A Case Series of 27 Primary Ovarian Neuroendocrine Tumors

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Background: Primary ovarian neuroendocrine tumors are rare and are often found incidentally. Differentiation of primary ovarian carcinoids from metastatic carcinoids is difficult. We examined 27 primary ovarian carcinoids treated over the course of 21 years, one of the largest series in the literature.

Methods: All patients with ovarian neuroendocrine tumors at a single institution between 1994 and 2015 were retrospectively reviewed in this IRB approved analysis. Pathologic review confirmed 27 primary ovarian carcinoids.

Results: The mean age of our cohort was 48.7 years (range 23-75 years). The majority of tumors were found to be benign. Nineteen patients (70.4%) had carcinoids associated with dermoid cysts, of which 9 were associated with thyroid tissue (strumal carcinoids). Seven patients (25.9%) had ovarian mucinous tumors, of which 3 were benign and 4 were malignant. Three of the malignant carcinoids were of the mucinous type, and one malignant tumor was of the insular subtype. Bilateral disease was found in 2 patients. All patients with benign tumors had no evidence of disease at time of review. Of the 4 patients with malignant tumors, one is alive with disease, two have died of disease, and one was lost to follow up. The estimated 5-year overall survival for patients with benign disease was 86%. Immunohistologic stains were positive for both chromogranin and synaptophysin in 16 cases. 6 cases were positive for neuron-specific enolase, 2 were positive for thyroglobin, and 7 were positive for CDX2.

Conclusion: Our case series is one of the largest reported. The average age of our patients was younger than previously reported. Most of the carcinoids were associated with teratomas and the majority of the malignant tumors were of the mucinous variant. In contrast to the existing literature, our cohort had a significant portion of strumal carcinoids.