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An unusual presentation of carcinoid syndrome

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CASE REPORT

Thirteen years old male patient has admitted to oncology department for tumor that made extensive liver metastases with an unknown primary origin. As a result of investigations it was diagnosed as a neuroendocrine tumor.
Repeated echocardiographic studies have showed progressive right ventricular failure, loss of coaptation and thickening of the tricuspid valve, moderate pulmonary stenosis and hypoplasia. Primary or metastatic cardiac involvement was not observed in whole body scintigraphy scan. Therefore, these symptoms were thought to be due to circulating neurotransmitters rather than primary tumoral invasion. Tricuspit biological valve replacement and pulmonary valvotomy were performed. Aggressive anti-tumor treatment was started simultaneously.

DISCUSSION

Carcinoid tumors are neuroendocrine tumors which usually originate from appendix and terminal ileum. Carcinoid syndrome is a clinical entity characterized by episodic flushing, secretory diarrhea, hypotension and bronchospasm linked to neurotransmitters. Cardiac involvement is observed in 50-60% of patients with carcinoid syndrome. Carcinoid heart disease is the most important cause of morbidity in patients with carcinoid syndrome. It is speculated that serotonin is the main neurotransmitter responsible for the carcinoid heart disease. Particularly activation of 5-HT2A give rise to myofibroblast deposition and accumulation of extracellular matrix on valves surface. Thereby possibility of valvular proliferation and fibrosis may increase. Due to high level of neurotransmitters are metabolized in the lung, the primary target of the carcinoid syndrome are right heart valves. However, 10% of those can be found with left heart involvement. Although carcinoid tumors have a slow progression; presence of cardiac involvement significantly decreases the life expectancy and quality. The diagnosis of this disease might be difficult and requires high index suspicion. Medical therapy, is performed to reduce the tumor burden, neurotransmitter activity and heart failure symptoms. In resistant cases valve surgery is preffered.

CONCLUSION
Early diagnosis and treatment is important in carcinoid heart syndrome.