Left Atrial Myxoma and Trichilemmal Cysts

Abstract: A 56-year-old man had a 6 x 5-cm mass originating from the fossa ovalis in the atrial septum found by echocardiography and multiple enlarging scalp masses. The mass had a regular surface and was prolapsing into the mitral valve during diastole. The tumor was successfully treated by surgical excision, which revealed a well-defined mass with a narrow-base stalk originating from the fossa ovalis in the atrial septum. Histological examination confirmed the diagnosis of myxoma. The scalp masses were also removed surgically; histopathologic examination corresponded to benign trichilemmal cyst. The patient recovered without complication and was discharged 10 days after the operation.

Key Words: Myxoma, trichilemmal cyst

Sol Atriyal Miksoma ve Trikilemmal Kist


Anahtar Sözcükler: Miksoma, trikilemmal kist

Introduction

Atrial myxoma is the most common cardiac neoplasm, and up to 80% of myxomas are localized in the left atrium, of which 75% involve the interatrial septum. Although they are benign mesenchymal and usually polypoid myxomatous or pedicile tumors, they may cause 3 types of clinical presentation: obstruction, embolism and constitutional (1,2).

Trichilemmal cyst (TC), a rare tumor originating from the outer root sheath of the hair follicle, is typically seen in middle-aged or elderly patients, with a strong predilection for women, and is particularly localized on the scalp (3,4).

We present the case of a 56-year-old man with a left atrial myxoma and multiple enlarging scalp masses.

Case Report

A 56-year-old man presented with exercise dyspnea, and slowly growing masses on the scalp. Dyspnea had been present for 1 year and was prominent with physical activity. The masses on the scalp had been growing slowly since he had first noticed a small lump 15 years earlier. On examination, a grade 2/6 holosystolic murmur and a low mid-diastolic murmur were audible in the apex. Pulse rate was 100 beats/min/regular and blood pressure was 100/60 mmHg. On auscultation, the lungs were clear. Multiple masses were noted on the scalp, ranging in size from 1-7.5 cm, although the largest was 7.5 x 6.0 cm in size and soft, immobile, fluctuating; the others were hard, non-tender, immobile and of varying diameters. The overlying skin was normal in color and texture (Figure 1). No abnormal neurological symptoms or signs were found. Neither electrocardiogram nor chest X-ray showed any abnormalities.
Transthoracic and transesophageal echocardiography identified a left atrial mass, 6 cm in maximal diameter, hemodynamically similar to mitral stenosis. The tumor had a stalk that allowed it to extend through the mitral valve into the left ventricle during diastole (Figure 2A). The stalk of the tumor seemed to originate from the atrial septum in the fossa ovalis region (Figure 2B). Because of the localization, this mass caused mitral inflow obstruction with a maximal gradient of 14 mmHg and mean gradient of 8 mmHg (Figure 3 upper panel). A flow of mild grade mitral regurgitation along the lateral wall of the left atrium was seen by color Doppler echocardiography. Left ventricular function and dimensions were normal.

With an assumption of tumor metastasis, contrast enhanced computed tomography (CT) of the head and thorax was performed the same day. The CT study of the thorax showed a well-defined left atrial mass attached to interatrial septum extending into the left ventricle (Figure 4A). The CT of the head revealed multiple large, subcutaneous, hypodense solid masses with rare calcific areas.

The patient was taken to cardiac operation in order to excise the tumor, and the gelatinous mass with a stalk attached to interatrial septum was removed with its pedicle completely (Figure 4B, Figure 5A). Microscopically, the tumor consisted of an abundant mucopolysaccharide matrix and vascular channels with varying numbers of distinctive stellate or plump “myxoma” cells. The mucoid matrix stained positively with Alcian blue and mucicarmine (Figure 5B).

After the operation mitral inflow pattern was normal (Figure 3 lower panel, Figure 6)

One week after the first operation, all masses on the scalp were resected completely. The macroscopic appearance of the dominant masses appeared as firm, smooth, white-walled cysts (Figure 7A). Microscopic examination revealed a well-circumscribed, partly cystic mass. Cysts were lined by stratified epithelium showing trichilemmal keratinization in which the individual cells increase in bulk. The cells were cytologically benign. The cystic areas were filled with trichilemmal-type keratin (Figure 7B).

The patient did well after the operation and was discharged on postoperative day 10. A schedule of routine follow-up visits was arranged to monitor the recurrence of both atrial myxoma and trichilemmal cysts.
Discussion

We herein report a case with a large left atrial myxoma producing symptoms of mitral valve obstruction. In patients with symptoms of left heart failure, left atrial myxoma remains an important differential diagnosis (1,2,5). As clearly seen in our case, most myxomas arise from the interatrial septum, with a pedicle at the border of the fossa ovalis and prolapsing across the mitral valve orifice. Prolapsing and polypoid myxomas have been associated with systemic and coronary embolism (2,5). A higher risk of embolization has been reported and events occur in 30% to 43% of the patients (6). Our patient did not experience any embolic event despite prolapsing myxoma. Transthoracic and transesophageal echocardiography can generally be used to determine the location, size, shape, attachment and mobility (1,5,7).
Figure 5. (A) Macroscopic appearance of the resected myxoma with smooth, shiny surface (B). Microscopic appearance of cardiac myxoma shows minimal cellularity. Only scattered spindle cells with scant pink cytoplasm are present in a loose myxoid stroma (Hematoxylin eosin stain x 100).

Figure 6. Apical 4-chamber view after the operation (LV: Left ventricle, LA: Left atrium, RA: Right atrium, RV: Right ventricle).

Figure 7. (A) Macroscopic appearance of the trichilemmal cysts. (B) Trichilemmal cyst lined by stratified squamous epithelium exhibiting trichilemmal keratinization (Hematoxylin eosin stain x 200).
Trichilemmal cyst most commonly occurs on the scalp during the fourth to eight decade of life as a solitary lesion and is seen more frequently in women than men. They are benign lesions but rarely exhibit malignant transformation (4,8,9). The tumor presented in this case is worth further mention. It occurred in a man and originated from the scalp, the usual location for this tumor, but there were multiple masses, which has never been reported previously. It is also surprising that careful histological examination did not indicate malignant transformation despite multiple formations in size and number. On further examination the patient also had no evidence of metastatic disease.

In this report, we describe a patient with different benign tumors found together incidentally.

References