Editorial

Primary ovarian signet ring cell mucinous carcinoma: is a real entity or is a missing primary neoplasm in another organ?

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Of all ovarian epithelial neoplasms, the primary ovarian mucinous carcinoma represents 2-3%.[1] The presence of signet ring cells in primary ovarian tumors simulates the metastatic signet ring cell carcinoma known as Krukenberg tumor.[2] The gastrointestinal tract is the most common primary site reported to metastasize to the ovary,[2,3] followed by the pancreas, biliary tree, breast, urinary bladder, uterine cervix, and renal pelvis.[4,5] Pathologists consider the presence of signet ring cells a significant pathological indicator leaning towards a metastatic rather than a primary neoplasm, however, this finding by itself does not definitively indicate a metastatic lesion.

Furthermore, characteristics such as bilaterality, small size, a nodular gross or microscopic appearance, notable histological variation within the tumor, invasive stromal destruction, surface tumor implants, presence of tumor cells within mucin, solitary cell invasion of stroma, extraovarian dissemination, and extensive lymphovascular invasion, particularly at the ovarian hilum, are indicative of a secondary mucinous neoplasm.[6] To differentiate between primary and metastatic signet ring cell mucinous carcinoma of the ovary, immunohistochemistry may be applied as an additional method. However, the distinction between primary and secondary ovarian mucinous neoplasms remains challenging due to the fact that many primary tumors demonstrate intestinal differentiation and express enteric markers such as CK20, CA 19-9, ACE, CDX2, and CK7.[7,8]

In the rare cases of primary ovarian signet ring cell mucinous carcinoma documented in the literature,[9,10] the authors support their hypotheses based on:

a) Unilateral presentation, substantial tumor size, presence of malignant glands within a fibrous stroma, absence of surface implantations, lack of lymphovascular invasion, and absence of extraovarian dissemination.

b) The non-existence of various other features typical of metastatic neoplasms and the presence of mixed benign-appearing areas.

c) And the absence of other lesions in the gastrointestinal tract in different explorations.

The main counterargument to recognizing the primary ovarian Krukenberg tumors revolves around the potential oversight of a small occult primary neoplasm, particularly in organs like the stomach. In 1968, Joshi[11] reported a case of "primary Krukenberg tumor of the ovary" and examined 38 cases documented in the literature. Among these, 17 cases were deemed acceptable based on either meticulous autopsy findings or survival extending to 5 years or more. So, the question sill, How many years of follow-up are required to definitively confirm the diagnosis of primary ovarian signet ring cell mucinous carcinoma?

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Conflicts of interest

None.

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