year history of paroxysmal headaches, chest distress, palpitation and elevated levels of urinary vannillylmandelic acid (VMA). Consequently, after thorough presurgical preparation, the patient again underwent excision of the vaginal mass uneventfully. She has been followed-up for three years since surgery without any evidence of recurrence. The clinical features and perioperative management of functional vaginal paraganglioma are described.

Key word: Paraganglioma; Vagina; Perioperative management; Hypertension crisis.

Introduction

Paragangliomas are rare neuroendocrine tumors with a highly variable clinical presentation. They are rarely reported along the genitourinary tract: the uterus, ovary, vagina, and cervix. Paragangliomas of the vagina are extremely rare. Only four cases have been reported in the literature [1-4]. Only one of these four cases was identified as a functional one. It has been well documented that surgical excision of an untreated functional paraganglioma may cause a serious and potentially lethal cardiovascular crisis. This poses great diagnostic and management challenges for gynecologists. We report the second case of a patient with a functional vaginal paraganglioma who experienced a hypertension crisis during surgery.

Case Report

A 38-year-old Chinese woman, gravida 2, para 2, was found to have a mass located in the anterior wall of the vagina during routine gynecologic examinations over a 7-year period. However, the most recent gynecologic examination revealed an increase in size of the mass to 3.0 cm in diameter. The patient had disregarded 16 years of paroxysmal headaches, chest distress, and palpitations.

Vaginal mass excision was performed under epidural anesthesia in an outpatient operating room. Standard monitors showed her baseline blood pressure was 110/58 mmHg and baseline heart rate was 70 beats/min. Precisely when the surgeon intraoperatively touched the tumor, the patient's arterial blood pressure and heart rate dramatically increased to 200/120 mmHg and 200-220 beats/min, respectively, accompanied by a severe headache, paleness, polypnea, blurred vision, limb anesthesia and nausea. Immediately, she was given 3 mg of midazolam and 20 mg of furosemide intravenously. Five minutes later, the symptoms disappeared and the operation was continued. However a similar episode occurred again when palpating the mass and an electrocardiogram showed ST-segment depressions in certain leads. The operation was then canceled and the patient was hospitalized for further management.

She recalled her 16-year history of paroxysmal strong headaches, palpitations and chest distress lasting for 3-5 min. Such spells were usually unpredictable but occasionally evoked by sexual activity or heavy exercise. Unfortunately, she had never seen a doctor for these symptoms. Laboratory data revealed elevated urinary vannillylmandelic acid (VMA) (15-18.5 mg/24 hours). Transvaginal ultrasound (Figure 1) showed a low echo-level solid tumor on the anterior wall of the vagina measuring 2.3 x 2.3 x 2.0 cm. Her blood pressure was normal between occasional fluctuations (ranging from 77/45 mmHg to 202/118 mmHg). Paraganglioma of the vagina was diagnosed. After being controlled with alpha-blockade, the tumor was extirpated. During the operation there were several hypertensive episodes which were managed by nitroprusside and phentolamine. Immediately after resection of the tumor, the patient had a severe hypotensive episode (65/40 mmHg) managed by norepinephrine. The postoperative period was hemodynamically stable. The pathohistologic diagnosis was vaginal paraganglioma (Figure 2). The patient has been followed-up for three years since surgery without any evidence of recurrence.

Discussion

Paraganglioma is a rare tumor from extra-adrenal chromaffin tissue or derived from parasympathetic tissue. Paraganglioma may occur in any part of the body where extra-adrenal chromaffin tissue or paraganglionic tissue is present. Parasympathetic-associated paragangliomas have a predilection for the head and neck region, most of which do not produce catecholamines, whereas catecholamine-producing extra-adrenal paragangliomas are usually found in the abdomen [5]. Paragangliomas of the vagina are extremely rare. Only four cases have been reported in the literature up to now and our reported case

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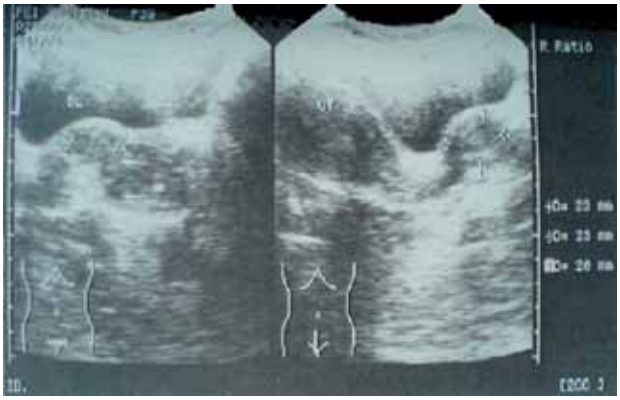

Figure 1. — Transvaginal ultrasound showing a low echo-level solid tumor of the anterior wall of the vagina measuring 2.3 x 2.3 x 2.0 cm.

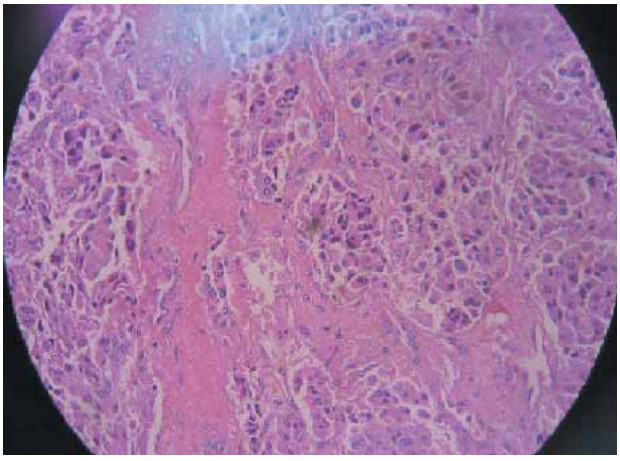

Figure 2. — Tumor cells had a large amount of granular cytoplasm, hyperchromatic round nuclei and formed nests of Zellballen organoid structures separated by a framework of fibrovascular connective tissue with partial hyperplasy of collagen.

is the second case of vaginal paraganglioma that produced catecholamines [1-4].

Clinical presentation of functional paraganglioma can vary greatly, commonly presenting with paroxysmal headaches, sweating, and palpitations while some cases are asymptomatic. Sometimes tumors may be functionally dormant before anesthesia and tumor manipulation during surgery. Among the reported four cases of vaginal paragangliomas, only one case presented with episodes of hypertension during the biopsy and the other three cases had only manifestations of a vaginal mass or vaginal bleeding. In our reported case, the woman had a long-time history of paroxysmal headaches, palpitations and chest distress, first stimulated by her primiparity. Symptoms were usually evoked by menstruation, breath-holding to defecate, and occasionally by sexual activity. Manipulation or palpation of the tumor even evoked a hypertensive crisis.

The potential morbidity and mortality associated with surgery in a patient with an undiagnosed functional paraganglioma is high, and adequate preoperative management is very important, as well reviewed by Lenders *et al*. [6]. Emergency tumor resection without proper preparation results in poor survival. Complications during surgery include hypertensive crises, cardiac arrhythmias, pulmonary edema, cardiac ischemia, and hypotension or even shock, which are all potentially life-threatening. With adequate pretreatment, perioperative mortality has fallen to less than 3%. This emphasises the importance of adequate preoperative management. The major aim of medical pretreatment which often includes the blockade of α-adrenoceptors is to prevent catecholamine-induced episodes and to reduce potentially life-threatening perioperative situations. Our reported case experienced a hypertensive crisis during the first unwitting surgical attempt but the second thoroughly preoperatively prepared surgery was uneventful. In conclusion, the possibility that a tumor is a paraganglioma should be considered before surgically removing a vaginal mass, and emergency methods for the treatment of catecholamine crises should be handled, so that the risks of operating on these tumors can be lessened.

Most paragangliomas are benign and curable by surgical resection, but some are clinically malignant. The differential diagnosis of malignancy with benign lesions can be difficult even with pathology. Our case had a clinical course of 16 years and no evidence of metastases have been found after a postoperative follow-up of three years. Although none of the previous paragangliomas of the vagina were associated with malignant behavior, close follow-up is recommended.

References

[1] 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.

[2] Pezeshkpour G.: "Solitary paraganglioma of the vagina: Report of a case". *Am. J. Obstet. Gynecol*., 1981, *138,* 219.

[3] 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.

[4] Plate W.P.: "Phaeochromo-blastoma of the vagina". *Gynecologia,* 1955, *139,* 35.

[5] Erickson D., Kudva Y.C., Ebersold M.J., Thompson G.B., Grant C.S., van Heerden J.A. *et al*.: "Benign paraganglioma: clinical presentation and treatment outcomes in 236 patients". *J. Clin. Endocrinol. Metab.,* 2001, *86,* 5210.

[6] Lenders J.W., Eisenhofer G., Mannelli M., Pacak K.: "Phaeochromocytoma". *Lancet,* 2005, *366,* 665.

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