Toothache uncovered the biatrial masses

Biatriyal kitleyi ortaya çıkaran diş ağrısı

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Right Answer: 2. Carney syndrome

Cardiac mass is defined as an abnormal structure within or neighboring to heart. There are basically 3 subtypes of cardiac masses including tumor, thrombus and vegetations. Although it is not possible in all cases to differentiate these masses from each other, a reasonably secure diagnosis often can be made by integrating the clinical data, echocardiographic appearance and other imaging modalities (1, 2).

Primary tumors of the heart are less frequently seen than non-primary tumors. Also a tumor can only be defined as a benign or malign via pathological examination of tissue. Although 75% of the primary cardiac tumors are benign, they can exert malignant physiological consequences like obstruction in the normal blood flow pattern. Myxomas are the most common primary cardiac tumors in adults. Cardiac myxomas most often are single, arising from the fossa ovalis of the interatrial septum and protruding into the left atrium (in 3/4 of the cases). It can be seen at >1 site in about 5% of cases (3). Due to tumor localization and size, it may protrude into the other cardiac chambers like left or right ventricle through atrioventricular valves in whom tumor "plop" on auscultation may be heard and may guide physicians for further cardiac evaluation. In approximately 7% of cases, myxomas are familial, with most pronounced case being the Carney complex. This syndrome is an autosomal dominant inherited disease, where multiple recurrent myxomas, extracardiac myxomas (breast, skin, testis, thyroid or adrenal gland), Schwannomas, spotty pigmentation of the skin or mucosal surface and endocrine overactivity or endocrine tumors (like pituitary adenomas with gigantism or acromegaly), may coexist (4). In patients with atrial myxomas in the setting of the Carney complex, relapses are frequent, and close follow-up of the patients is essential (5).

The Carney complex should be distinguished from other syndromes named with the term "Carney" which may cause confusion. Prominent among them are the Carney-Stratakis syndrome and the Carney triad, neither of which include cardiac tumors (6). Carney triad is a usually sporadic association of pulmonary chondroma, gastrointestinal stromal tumors, and paraganglioma. The most of the patients have two of these tumors, the gastric and pulmonary tumors being the most common combination. Carney Stratakis syndrome is an association of familial paraganglioma and gastric stromal sarcoma and it is considered to be a distinct condition from Carney triad as it is dominantly inherited and not associated with pulmonary chondroma (6). Our patient had no paraganglioma, pulmonary or gastrointestinal tumors which excludes Carney Stratakis syndrome and the Carney triad.

Also, the diagnosis of neurofibromatosis type I is based on the presence of several criteria including café-au-lait spots, neurofibromas, axillary or inguinal multiple freckles, ocular and/or orthopedic problems and epilepsy. Due to absence of these diagnostic criteria, neurofibromatosis type I was also excluded in our patient.

Our patient was diagnosed as "Carney syndrome" because of the presence of multiple endocrine overactivity and multiple recurrent atrial myxoma. So, careful history taking and cardiac auscultation which were the main components of patient evaluation have helped us for correct diagnosis of the patient.

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