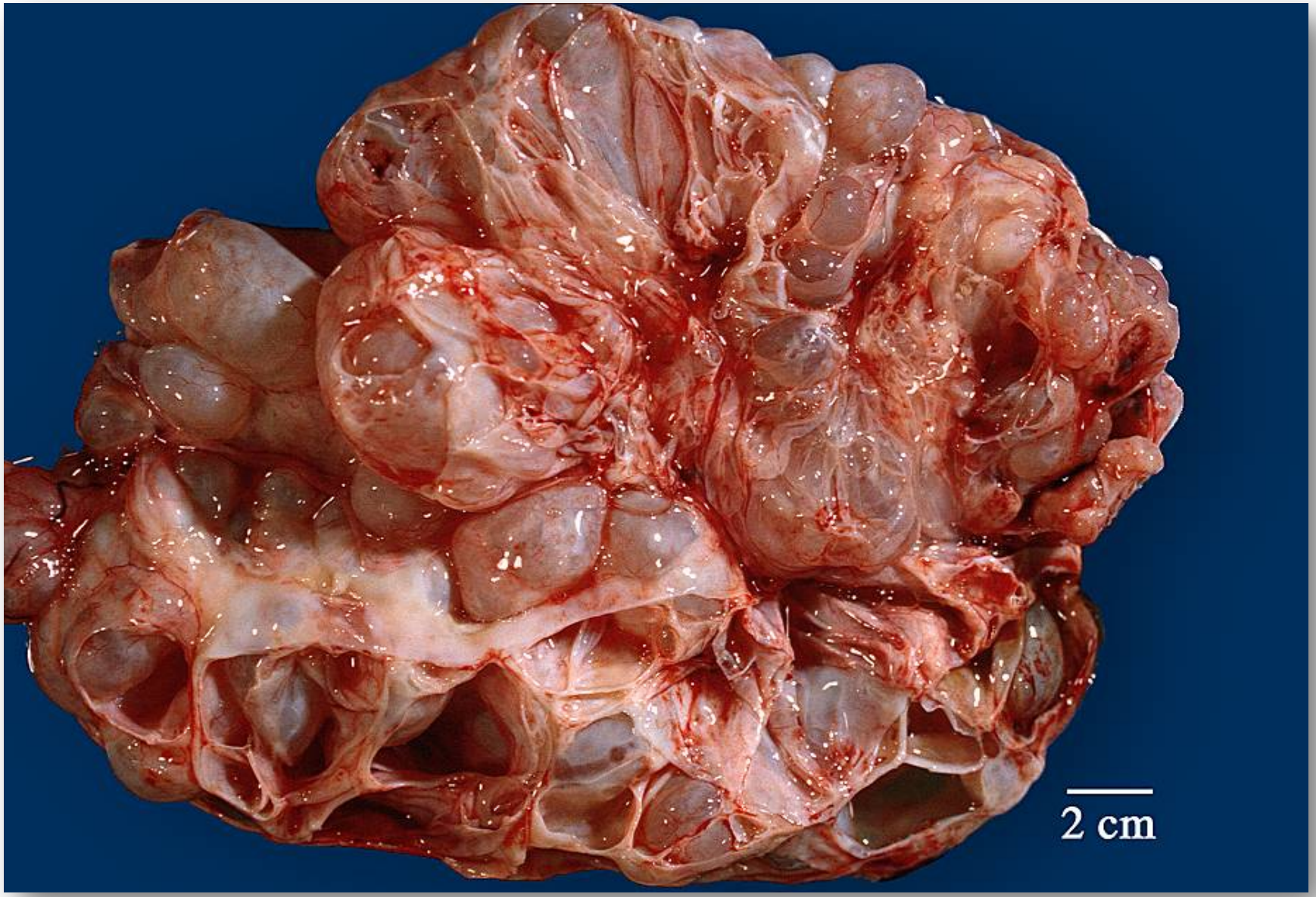
Ovarian mucinous tumor



- **Most of mucinous carcinomas in ovary are metastatic.**
- **The majority of ovarian mucinous tumors are mucinous borderline tumors.**
- **True ovarian mucinous carcinomas are uncommon.**
- **They come from mucinous borderline tumors and teratomas.**
- **Mutations in *KRAS*, *TP53* and *RNF43*.**

Most ovarian primary mucinous carcinomas are well differentiated and moderate and poorly differentiated tumors are relatively uncommon. Mucinous carcinomas are histologically heterogeneous, containing areas of cystadenoma and atypical proliferative tumor intimately admixed with areas of carcinoma. Next-generation sequencing reports that KRAS-activating mutation is the most common molecular genetic alteration in mucinous carcinomas, occurring in 65% of tumors. More importantly, mutations in KRAS , BRAF, and/or ERRB2 amplification are present in > 90% of mucinous carcinomas, suggesting that RAS/MEK pathway activation is common in this neoplasm. Another sequencing study identified mutations in a novel gene, RNF43 and 21% of mucinous carcinomas harbor the inactivating mutations of RNF43 , a zinc finger-dependent E3 ubiquitin protein ligase, suggesting that RNF43 inactivation may characterize a proportion of mucinous cancers. In contrast to other type I ovarian carcinomas, *TP53* mutation is frequent in mucinous carcinomas, being present in approximately one-half of cases. In contrast to serous, endometrioid, and clear cell tumors, mucinous tumors do not express ER or PR, which is consistent with our proposal that they are none Müllerian derived tumors. However, paired box 8 (PAX8), a Müllerian marker, is expressed in approximately 50% of mucinous tumors. We have postulated that the nongerm cell mucinous tumors develop from Brenner tumors, which are in turn derived from nests of transitional epithelium at or near the tuboperitoneal junction. Accordingly, a possible explanation for expression of PAX8 by a substantial number of mucinous tumors may be that in the region of the tuboperitoneal junction, where Müllerian-derived tubal epithelium is in close contact with the mesothelium of the tubal serosa and ovarian surface epithelium, overlapping PAX8 expression may occur in both Müllerian and non-Müllerian epithelium (Kurman and Shih, Am J Pathol, in press, 2016)