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Primary Vaginal Adenosarcoma: Report of a Case and Review of the Literature

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Vaginal tumour; Adenosarcoma; Endometriosis

Abbreviations:

WT1: Wilms tumor protein; ER: estrogen receptor; PR: progesterone receptor; CD: cluster designation or cluster of differentiation; MDT: multi-disciplinary team meeting

1. Abstract

1.1. Background: Adenosarcoma is a rare biphasic neoplasm of the female genital tract wich is composed of a benign epithelial component and a malignant mesenchymal component. While several cases of uterine adenosarcoma have been reported in the literature, there are only a few reports of vaginal adenosarcoma . This is, to our knowledge, the 8th published case report of primary vaginal adenosarcoma.

1.2. Case Presentation: In this case report, we present an unusual case of a primary vaginal adenosarcoma that developed in 43 years old patient without a clinical or radiological sign of endometriosis.

1.3. Conclusion: In this case we aim to describe the clinical, histological and immunohistochemical features of this exceptionnel tumour, and to discuss the different therapeutic management opted for our patient and the other ones reported in the literature .

2. Introduction

In 1974, Clement and Scully were the first to describe uterine adenosarcoma as mixed tumors with benign Mullerian epithelial and malignant mesenchymal components [1]. Since then, several cases of adenosarcomas of the female genital system have been reported. Most of these cases were uterine, but other, rarer sites have been described, such as the cervix, ovary, fallopian tube, vagina and peritoneum.

3. Case Report

We report the case of a 34 years old, nulligest and nulliparous patient who was referred to our structure for vaginal pain with a prolapse mass appeard 8 weeks ago. She did not have any medico-surgical or gynecologist-obstetric history. Gynecological examination under general anesthesia showed a mass developed on the right side of the vagina wall and away from cervix. The rest of the exam was normal.

Magnetic Resonance Imaging (MRI) of the Pelvis showed a 29x93x70 mm vaginal mass infiltrating the vaginal wall. On T2-weighted images , the mass showed a low signal intensity with intense enhancement after injection of gadolinium (figure 1).

Mass excision was performed and the specimen was sent to pathology. The macroscopic examination showed a polypoid mass measuring 10x6x3 cm in size. The cut surface showed cystic and solid areas. Histopathologic examination revealed a biphasic tumor composed of benign endometrial glands surrounded by hypercellular stroma with leaf-like growth patterns. The stromal cells showed mild to moderate atypia with periglandular stromal condensation. Four mitotic figures per 10 high-power fields were observed.

Immunohistochemical stainings showed positivity for cytokeratins and hormonal receptors in the glandular epithelium. In the mesenchymal component, tumour cells showed positive labelling with vimentin, WT1, CD10 and hormonal receptors. Tumour cells were negative for myogenine, and CD 34.

These pathology results were consistent with the diagnosis of a low-grade adenosarcoma. The resection margins were tumor-free (Figure2).

The patient underwent Computerized Tomography (CT) scans of the thorax, abdomen and pelvis that revealed no metastases. After the MDT discussion, the patient did not benefit from additional surgery (given her particular context) and was referred to oncology for chemotherapy. 9 months later, our patient is still free of disease.

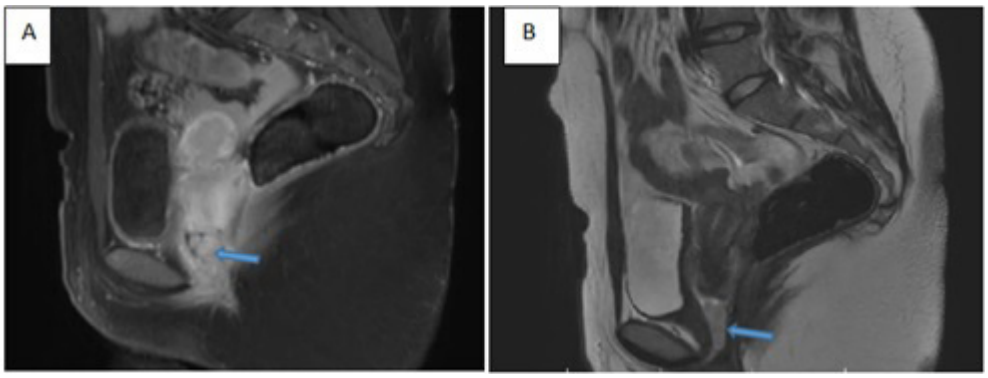

Figure 1 : Magnetic resonance imaging showing a mass infiltrating the vaginal wall on sagittal T2-weighted images (A) and strongly enhanced after injection of gadolinium on sagittal T1-weighted images (B).

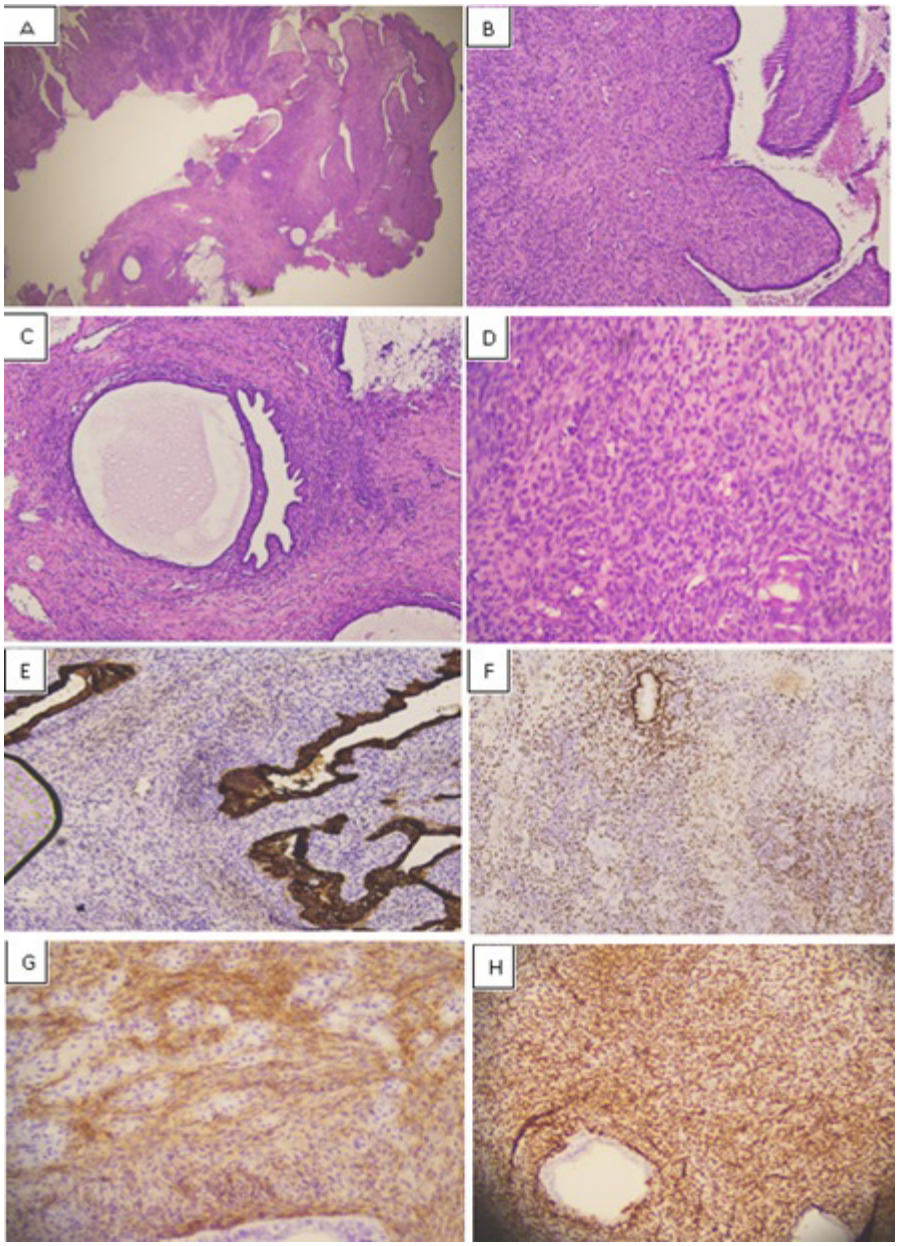

Figure 2 : Low grade adenosarcoma of the vaginal wall : (A-B) low power reveals a biphasic tumor with phyllodes-like architecture. (hematoxylin & eosin _20 et _100). (C) the epithelial component characterized by endometrioid gland surrounded by dense stromal cells (hematoxylin & eosin _100). (D) The stromal cells have atypical nuclei. Some mitotic figures are observed. (hematoxylin & eosin _400).
(E) Immunohistochemical stainings showed positivity for cytokeratins in the glandular epithelium(x100) (F) and for hormonal receptors in both component (x100). (G) CD10 stain showing mainly membranous pattern in stromal cells (x200) (H) Diffuse Nuclear WT1 staining in stromal cells (x100).

4. Discussion

Adenosarcoma is a rare mixed neoplasm of the female genital system consisting of a benign epithelial component and a malignant stromal component [1]. Several cases were subsequently published, the majority of these series of studies report uterine localization of adenosarcoma [2]. However, these tumors can occur in other gynecologic tissue such as ovaries, cervix, fallopian tube, vagina and peritoneum [3].

Primary vaginal adenosarcoma is extremely rare. Research of English language reports using the keywords ‘‘vaginal adenosarcoma,’ was performed. Articles were further considered after reviewing the titles and abstracts when available. This research revealed only 7 cases of primary vaginal adenosarcoma (Table 1) [9-15].

The review of these cases found that the age distribution ranged from 34 years to 58 years with a mean age of 47 years. All patients have been presenting a history of endometriosis. Unlike our patient that presented no clinical or radiological sign of endometriosis.

The etiology of vaginal adenosarcoma is unknown. These neoplasms may arise in association with endometriosis.

Endometriosis is defined as the presence of endometrial tissue outside of the endometrium and myometrium. It can be considered to be a disease with the potential to develop malignancy [5], especially if related to immune response alterations [6]. Clear cell and endometrioid neoplasms are the most common tumours arising in endometriosis. More rarely, other associated neoplasms including adenosarcoma were reported [5]. However, Some extrauterine tumors have not been associated with endometriosis, they may have arisen from the pluripotent mesothelial and mesenchymal cells in the pelvic cavity [7].

Patients usually present with symptoms of a vaginal mass, including pain, vaginal bleeding and urinary frequency or incontinence.

Histologically, low power reveals a biphasic tumor with phyllodes-like architecture composed of benign glands surrounded by hypercellular stroma. Depending on stromal cytologic atypia, we distinguish low and high-grade adenosarcomas [4].

Immunohistochemically, the glandular component shows positive staining for pancytokeratin (AE1/AE3) , ER ,PR, and may be positive for CD10 but negative for androgen receptors. However the mesenchymal component expresses ER, PR, and CD10. Smooth Muscle Actin (SMA), desmin, CD34 and cytokeratins can have variable positivity in the stromal component [8].

The main differential diagnoses of adenosarcomas are adenomyoma, endometrial polyps and carcinosarcoma [8].

Treatment of vaginal adenosarcoma is not yet standardized. Among the 7 patients reported in the literature, the treatment in 5 patients was mainly based on complete surgical resection of the tumor with adjuvant radiation or chemotherapy therapies. The treatment of the other two patients was based solely on surgery without adjuvant radiotherapy or chemotherapy, although one of the two presented sarcomatous overgrowth(without infiltration of the surgical margins). Neither of these two patients died or had recurrence at the time of their reports.

Few data are available concerning the prognostic factors. In general, vaginal adenosarcomas are presumed to recur or metastasize. According to the literature, two-thirds of patients presented with recurrences before they became free of disease. One patient presented pulmonary metastases and died.

Table1: Summary of published reports of primary vaginal adenosarcoma in the literature

	Author	Year	Country	Age year	Size cm	Stromal mitotic figures (/10 HF)	Treatment	Metastasis	Evolution	History of endo metriosis	Previous surgery	Medical therapy	Interval between hysterectomy and diagnosis of adenosarcoma
1	**Judson, et al Judson, et alnnn Judson, et al.**	1999	USA	42	7	3 mitoses	6 courses of PC and external beam and brachyradiotherapy	NO	Regular and free of disease	yes	Hysterectomy and bilateral salpingo-oophorectomy	Progestins	2 years
2	**Anderson, et al.**	2001	USA	46	10	4 mitoses	Extetnal radiotherapy and interstitial brachytherapy	NO	Regular and free of disease	yes	Hysterectomy and bilateral salpingo-oophorectomy	Oestrogen/ Progestin	23 mounths
3	**Liu et al.**	2003	*USA*	56	16	Occasional mitoses	PI chemotherapy and pelvic adiotherapy	NO	Regular and free of disease	yes	Hysterectomy and bilateral salpingo-oophorectomy	Oestrogen/ Progestin	22 mounths
4	**Toyoshima, et al**	2004	Japan	52	11	Associated with sarcomatous overgrowth	3 ourses of CP and 1 course of PC	Both Lungs	Died from disseminated disease 9 months after surgery	yes	Hysterectomy and bilateral salpingo-oophorectomy	No	10 years
5	**Han, et al.**	2010	China	34	7	Rare (low-grade neoplasm)	3 courses of PVP and 1 course of PEI	NO	Regular and free of disease	yes	Hysterectomy and bilateral salpingo-oophorectomy	No	6 mounths

6	**Pontrelli**	2016	italy	58	3	With sarcomatous overgrowth 9Mitoses	No chemotherapy and radiotherapy	NO	Regular and free of disease	yes	total hysterectomy and right salpingectomy	not mentioned	19 years
7	**Li and Duan, et al.**	2019	china	43	4	Low grade 1-2 mitoses	No chemotherapy and radiotherapy	NO	Regular and free of disease	yes	Hysterectomy and bilateral salpingo-oophorectomy	Oestrogen/ Progestin	10 years

PC: Paclitaxel and Carboplatin; PI: Cisplatin and Ifosfamide; CP: Carboplatin and Pirarubicin; PVP: Cisplatin, Vincristine and Pingyangmycin; PEI: Cisplatin, Epirubicin and Iphosphamide.

5. Conclusion

Vaginal adenosarcoma is an extremely rare biphasic neoplasm that is most often linked to endometriosis. The diagnosis is based on histological study because of non-specific clinical and radiological features. Treatment and prognostic factors have not been established yet.

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