Dedifferentiated Endometrial Carcinoma with Synchronous Uterine Leiomyosarcoma: A Rare Collision Tumour – Case Report and Literature Review

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Abstract

Dedifferentiated endometrial carcinoma (DEC) is a rare, aggressive uterine malignancy. Synchronous occurrence with uterine leiomyosarcoma (collision tumour) is extraordinarily uncommon. We report a postmenopausal patient whose hysterectomy demonstrated DEC with subclonal MMR deficiency and a distinct leiomyosarcoma. Integrated morphology, immunohistochemistry (IHC) and targeted molecular testing confirmed the diagnoses.

Introduction

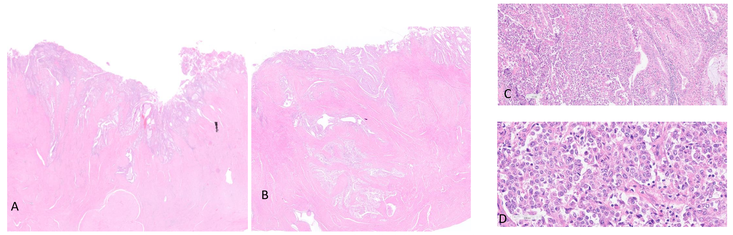
DEC comprises juxtaposed low-grade endometrioid adenocarcinoma and undifferentiated carcinoma and behaves aggressively. Leiomyosarcoma is the most common malignant mesenchymal uterine tumour with poor prognosis. Collision of DEC with leiomyosarcoma appears exceedingly rare and diagnostically challenging.

Case Presentation

Postmenopausal woman with abnormal bleeding underwent TAH-BSO. Uterus 120×80×51 mm; intracavitary endometrial mass 65 mm (6 mm from serosa) and separate intramural mass 43 mm; ovaries/tubes unremarkable except right hematosalpinx.

Histopathology

Endometrium: FIGO grade 1 endometrioid carcinoma with >50% myometrial invasion and abrupt transition to undifferentiated carcinoma; extensive lymphovascular invasion (>5 vessels), cervical stromal involvement, and tumour at lateral cervical margin. Intramural mass: moderate atypia, patchy coagulative tumour cell necrosis, 7/10 HPF mitoses → leiomyosarcoma.



Sections from the endometrial tumour showed areas of well-differentiated endometrioid adenocarcinoma (FIGO grade 1) with more than 50% myometrial invasion (Fig 1 A and B), with an abrupt transition to undifferentiated carcinoma composed of solid sheets of monomorphic tumour cells displaying vesicular nuclei, high mitotic activity, and scant cytoplasm (Fig 1 C and D).

Immunochemistry & Molecular Findings

Differentiated component: AE1/3+, PAX8+, ER/PR+, p53 wild-type, weak p16+. Undifferentiated component: EMA+, PAX8+, CD56+, p16+, focal ER/PR+, AE1/3−, CK7−, p53 wild-type. Leiomyosarcoma: Desmin+, SMA+, CD10−, Ki-67 ~4%. MMR IHC: MLH1/PMS2 loss in undifferentiated areas (subclonal deficiency).

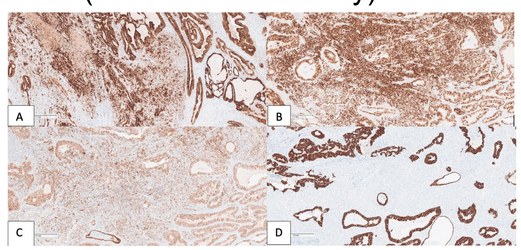


Figure 3: A) EMA +; B) PAX-8 +; C) P16 Focal +; D) AE1/3 - (IHC; x40)

Discussion

DEC is often under-recognized; subclonal MMR deficiency reflects heterogeneity/evolution and has therapeutic implications. MMR-deficient tumours may benefit from immune checkpoint inhibitors in advanced/recurrent settings. Recognition of synchronous leiomyosarcoma alters prognosis and may influence adjuvant strategies.

Conclusion

This rare collision tumour (DEC with subclonal MMR deficiency plus leiomyosarcoma) underscores the need for thorough sampling, broad IHC and selective molecular testing to guide diagnosis, prognostication, genetic counselling and therapy selection.

Key References

References (key): 1) Silva EG et al. Undifferentiated carcinoma of the endometrium. Int J Gynecol Pathol. 2006;25(1):1–10. • 2) Ramalingam P. Undiff & dediff endometrial carcinomas. Adv Anat Pathol. 2016;23(4):226–234. • 3) Singh R et al. Dedifferentiated EC: clinicopath & molecular. Front Oncol. 2022;12:812290. • 4) Han G et al. DEC series of 17 cases. Histopathology. 2020;77(6):866–875. • 5) Makker V et al. Lenvatinib + pembrolizumab in EC. Lancet Oncol. 2020;21(5):719–729. • 6) Le DT et al. dMMR predicts response to PD-1. Science. 2017;357(6349):409–413. • 7) WHO Classification of Female Genital Tumours. IARC, 5th ed. 2020. • 8) Baiocchi G et al. Uterine collision tumour: EC + LMS. Clinics (São Paulo). 2016;71(2):92–95.