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CASE REPORT

Adenoid Cystic Carcinoma of the Vagina: A Case Report

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ABSTRACT

Adenoid cystic carcinoma (ACC) of the vagina arising from Bartholin's glands is a rare type of female malignancy. ACC of the vulva is rare, and treatment protocols are often based on experience with similar tumours in other locations (e.g. head and neck) and case reports. Here, we report a case of a 52-year-old female patient who presented with a pain above perineum and lower end of vagina leading to a diagnosis of ACC of the vagina after biopsy and confirmation on immunohistochemical analysis. Often, such lesion is clinically misdiagnosed as a cyst or inflammation. The present case was clinically suscpected to be either a bartholin cyst or lipoma or parasitic fibroid or calcification. The possibility of cancer should be considered in any female older than 40 years of age with a lesion near the Bartholin's glands.

KEYWORDS

• Adenoid Cystic Carcinoma • Bartholin's Glands • Bartholin's Cyst • Immunohistochemistry

INTRODUCTION

Adenoid cystic carcinoma notably targets the salivary glands, breasts, and female genital tract, especially the cervix.

Carcinoma of Bartholin's glands has been variously estimated as accounting for between 2 to 7% of malignant vulvar neoplasms. If adenoid cystic carcinomas are excluded then Squamous cell carcinomas and adenocarcinomas, in approximately equal proportions, make up about 85% of Bartholin's glands carcinomas, the remainder being transitional carcinomas, adenosquamous carcinomas, adenoid cystic carcinoma and anaplastic carcinomas.[1]

ACC comprise about 10% of malignant Bartholin's glands tumours and are distinctive

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in both their histological appearance and their behaviour.[1]

To date, there are only a few cases documented in the literature.

Bartholin's glands carcinoma most commonly presents as a vulvar mass with pain or irritation and occasionally accompanied by vaginal discharge and bleeding. In very high proportion of cases the initial clinical diagnosis is of a cyst or abscess.[1]

The general approach for vulvar cancer involves surgery as the primary treatment, with lymph node status being a major determinant of further treatment. For ACC, surgical resection with clear margins is crucial, and adjuvant radiation therapy may be considered, especially if margins are positive or perineural invasion is present, according to the International Journal of Gynaecological Cancer and National Institutes of Health (NIH) | (.gov).[2]

CASE REPORT

A 52-year-old female patient, presented with pain above perineum and lower end of vagina. On examination, one 1x1 cm size cyst was felt at lower end of vagina at 7 o'clock position. It was tender on touch. There was no vaginal discharge or bleeding.

Incision and excision biopsy was performed for final diagnosis.

Gross examination: Received 4 soft tissue fragments measuring 0.3 to 1 cm in size. One is divided and all processed.

Microscopic examination: Sections show histological features of adenoid cystic carcinoma. Perineural invasion is present. (Figure 1-3)

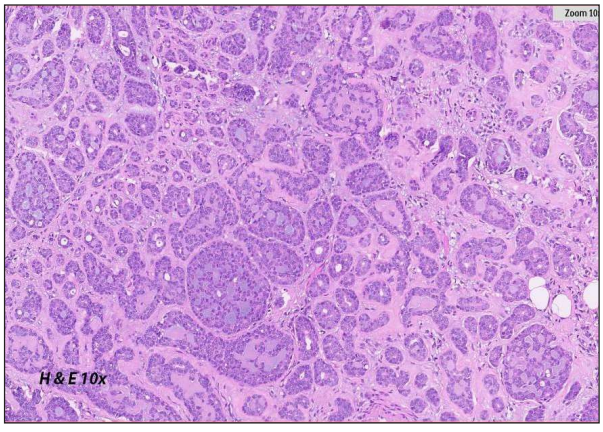

Figure 1:

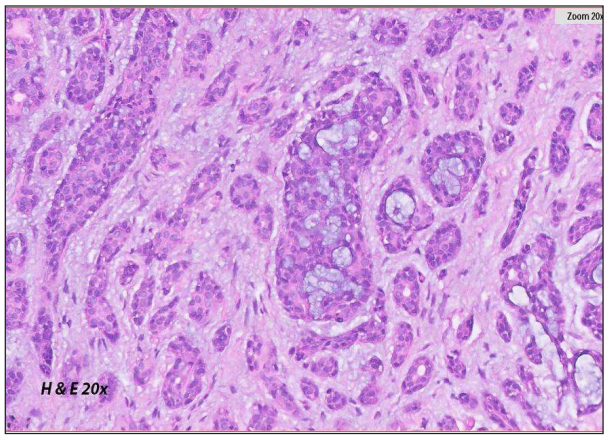

Figure 2:

Figure 1 & 2 Adenoid cystic carcinoma with tubular and cribriform pattern. Tubular pattern contains simple tubules composed of inner ductal and outer myoepithelial cells. Cribriform pattern is composed of myoepithelial cells with myxoid or hyaline globules.

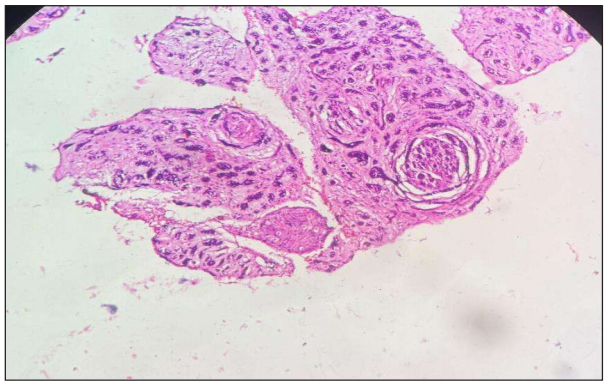

Figure 3: Adenoid cystic carcinoma with perineural invasion

Further immunohistochemical analysis showed that tumor cells expressed p63 and CD 117 positivity with patchy positivity for p16. Ki-67 showed low index. ER and CEA are negative. (Figure 4-8)

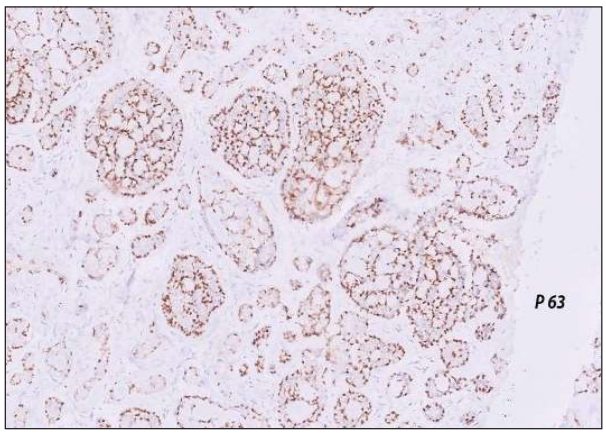

Figure 4: p63 Positivity

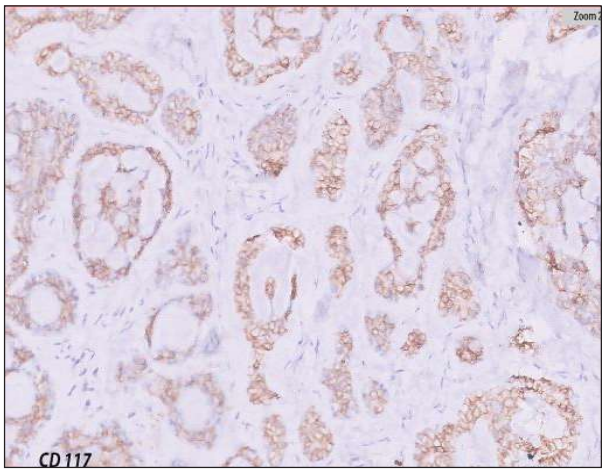

Figure 5: CD117 Positivity

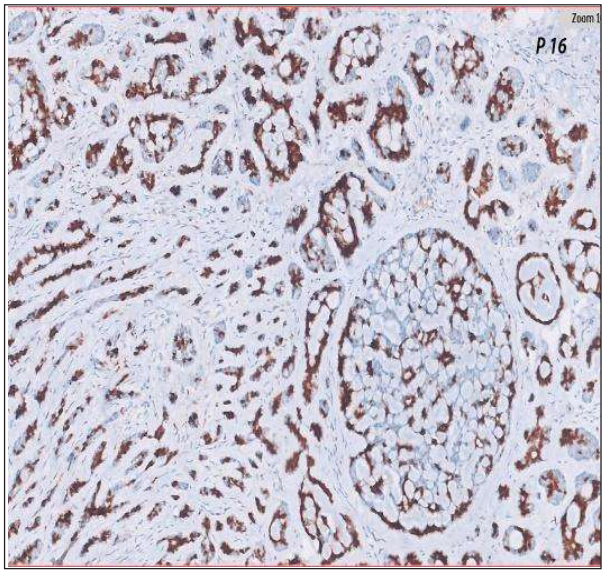

Figure 6: Patchy positivity for p16

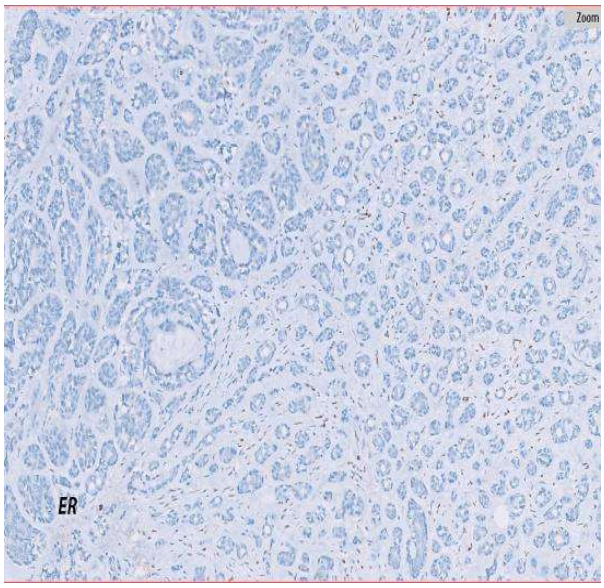

Figure 7: ER Negative

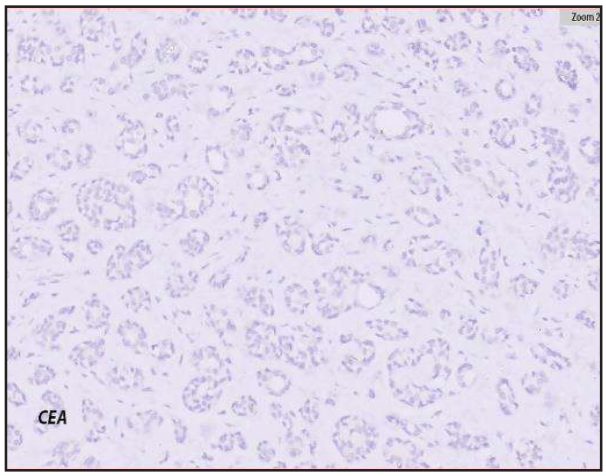

Figure 8: CEA Negative

Radiology investigations: MRI Pelvis contrast was carried out after HP examination report.

Observation: Evidence of poorly defined nodular soft tissue thickening is seen along the right side of vulval soft tissue and extending into the overlying perineal subcutaneous fat pad. It is seen infiltrating the anterior margin of the vulval soft tissue on right side at the vaginal introitus. surrounding perilesional soft tissue edema noted. The lesion exhibits intermediate signal on T2W images with accentuated heterogenous thick peripheral wall enhancement and reveals patchy restriction of diffusion. Rest of the vaginal vault appears normal. No deeper extension is seen into the rest of the vaginal and cervix. No bony involvement is noted. The lesion approx. measures 1.9 x 1.8 x 2.1 cm.

Few small volume bilateral inguinal and external iliac lymph nodes noted indeterminate. Uterus is post-menopausal atrophic in size and shows normal signal intensity. No obvious focal lesion is seen. Endometrium is linear and regular. Junctional zone is preserved. Both adnexa appear unremarkable.

DISCUSSION

Adenoid Cystic Carcinoma was initially mentioned by Billroth as a histological variant of adenocarcinoma occurring in glandular mucosae.[3,4] It is most commonly seen in the salivary gland, breast, and skin.[5] In the female genital tract, the cervix is the most common site of ACC but is rare in Bartholin's gland.[5,6]

These tumours are distinctive in both their histological appearance and their behaviour.

The tumour is consisting of small, angular or oval cells with oval hyperchromatic nuclei and scanty amount of cytoplasm, which resemble the luminal cells of the ductal epithelium, and modified myoepithelial cells which have large angular nuclei without nucleoli. The cells form single or anastomosing strands, trabeculae and well-circumscribed nests with a cribriform growth pattern within which there are lumina and pseudolumina containing basophilic, PAS-positive after diastase digestion, amorphous material. It is a combination of reduplicated basal lamina material and proteoglycans (Orenstein *et al,* 1985). The cell nests are set in a fibrous stroma.[1]

The histogenesis of an adenoid cystic carcinoma in this site is not certain; Addison & Parker (1977) suggested an origin from either ductal epithelium or from vascular myo-epithelial cells whilst a more recent study has suggested that they are derived from reserve cells in the intercalated small ducts of the gland that have the capacity to differentiate into luminal epithelial cells or into myoepithelial cells (Tsukahara *et al,* 1991). Cytogenetic study of one neoplasm showed complex chromosomal changes involving chromosomes 1, 4, 6, 11, 22 and 14 (Kieche-Schwartz *et al,* 1992) but whether this is a consistent pattern of abnormalities in these tumours remains to be determined.[1]

These tumours mainly invades and spreads via the perineural spaces and is associated with a high incidence of local recurrence (Sayre, 1949; Dennis *et al,* 1955; Murphy *et al,* 1962; Abell, 1963; Eichner, 1963; Dodson *et al,* 1978; Ortner et al, 1980; Bernstein *et al,* 1983b; Copeland *et al,* 1986a; Rosenberg *et al,* 1989).[1]

The tumour follows, an indolent and prolonged course with the mean time interval between primary treatment and recurrence being about eight years (Copeland *et al,* 1986a; Lelle *et al,* 1994). Spread to regional lymph nodes does occur but is uncommon, at the same time eventual haematogenous spread is to the lung, skeleton and liver (Abroa *et al,* 1985; Chapman *et al,* 1985; Copeland *et al,* 1986a; Rose *et al,* 1991), patients sometimes succumbing to their disease as long as 27 or 31 years after initial treatment (Sayre, 1949; Lelle *et al,* 1994). The five-and 10-year survival rates have been reported as 71 and 59% respectively (Copeland *et al,* 1986a) but in a recent series of 11 patients all three deaths occurred more than 10 years after initial surgical treatment (Lelle *et al,* 1994).[1]

General NCCN Guidelines for Vulvar Cancer (including ACC)

Surgical resection is the primary treatment for early-stage ACC, with lymph node management being crucial. Adjuvant radiation therapy and/or chemotherapy may be recommended based on factors like lymph node status and surgical margins.

Surgery:

Radical wide local excision is the standard for early-stage vulvar cancers, including ACC.

Lymph Node Management:

The status of groin lymph nodes is a critical factor in determining the need for further treatment.

Adjuvant Therapy:

Radiation therapy (RT) and/or chemotherapy may be used after surgery, especially if lymph nodes are positive or margins are close or positive.

Advanced or Recurrent Disease:

For unrespectable or recurrent ACC, chemo radiation is often recommended, with surgery considered for persistent disease.

Considerations for ACC:

- **Perineural Invasion:** A characteristic feature of ACC, which may influence treatment decisions.
- **Positive Margins:** Adjuvant radiation therapy is often recommended for positive or close surgical margins.
- **Clinical Trials:** Participation in clinical trials is encouraged for ACC, as outcomes data is still evolving.[7-17]

CONCLUSION

Role of pathologist is central for diagnosis of this rare entity in this location as it it can be misdiagnosed clinically as a benign lesion. The pathologist can also guide the clinician regarding adequacy of margins, perineural invasion and lymph node status. Given the rarity and complexity of ACC, a multidisciplinary team approach is essential, involving gynaecologic oncologists, radiation oncologists, and possibly other specialists.

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