Gynecologic Oncology Reports 24 (2018) 99–101

Contents lists available at ScienceDirect

Gynecologic Oncology Reports

journal homepage: www.elsevier.com/locate/gynor

Case report

Clitoral priapism due to distant clitoral metastasis of high-grade serous ovarian carcinoma: A case report and review of the literature

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1. Introduction

Approximately 22,440 new cases of ovarian cancer were diagnosed in 2017, compromising 1.3% of new cancer diagnoses. The overall 5 year-survival rate for ovarian cancer is approximately 46.5%, however, the 5-year survival rate with distant metastasis on presentation decreases to 28.9% (Howlader et al., 1975–2014). Although direct extension within the peritoneal cavity is the most common route of metastasis, distant metastases can occur through lymphatic and hematogenous spread. The most common extra-abdominal metastatic sites are the pleura, lungs, central nervous system, skin, mediastinal lymph nodes, and bones. Skin metastatic lesions are rare, with a reported prevalence ranging from 1.9–5.1% (Cormio et al., 2003a).

Priapism in women is a prolonged, painful erection of the clitoris not related to sexual arousal. It is most commonly caused by an obstruction of the venous outflow tract from the corpora cavernosa either due to impaired relaxation of the alpha-adrenergic receptors or to mass effect. This is most commonly caused by psychotropic drugs that cause alpha blockage, but has also been associated with sickle cell disease and malignancy (Medina, 2002; DiGiorgi et al., 2004). Clitoromegaly is also in the differential diagnosis for women with clitoral swelling, and presents most commonly in the setting of virilization from excess testosterone.

We present a rare case of a patient who presented with clitoral swelling and a pelvic mass. After her primary debulking surgery for metastatic serous ovarian cancer, the patient's clitoral pain and swelling worsened. She was diagnosed with clitoral priapism with ovarian cancer metastatic to the clitoris. We will review her clinical course and treatment, as well as review the literature for management of clitoral priapism in the setting of malignancy.

2. Case report

A 66-year-old female presented with a two-month history of vague abdominal pain as well as a new painful clitoral lesion. During her work up, a CT scan demonstrated a 14 cm pelvic mass and she was scheduled with gynecologic oncology for evaluation. At her initial appointment, the patient's abdominal pain was noted to be acutely worsened. Due to her pain the patient was taken to the operating room the same day for evaluation and debulking. Given her tender clitoromegaly, pre-operative testosterone and sex hormone binding globulin levels were drawn. Other tumor markers were also notable for a Ca 125 of 34.9.

She underwent a total abdominal hysterectomy, bilateral salpingo-oophorectomy, radical tumor debulking with pelvic peritoneal stripping, sigmoid resection, ileocecectomy, and omentectomy. Intraoperatively, she was noted to have a large, friable pelvic mass adherent to bladder, uterus, sigmoid colon, terminal ileum and cecum. No other disease was noted on small bowel, upper abdomen, diaphragm, liver edge or spleen. At the completion of the procedure, all gross tumor had been removed. Due to the extent of her disease, her clitoromegaly was considered to likely be hormonal or due to venous stasis due to tumor burden. Final pathology demonstrated high grade serous carcinoma.

The patient's immediate post-operative course was uncomplicated. One week after surgery, however, she reported worsening clitoral pain and enlargement. Her exam was significant for an enlarged, firm, tender clitoris. Clitoral priapism was suspected, and she was prescribed a trial of oral phenylephrine. Her symptoms improved temporarily.

On POD#26 she presented for her first cycle of adjuvant carboplatin and paclitaxel. Prior to starting treatment that morning, however, she reported exacerbation of her clitoral symptoms as well as bleeding from the area. Chemotherapy was held, and a pelvic MRI was performed which noted a marked abnormality within the perineum with hyperintense bilateral tubular structures, converging at the midline near the clitoris (Fig. 1). Also near the clitoris there was an approximate 2.2 cm abnormality extending bilaterally along the inferior pubic rami and posteriorly to the ischial tuberosities. That afternoon she underwent an exam under anesthesia and was noted to have a massively enlarged clitoris, measuring 3 cm in width, bulging past the clitoral hood. The clitoris was very tense with bleeding at the tip, and this was thought to be due to ischemia. The clitoral tip was very friable and was debrided. With concern for ischemia due to clitoral priapism, phenylephrine (250 μg in 0.3 ml normal saline) was injected into the corpus cavernosum bodies at 3:00 and 9:00 of the clitoris. Within 30 s of the

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https://doi.org/10.1016/j.gore.2018.04.011
Received 24 March 2018; Received in revised form 24 April 2018; Accepted 30 April 2018
Available online 01 May 2018

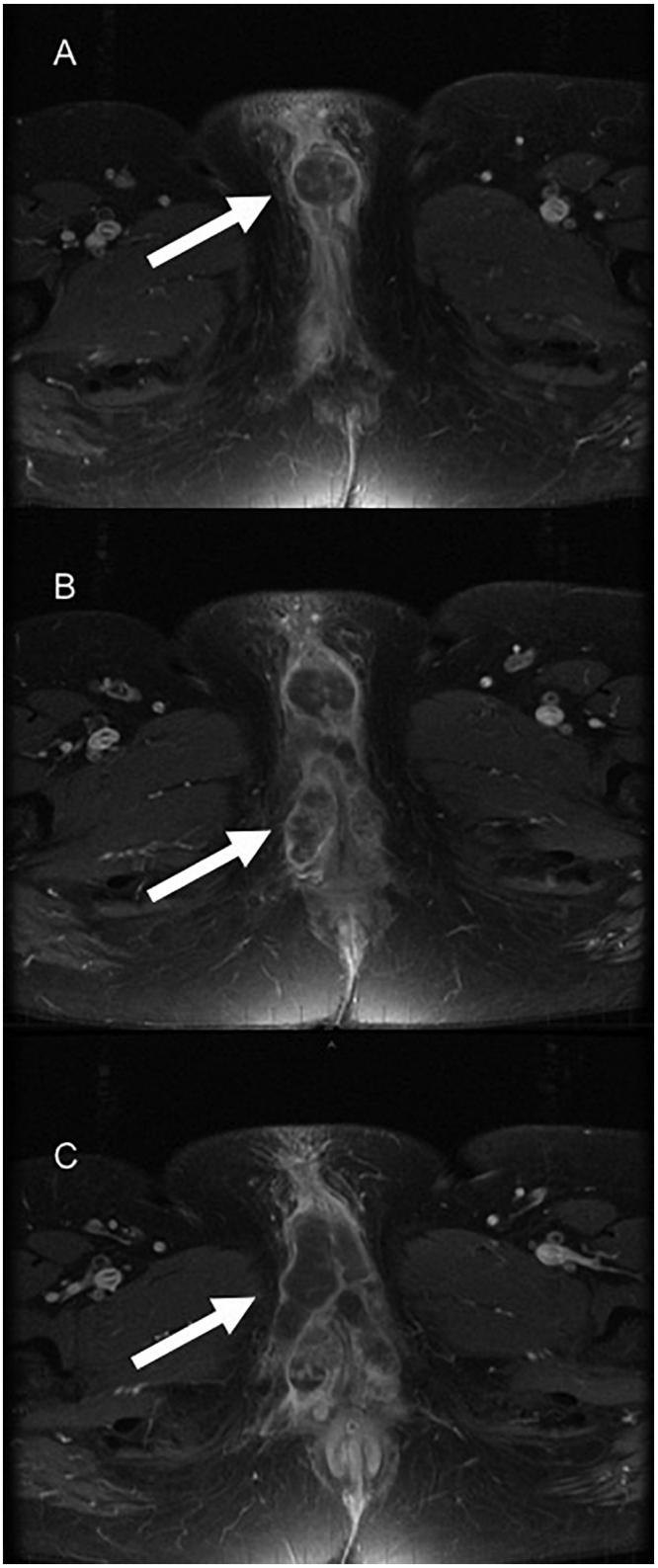

Fig. 1. MRI of the pelvis, T2 axial images, arrows highlighting enlarged clitoris (A), and engorged corpus cavernosum bodies (B and C).

injection, swelling of the clitoris decreased, and the clitoris was noted to retract below the clitoral hood.

On POD#1 she reported marked improvement of her pain and received her first cycle of paclitaxel and carboplatin. Pathology from the clitoral debridement tissue from surgery revealed high-grade serous carcinoma (Fig. 2). Her symptoms resolved during her adjuvant chemotherapy treatment.

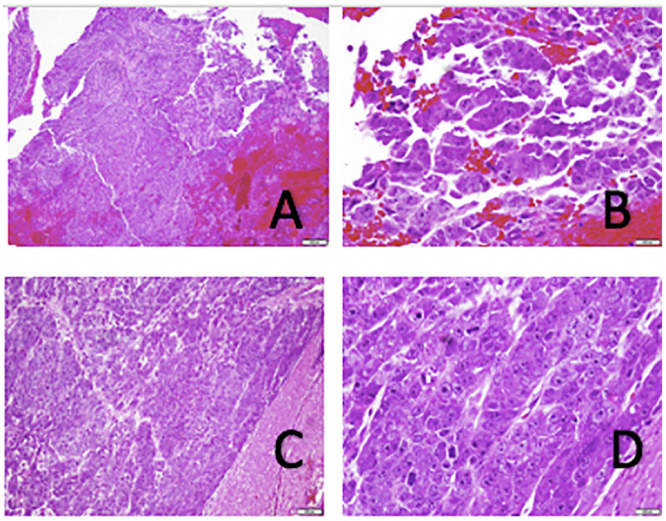

Fig. 2. Images A and B are from the patient's clitoral biopsy and demonstrate high-grade serous carcinoma. Images C and D are from the patient's original debulking demonstrating high-grade serous carcinoma of the ovary.

3. Discussion

This case represents a rare presentation of high-grade serous carcinoma of the ovary presenting with clitoral priapism and a cutaneous metastasis to the clitoris. Clitoromegaly can be a presenting sign in gynecologic malignancies; however, the most common etiology is virilization due to androgen excess. Therefore, if a thorough history and physical exam demonstrate no other signs of androgen excess, clitoral priapism should remain in the differential diagnosis.

As discussed previously, clitoral priapism can be caused by obstruction due to mass effect or medication side effects causing alpha-adrenergic blockade. The majority of cases reporting female priapism are due to psychotropic agents such as trazodone and serotonin reuptake inhibitors. In contrast, there are only a few case reports that describe clitoral priapism due to malignancy (Medina, 2002). Two cases of bladder carcinoma and one case of a locally aggressive granular cell tumor have been reported to cause clitoral priapism (DiGiorgi et al., 2004; Slavin et al., 1986; Monllor et al., 1996). Only two cases associated with a gynecologic malignancy have been reported, one presenting as a clitoral tumor in vulvar lymphoma and another patient with cervical cancer (Ferrando-Marco et al., 1992; Lozano and Castañeda, 1981). Using the search terms “clitoral metastasis”, “clitoral priapism” and “cutaneous metastasis ovarian carcinoma” in PubMed, no cases to date have described high-grade serous carcinoma of the ovary presenting initially as clitoral priapism.

Ovarian cutaneous metastases on initial presentation are rare, and the prognosis of distant metastasis is relatively poor. A retrospective review by Dauplat et al. found that only 3.5% (9 of 255) of patients with ovarian cancer had subcutaneous nodules on initial presentation. These patients had a median survival of only 12 months (Dauplat et al., 1987). A review by Cheng et al. reported 6 cases out of 665 epithelial ovarian carcinoma patients with extra-abdominal metastases, with a median survival time of 11 months (Cheng et al., 2009). A review of 9 cases of skin metastases in ovarian carcinoma by Cormio et al. revealed that the most common sites of skin metastasis included prior surgical sites, neck, chest and arms. This review only reported one case of metastasis to the groin and vulva and did not specifically involve the clitoris (Cormio et al., 2003b).

While any cutaneous metastasis is uncommon, distant clitoral

metastases are extremely rare at initial presentation and, to date, have not been reported in high-grade serous carcinoma of the ovary. Case reports of clitoral metastases include patients with a retroperitoneal leiomyosarcoma as well as bladder, breast, renal, stomach, cervix, uterus and anorectal carcinomas (Cokmert et al., 2014; Julien et al., 2012; Filho et al., 2014). One report by Karpate et al. reports a patient with probable ovarian adenocarcinoma presenting with a recurrence as clitoral and skin nodules (Karpate et al., 2009). The only cases involving gynecologic malignancies are patients with cervical and uterine cancers; to our knowledge there have been no reported cases of clitoral metastasis on primary presentation of ovarian cancer or metastasis due to high-grade serous carcinoma of the ovary.

In summary, our patient's presentation is the first reported case to date of high-grade serous carcinoma of the ovary presenting with a clitoral metastasis causing clitoral priapism. This metastatic site likely caused blockage of the venous drainage system in her clitoris causing severe pain and swelling. Ultimately, her symptoms were relieved with intracavernosal injection of phenylephrine and initiation of chemotherapy. Our case demonstrates that in the absence of other signs and symptoms of virilization, clitoral priapism should be further investigated and may represent a distant metastatic site of malignancy.

4. Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflicts of interest statement

The authors declare that there are no conflicts of interest associated with this paper.

Financial support

None.

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