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Carcinoid Heart Disease in Patients Diagnosed with Small Bowel and Lung Neuroendocrine Tumors

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BACKGROUND

Carcinoid heart disease (CHD) is a complication of neuroendocrine tumors (NETs). While its pathophysiology and manifestations are described, little is known about its occurrence in all patients diagnosed with NETs. We examined the occurrence of CHD and explored the use of echocardiography after diagnosis of NET.

METHODS

We conducted a population-based retrospective cohort study of adults diagnosed with small bowel and lung NETs (2000-2019). CHD was defined as new congestive heart failure or valvular disease. Cumulative incidence functions (CIF with 95%CI) of CHD and use of echocardiography were computed accounting for the competing risk of death. Fine-Gray models examined factors associated with CHD (reported as sub-hazard ratios – sHR with 95%CI).

RESULTS

Of 5,735 patients with NETs, 54.1% had small bowel primaries and 48.8% metastatic disease. Median follow-up was 52 months (inter-quartile range 20-99). The CIF of CHD in all patients were 7.8% (7.0-8.4%) at 5 years and 10.7% (9.8-11.7%) at 10 years. CHD was more frequent for small bowel (10-year CIF 12.7% [11.2-14.2%]) than lung (10-year CIF 9.1% [8.0-10.3%]) NETs. No difference was observed by metastatic status. Of 1,864 patients with available urinary 5HIAA data, 64.0% had elevated results. CHD was more frequent with elevated serotonin (10-year CIF 13.5% [11.3-15.9%]) than without (10-year CIF 9.0% [95%CI 6.4-12.2%]). Higher comorbidity burden (sHR 1.3 [1.1-1.6]), small bowel primary (sHR 1.4 [1.2-1.6]), and serotonin secretion (sHR 1.5 [1.1-2.1]) were associated with increased incidence of CHD. CIF for use of echocardiography in all patients was 64.7% (63.3-66.1%) at 10 years. Use of echocardiography reached >50% in all sub-groups. Patterns of echocardiography CIF mirrored those of CHD across sub-groups.

CONCLUSIONS

CHD occurred in 10 out of 100 patients in the 10 years after small bowel and lung NETs diagnosis. Patterns of echocardiography use suggest that testing is not influenced by NET disease characteristics, with risks of under-detection in at-risk individuals. Knowledge of factors associated with CHD can be used to target future screening efforts for early diagnosis of CHD.