Carney's complex remains a puzzle

THE CASE of Carney's complex described by Wallace and associates in this issue of the *Cleveland Clinic Journal of Medicine* is fascinating and instructive. A family's index case of this autosomal dominant disease could present first to any one of several specialists, including endocrinologists, cardiologists, internists, dermatologists, and pathologists—all of whom need to be familiar with the syndrome and its management.

■ *See Wallace and associates (pp248–256).*

I first learned of the patient described in this issue in 1974, during a review of the cases of Cushing's syndrome seen at the Cleveland Clinic from 1950 to 1974; I did not realize what entity plagued this patient until about 10 years later, when Carney and his colleagues first began to write the series of articles that identified and characterized Carney's complex.[1–3]

The complex includes a rare form of Cushing's syndrome caused by primary pigmented micronodular hyperplasia (or dysplasia), cardiac and cutaneous myxomas, and other pathologic lesions.

Low ACTH levels, lack of response of steroid levels to dexamethasone administration, and normal adrenal size on computerized tomography or magnetic resonance imaging should alert one to the diagnosis of this unusual form of Cushing's syndrome associated with Carney's complex. If Cushing's syndrome is the first manifestation of the complex, precise diagnosis needs to be made, since only adrenal surgery can cure Cushing's syndrome associated with Carney's complex.

When the diagnostic criteria strongly suggest pigmented micronodular adrenal hyperplasia or it is confirmed after adrenal surgery, the patient's problems are only beginning. Given our present understanding of the disease, it is proper to assume that all patients with this type of Cushing's syndrome have Carney's complex and should be examined for cardiac myxomas and other tumors. Since the disease is transmitted as an autosomal dominant trait,[4] family members should be screened for Cushing's syndrome, cardiac myxomas, and other tumors. Although the patient reported by Wallace and associates died from thromboembolic events related to cardiac myxomas, progress in cardiac surgery has improved the chances for successful removal of cardiac myxomas.

Many questions remain about the management of these patients. For example, if a patient has had cardiac myxomas successfully removed, what are the chances of new myxomas developing in the future? If no cardiac myxoma is found at the time Cushing's syndrome is diagnosed, how often and for how many years should echocardiograms be performed? If clinically silent Cushing's syndrome is discovered, should one wait for some clinical damage or correct the syndrome when it is first discovered? Should affected males be screened yearly for testicular tumors by physical examination and ultrasound? For how many years? These issues will be resolved only after years of observation.

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REFERENCES

1. Shenoy BV, Carpenter PC, Carney JA. Bilateral primary pigmented nodular adrenocortical disease. Am J Surg Pathol 1984; **8**:335–344.
2. Carney JA, Gordon H, Carpenter PC, Shenoy BV, Go VLW. The complex of myxomas, spotty pigmentation, and endocrine overactivity. Medicine (Baltimore) 1985; **64**:270–283.
3. Grant CS, Carney JA, Carpenter PC, Van Heerden. Primary pigmented nodular adrenocortical disease: diagnosis and management. Surgery 1986; **100**:1178–1183.
4. Carney JA, Hruska LS, Beauchamp GD, Gordon H. Dominant inheritance of the complex of myxomas, spotty pigmentation, and endocrine overactivity. Mayo Clin Proc 1986; **61**:165–172.