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CORE AND MANAGEMENT OF UTERINE PECOMA

- REVIEW FROM A CASE-REPORT -

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OBJECTIVE

Perivascular epithelioid cell tumours (PEComas) are unusual mesenchymal neoplasms with distinctive histological and immunohisto-chemical characteristics (reactivity to myogenic and melanocytic differentiation biomarkers). PEComas can appear on gastric, intestinal, pulmonary and genitourinary conjunctive tissue. Although they are mainly benign, some malignant types have been identified and are still being studied and classified. Uterine PEComas represent < 25% of the total, finding less than 70 cases reported in the literature. Their differential diagnosis includes leiomyomas, angiomyolipomas, extrapulmonary clear cells tumours, myomelanocytic tumours of the broad ligament and other abdomino-pelvic sarcomas. Moreover, it is advisable to dismiss coexisting tuberous sclerosis because of its frequent association. The primary treatment of PEComas is not standarized yet but is normally based on surgical resection, saving adjuvancy to the high-risk cases.

Our aim is to review the bibliographic evidence about the nature, treatment and prognostic features of PEComas in the female genital tract in order to optimize its management; while providing a new case to the scientific literature.

METHODS – Case presentation

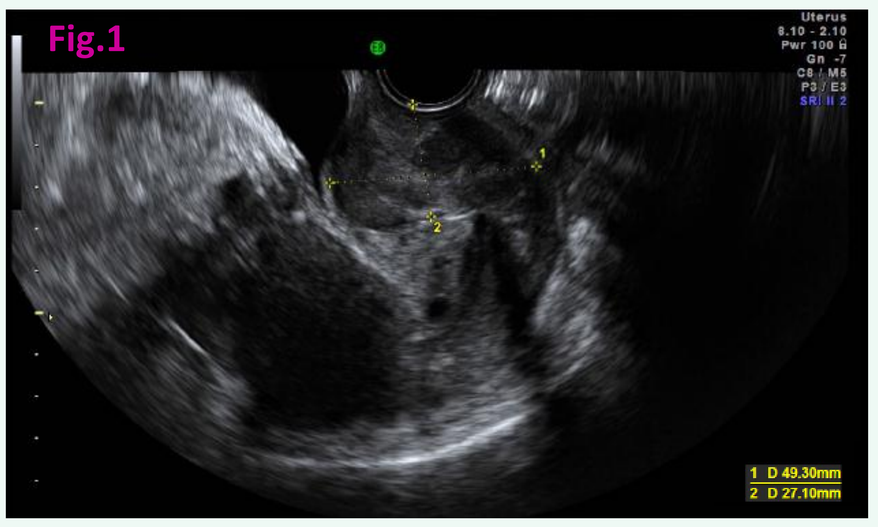
Nulliparous 38-year-old patient, without medical or surgical relevant history, turned to the Gynecologic consultancy recounting self-finding of a genital mass in dyspareunia, coitorrhage and intermittent metrorrhagia clinical context from 1 year. Speculoscopy showed an intravaginal tumour suggesting myoma calved, difficult to delimit. By manually touch, a hard yet elastic mass about 6 cm probably pedunculated from the anterior cervical lip could be palpated. **In the ultrasound stood out a pedunculated dense vaginal nodule of 49x27mm, apparently myoma calved (Fig.1)**. An office biopsy was performed, resulting compatible with granulation tissue.

A diagnostic hysteroscopy was indicated, in which a myomatous-like mass was observed ocuppying the whole vagina, seemingly pendant from the cervical anterior lip. Endocervical canal and endometrial cavity showed no macroscopic pathology. As long as the mass was liberated, its pedunculus could be objectified coming out from the vaginal lateral wall. Tissue sampling was sent for hystological study.

FINDINGS	
Anatomopathological results:	PEComa
Immunohystochemical study:	Tumoral cell immunoreactivity to HPB45 (melanocytic differenciation) and absence of CD10, vimentin, cheratines (Cam 5.2, AE1.3) and HNF

PREOPERATIVE

Fig.1



POSTOPERATIVE

Fig.2A

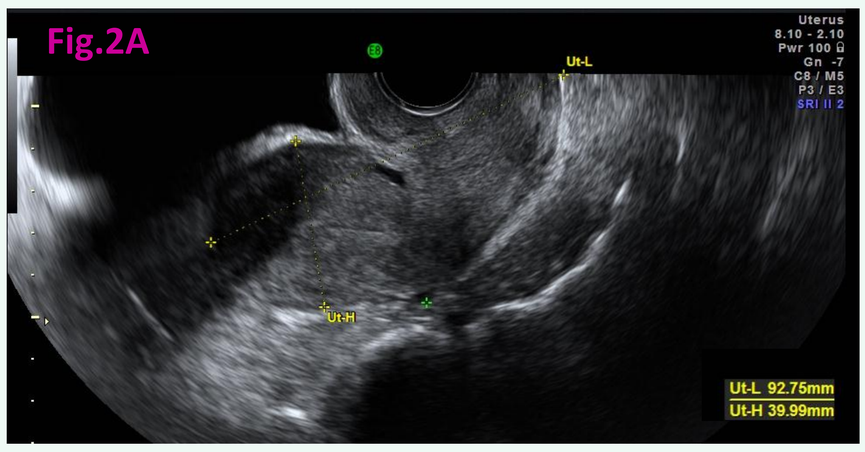


Fig.2B

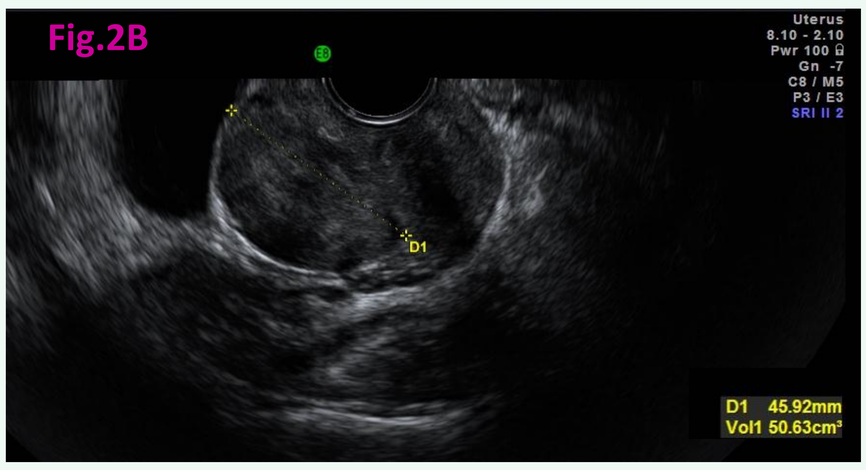
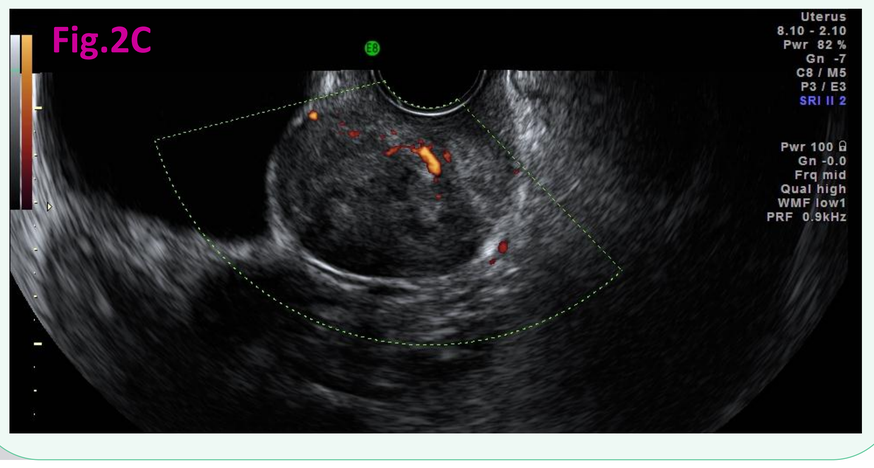


Fig.2C



RESULTS - Evolution

During the follow-up after the resection, the patient explains clinical improvement. In the consecutive visits, physical examination evidenced non-altered visible vagina and cervix. Transvaginal ultrasound showed a **46mm well-defined rounded solid image at the cervical-isthmic level, positive power Doppler-signal, compatible with an anterior cervical myoma (Fig.2A-C).**

Nowadays the patient remains asymptomatic, so an expectant management is decided while concluding the local extension study by MRI and a follow-up ultrasound in 4 months to assess growth or stability of the mass .

CONCLUSION

Uterine PEComas are exceptional tumors, difficult to diagnose and categorize, with a variable but defining and predictive histoimmuno-genotype. We exposed the case of an apparent cervical-origined PEComa shaping intravaginal extensive growth and extrusion, simulating a myoma calved. It expressed melanocytic and myoepithelial cell differenciation, but lacked poor prognosis molecular markers according to the reviewed literature. After having performed the initial surgical resection, it seemed reasonable to stand an expectant management of the remaining tissue with clinical and imaging follow-up.