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Ovarian Metastases of Appendiceal Tumors with Goblet Cell Carcinoid-Like Features: Limited Expression of Neuroendocrine Markers and Clinicopathologic Features of Aggressive Invasive Carcinoma Support Designation as Adenocarcinoma Rather Than Goblet Cell Carcinoid Tumor

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Background: Appendiceal tumors exhibiting goblet cell carcinoid-like features are often designated as goblet cell carcinoid tumors or “adenocarcinoids”, suggesting they are indolent neuroendocrine tumors or mixed tumors with behavior intermediate between carcinoid and adenocarcinoma. However, those infiltrative appendiceal tumors metastatic to the ovary have demonstrated clinicopathologic features more characteristic of aggressive invasive adenocarcinomas (Am J Surg Pathol 1997;21:1144-1155).

Design: Immunohistochemical analysis of neuroendocrine marker expression was performed on 16 ovarian tumors and 15 of the corresponding primary appendiceal tumors. Percentage of positive tumor cells was estimated to the nearest 5%, with <5% regarded as negative.

Results: All appendiceal tumors exhibited transmural invasion and the ovarian tumors were bilateral in 14 cases for which data was available for both ovaries. The appendiceal and ovarian tumors exhibited a variety of patterns of differentiation, including glandular and signet ring cell, with all displaying some goblet cell carcinoid-like patterns (nests, islands, or crypt-like tubules with goblet cells). Chromogranin was expressed in 5 of 14 appendiceal tumors (mean/median: 8%/0%; range: 0-50%) and synaptophysin was expressed in 3 of 13 of these (mean/median: 4%/0%; range: 0-25%). Chromogranin was expressed in 5 of 16 ovarian tumors (mean/median: 5%/0%; range: 0-20%) and synaptophysin was expressed in 1 of 15 of these (mean/median: 0.3%/0%; range: 0-5%). Follow-up was available for 12 patients: 7 patients had died of disease at intervals ranging from 4-30 months (mean/median: 15/12) and 5 patients were alive with disease at 1-40 months.

Conclusion: Some infiltrative appendiceal tumors may arise from pre-existing goblet cell carcinoid tumors and could represent mixed carcinoid/adenocarcinomas but limited expression of neuroendocrine markers combined with established clinicopathologic features of aggressive carcinoma support designation of these tumors as invasive adenocarcinomas. The ovarian metastases should be labeled as metastatic adenocarcinomas to reflect their clinicopathologic features and distinguish them from the rare true primary ovarian goblet cell carcinoid tumors of germ cell origin.