Case report

Papillary fibroelastoma of a mitral valve chorda

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Received 15 May 2001; received in revised form 22 August 2001; accepted 3 September 2001

Abstract

A 43-year-old woman presented with an ischemic stroke in the right middle cerebral artery territory. Cardiac echography disclosed a tumor of a primary chordae of the anterior leaflet of the mitral valve. After neurologic recovery, the patient was referred to surgery for excision of the tumor and plastic reconstruction using a chordal transfer technique. Histological examination of the tumor showed a typical papillary fibroelastoma. Papillary fibroelastoma is the third most frequent cardiac benign tumor. The high embolic potential of this tumor is in favor of an aggressive surgical attitude.

Keywords: Ischemic stroke; Mitral valve chordae; Fibroelastoma; Mitral valve reconstructive surgery

1. Introduction

Cardiac papillary fibroelastoma (CPFE) represent less than 16% of all primary cardiac tumors, but is the third most common primary cardiac neoplasm after myxoma and lipoma in necropsy series [1] and the most common valvular tumor [2]. In 1996, Hicks et al. [3] reviewed four cases of isolated chordal CPFE. We report here a new case in this location.

2. Case report

A 43-year-old woman presented at our hospital with a sudden onset of right upper-limb weakness and aphasia on January 6, 2000. Her past medical history was unremarkable except for an episode of aphasia subsiding in a few minutes in August 1999. On physical examination, a left facial palsy was associated with a motor deficit of the left upper limb. No other anomalies were present and routine investigations (electrocardiography (EKG), chest X-ray and usual laboratory work) were negative.

Cerebral computerized tomography (CT) scan and magnetic resonance imaging put in evidence an ischemic lesion with mass effect in the territory of the right middle cerebral artery, suggesting an embolic process. Carotid duplex ultrasound exploration was normal.

Transthoracic echocardiography (TTE) disclosed a mobile mass on the mitral valve apparatus. Transesophageal echocardiography (TEE) showed the mass to originate from a mitral chordae attached to the free edge of the central portion of the anterior leaflet, and close to this attachment (Fig. 1). The mass was estimated to be 5 mm wide. A small patent foramen ovale was also present.

Doppler ultrasonography examination of the lower limb veins, 24 h EKG and blood pressure recordings, coagulation screening in search of hypercoagulability disorders, right heart catheterization and coronary arteriography were performed and showed no abnormalities.

Diagnostic conclusion was embolic cerebral stroke related to a tumor of the chordal mitral valve apparatus, probably a papillary fibroelastoma.

Surgical excision of the tumor was programmed, but belated until full complete neurologic recovery. On March 3, 2001, the mitral valve was approached through a biatrial transeptal incision. A 6 mm diameter polylobular translucent mass was attached to a main paramedial chordae of the anterior leaflet, 2 mm far from the edge of this leaflet (Fig. 2). This mass and the adjacent chordae were excised. A secondary chordae was mobilized and reimplanted on the free edge of the leaflet. The minimal atrial septal defect was closed by suturing the biatrial incision. Immediate and postoperative TEE showed a normally functioning mitral valve. The post-operative course was uneventful. Histopathologic examination of the mass confirmed the diagnosis of papillary fibroelastoma.
One year later, the patient remains asymptomatic without any medication.

3. Discussion

CPFE derive from normal components of the endocardium and resemble a sea anemone with multiple papillary fronds. Histologically they present a central core of dense connective tissue. Surrounding this central core is a layer of hyperplastic endocardial cells, loose connective tissue and a mesh of elastic fibers. The tumor rarely exceeds 1 cm in diameter [2].

Grinda et al. [4] in a review of 198 cases of CPFE found the location of CPFE to be aortic in 29%, mitral in 25%, tricuspid in 17%, pulmonic in 13% and non-valvular in 16%. In the same year, Di Mattia et al. [5] collected only 34 observations of surgical resection of a mitral papillary fibroelastoma. Chordal CPFE as an isolated location appears more rarely in the literature [6–8].

The real incidence of CPFE remains controversial as evolution can be asymptomatic for years [8]. However, all patients are at risk of embolic neurological strokes [9]. Embolization may occur from the fragile papillary fronds [3] or from a platelet and fibrin aggregation forming on the tumor [10]. Chordal CPFE as other mitral valve tumors are potentially life-threatening tumors, likely to produce serious neurological symptoms or sudden death [2].

Transthoracic and specially transesophageal echography is the most sensitive and reliable investigation to establish the diagnosis, exclude multiple locations and plan the optimal surgical approach [4]. In the reported case, echography disclosed another potential cause of emboli, a patent foramen ovale; so the embolic responsibility of CPFE could not be fully established.

Even small CPFEs (3 mm) can be responsible for strokes [4]. The effectiveness of preventive systemic anticoagulation remains controversial [4,5,11]. Even if the patients are asymptomatic and if there are no major contraindications to operation, they should be surgically managed. In case of post-embolic brain damage, a delay of 4–6 weeks is generally recommended between the embolic episode and intra-cardiac surgery [12].

Conservative management of mitral lesions is generally possible [4,5]. Chordal lesions can be repaired by plication, chordal transfer or use of artificial chordae. Recurrence has
not been reported as long as the tumor has been completely resected [6].

Acknowledgements

We are grateful to Dr Y. Couvreur for the histopathologic examination and to Dr Yvan Pamjanov (Indianapolis medical centre) for reviewing it, and to Dr Alain Brunet (CHU St Pierre-Bruxelles) for reviewing the manuscript.

References