Primary intestinal-type adenocarcinoma of the vagina: clinicoradiologic-pathologic correlation and a stepwise approach to avoid metastatic overinterpretation

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Background

Primary intestinal-type adenocarcinoma of the vagina (PIAV) is an exceptionally rare, HPV-independent, non-Müllerian malignancy that morphologically and immunophenotypically recapitulates colorectal adenocarcinoma (CRA) and may occur along a spectrum from benign epithelium, adenoma to invasive carcinoma, analogous to colorectal tumorigenesis. We report a case illustrating this pitfall and propose a practical diagnostic workup.

Clinical findings

- 46-year-old.
- Vaginal bleeding and a large exophytic lesion (figure 1).
- Imaging demonstrated disease restricted to the vaginal topography (figure 2).
- Colonoscopy was negative.
- Histopathological and immunohistochemical (IHC) findings showed below (figures 3 and 4)

Figure 1. The physical examination reveals an exophytic and ulcerated lesion involving the vaginal introitus and posterior vaginal wall.

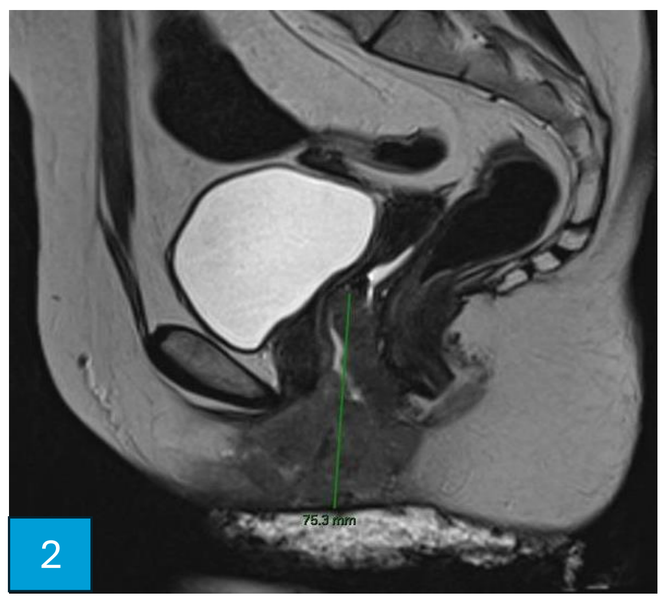
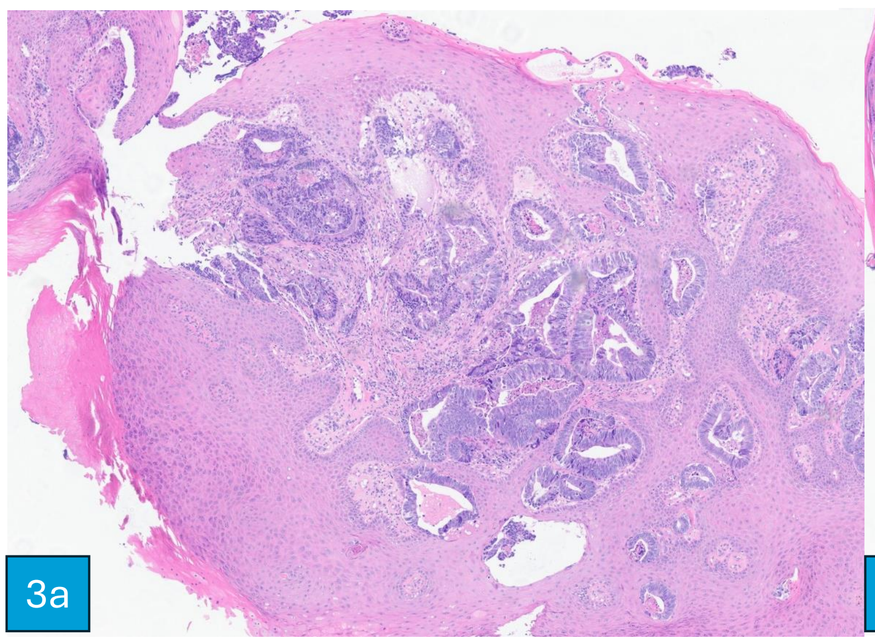
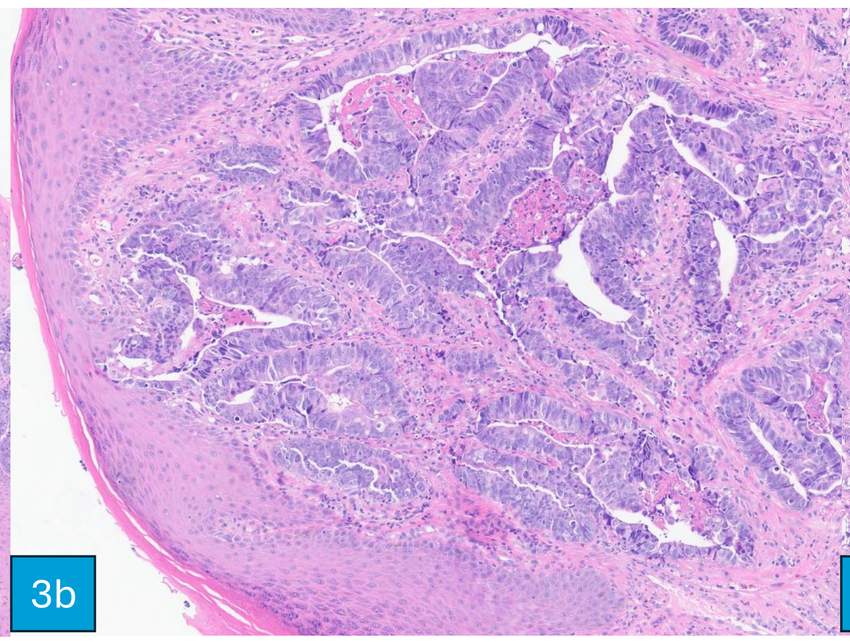


Figure 2. Lesion extending to the lower and middle thirds of the vagina, measuring 7.5 cm

Histologic and immunohistochemical images





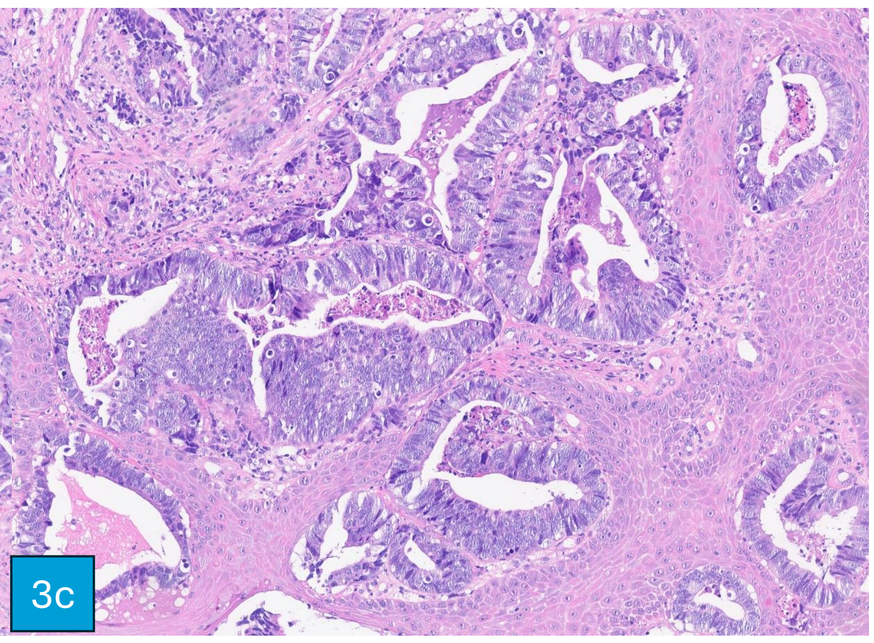
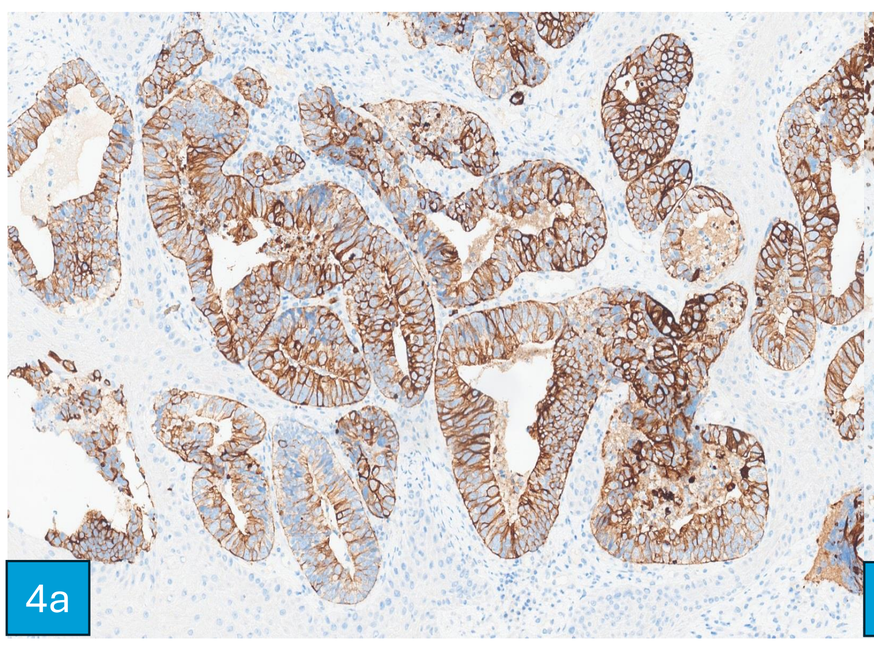
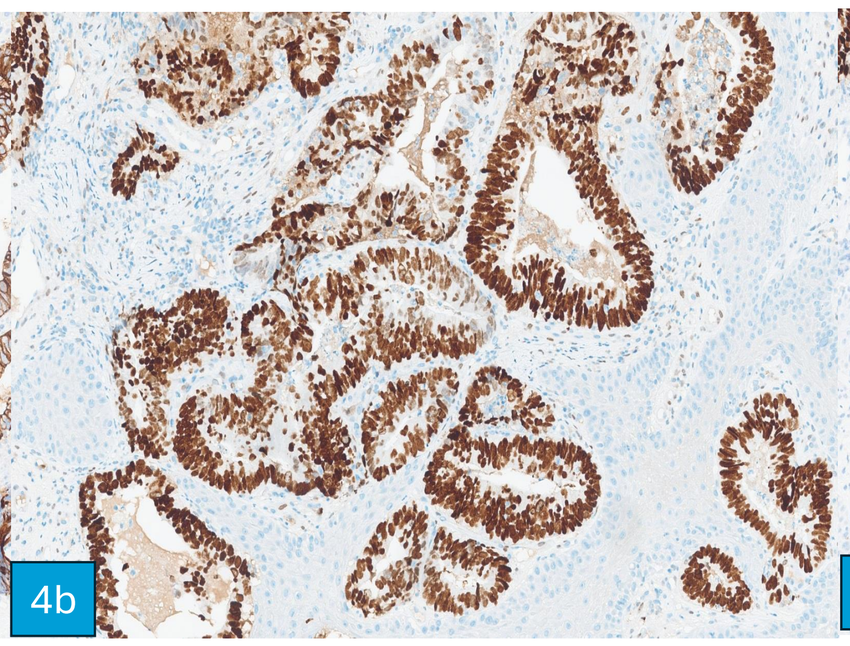


Figure 3a, 3b and 3c. Adenocarcinoma with intestinal differentiation and dirty luminal necrosis.





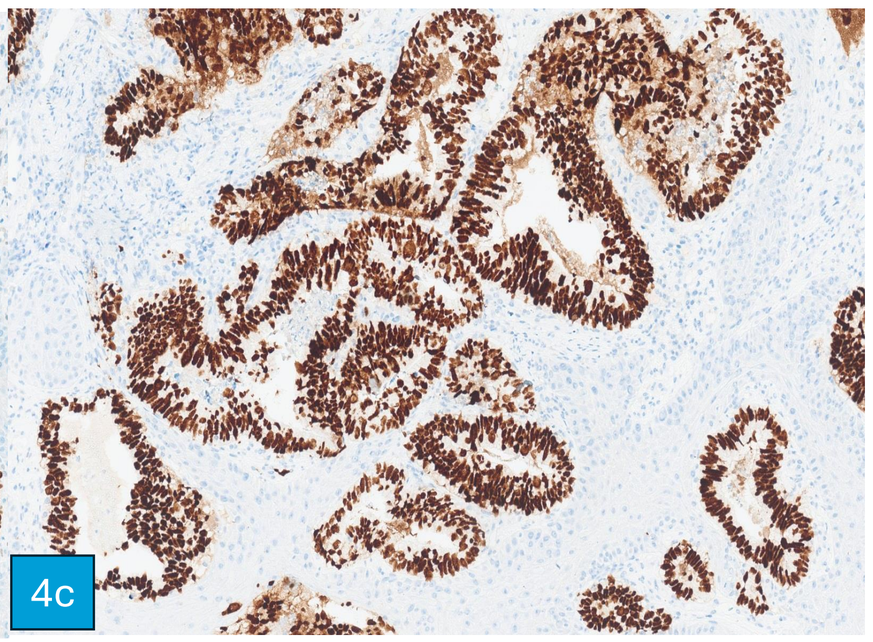


Figure 4a, 4b and 4c. Immunophenotype supported colorectal-like profile (CK20+, CDX2+, SATB2+ - figures 4a, 4b and 4c, respectively), absence of Müllerian markers (PAX8−, CK7−) and non-block-type p16.

Discussion

- Primary adenocarcinoma of the vagina is a rare entity (up to 2% of gynecologic malignancies)[1].
- Intestinal type is limited to case series[1].
- Features similar or identical to those seen in colorectal carcinomas[1].
- Histogenesis remains uncertain: origin from congenital remnants of intestinal epithelium (e.g., cloacal remnants), metaplasia with neoplastic progression (adenoma–carcinoma sequence, including acquisition of mutations such as KRAS and TP53), and, occasionally, secondary development in the setting of a rectovaginal fistula[2].
- Prognosis remains uncertain regarding long-term survival - reported cases of local recurrence and metastasis[3].

Conclusion

PIAV remains an exceptionally rare but increasingly recognized mimic of metastatic CRA. We propose a stepwise diagnostic checklist to prevent inappropriate management:

Recognize colorectal-type morphology → Confirm intestinal differentiation by IHC → Exclude Müllerian lineage and HPV association → Clinicoradiologic and endoscopic correlation

References