

Solitary thickened papillary muscle with mitral valve abnormalities: Uncommon form of hypertrophic obstructive cardiomyopathy.

## Hypertrophie isolée du muscle papillaire avec anomalies de la valve mitrale : Forme rare de la cardiomyopathie hypertrophique obstructive

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### Résumé

Nous rapportons le cas d'un patient de 40 ans se plaignant d'une aggravation progressive d'une dyspnée d'effort. L'échocardiographie a révélé un muscle papillaire (MP) antérolatéral hypertrophié de façon isolé avec déplacement apical et élongation de la valve mitrale occasionnant un mouvement systolique antérieur avec une obstruction intra-ventriculaire (gradient systolique maximal au repos de 168 mmHg) ainsi qu'une régurgitation mitrale excentrée et rasant la paroi inférolatérale de l'oreillette gauche. Par ailleurs Il n'y avait aucune hypertrophie pariétale évidente. L'hypertrophie isolée du muscle papillaire est un type peu fréquent de cardiomyopathie hypertrophique. Cela peut provoquer une obstruction intra-ventriculaire. Le traitement chirurgical dépend du MP concerné (principal ou accessoire), ses attaches aussi bien avec les feuillets valvulaire mitraux qu'avec avec le septum inter-ventriculaire ainsi que de la gravité de l'hypertrophie.

### Mots-clés

Muscle papillaire, Cardiomyopathie hypertrophique, Obstruction ventriculaire gauche, Insuffisance mitrale, Echocardiographie.

### Summary

We report a case of a 40-years-old man presented with a chief complaint of progressively worsening dyspnea. Echocardiography revealed isolated hypertrophied anterolateral papillary muscle (PM) with apical displacement and mitral valve elongation occasioning an anterior systolic motion with severe left ventricular outflow tract obstruction (Peak systolic gradient at rest of 168 mmHg) and eccentric mitral regurgitation inferolaterally oriented. There was no obvious parietal hypertrophy elsewhere. Solitary papillary muscle hypertrophy is an uncommon type of hypertrophic cardiomyopathy. It may cause intraventricular obstruction. Surgical treatment depends on the concerned PM (principle or accessory), its extent to the mitral leaflet or the septum or both and the severity of hypertrophy.

### Keywords

Papillary muscle, hypertrophic cardiomyopathy, left ventricular obstruction, mitral regurgitation, Echocardiography.

### Correspondance

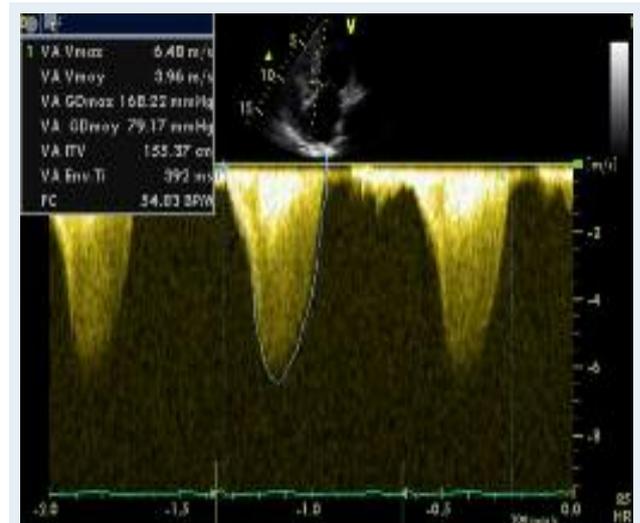
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## BACKGROUND

Solitary papillary muscle (PM) hypertrophy with mitral valve abnormalities could be a unique form of hypertrophic cardiomyopathy. Hypertrophy concerns only the papillary muscle and spares the other parietal left ventricle segments (1, 2). This entity has recently drawn attention owing to the left ventricular outflow tract (LVOT) obstruction it might cause.

## CASE REPORT

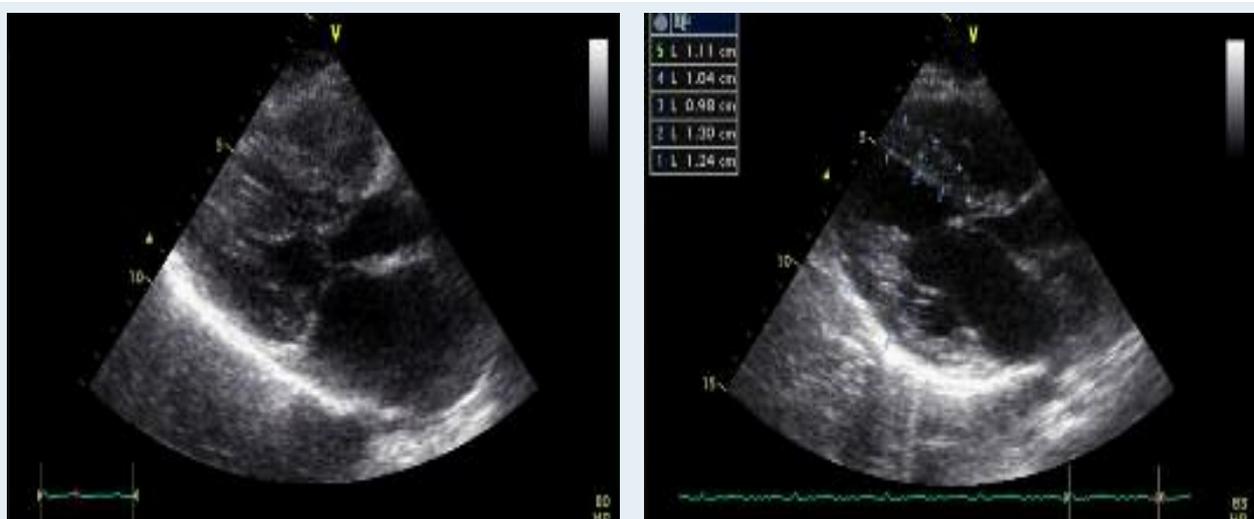
We report a case of a 40-years-old man whose chief complaint was a progressively worsening dyspnea. His past medical history was largely unremarkable, as was his family medical history. The cardiac examination found a markedly 4/6 harsh systolic ejection murmur in the aortic area with no radiation to the neck and holosystolic murmur heard at the apex and radiating to the axilla. Transthoracic echocardiography (TTE) showed hypertrophied anterolateral papillary muscle (a muscular strip making 13mm of thickness) without obvious parietal left ventricle hypertrophy revealed elsewhere (Figures 1a,1b, video1) and elongated mitral leaflets causing a systolic anterior motion (SAM) with severe obstruction of the LVOT (Resting LVOT maximum gradient=168 mmHg, mean gradient=79 mmHg) (video2) and late-peaking dagger-shaped appearance on continuous Doppler (Figure 2). Color Doppler showed severe eccentric mitral valve regurgitation (Figure 3). The patient was referred to surgery for a mitral valve replacement and PM resection.



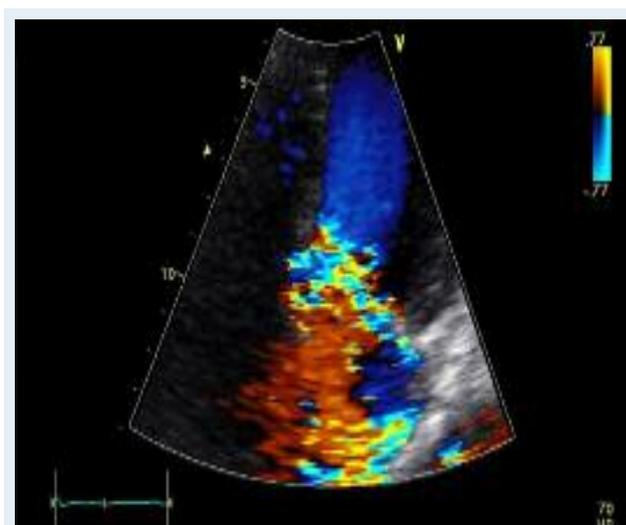
**Figure 2 :** Continue Doppler showing a resting LVOT maximum gradient at 168 mmHg and late-peaking dagger-shaped appearance.

## DISCUSSION

Our patient had an unusual association of solitary main papillary muscle hypertrophy with elongated mitral valves and SAM causing severe intra-ventricular obstruction without evidence of parietal left ventricle hypertrophy. This may be part of peculiar form of hypertrophic cardiomyopathy (HCM) manifested as predominant PM hypertrophy sparing the parietal LV segments.



**Figures 1 a/b.** Two dimensional echocardiography revealing thickened anterolateral papillary muscle (13mm). End-diastolic wall thickness of the rest of left ventricle walls is normal.



**Figure 3 :** Eccentric severe mitral regurgitation laterally oriented with left atrial enlargement

Current guidelines define HCM by a wall thickness  $\geq 15$  mm in one or more LV myocardial segments (3). In hypertrophic obstructive cardiomyopathy (HOCM), the significant dynamic LVOT obstruction is due to mechanical causes inherent to asymmetrical septal hypertrophy resulting in dynamic SAM of mitral leaflets (4). Rare cases of dynamic LVOT or midventricular obstruction due to different morphological papillary muscle abnormalities such as accessory, bifid, octopus and single papillary muscle or solitary papillary muscle hypertrophy have been cited in literature (2, 5-8). Solitary PM hypertrophy, defined as thickness of one or two of the PM more than 11mm, has been recently proved to be a phenotypic variant of HCM (8, 9). Its coexistence with mitral valve abnormalities could also be a unique form of HCM that can lead to a worsening of dynamic LVOT obstruction. Few isolated cases have been reported in literature with mid-ventricular obstruction

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(1-2, 10). It should be pointed out that those patients could potentially develop parietal hypertrophy (septal hypertrophy essentially) and classic HCM phenotype during their follow up, what can be explained by repeated episodes of LVOT obstruction that it is firstly due to solitary PM hypertrophy (1). The mechanism of obstruction in case of solitary PM hypertrophy is produced by an apposition of the papillary muscle with the septum. The hypertrophied papillary muscle can also have a subaortic muscular extension into the septum or can be inserted into it by anomalous chordae tendineae. This muscular midcavity apposition or fusion between papillary muscle and septum can be total, interesting the entire length of papillary muscle, or partial, interesting only the base. It could be at the origin of an additional LVOT tunnel-like obstruction (2, 11-15).

The therapeutic management and especially surgical treatment depends on the concerned papillary muscle (principle or accessory), its extent to the mitral leaflet or the septum or both and the severity of LV hypertrophy. Surgical techniques are varied. If and commonly the anterolateral papillary muscle is concerned, it can be dissected partially free from its attachment with the lateral left ventricular free wall to enhance PM mobility and reduce the anterior tethering of the mitral apparatus (13). PM realignment without septal myectomy through the aortic valve can be realized in case of solitary PM hypertrophy (1, 16). In other cases, the hypertrophied PM can be resected together with the attached normal anterior leaflet in block via the left atrium and the mitral valve is replaced with a mechanical one (17).

## CONCLUSION

Solitary papillary muscle hypertrophy without LV parietal hypertrophy is a rare entity of HCM which can coexist with mitral valve abnormalities and lead to LVOT obstruction. Identification of such atypical presentation of HCM is important to screen patients with sudden cardiac death risk irrespective of LV wall thickness.

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