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Aortic Bicuspidia: About 50 Cases

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Original Research Article

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Abstract: Aortic bicuspidia is the most common congenital heart defect. The two-leaf aortic valve remains functional up to adulthood in most patients, which explains its generally late diagnosis. Its clinical course is unpredictable and heterogeneous. The vast majority of patients develop aortic valvular and parietal complications. The overall prognosis remains good on recent data from the literature.
Keywords: Aortic bicuspidia, Diagnostic, Aortic stenosis, Aortic insufficiency, Treatment.

INTRODUCTION

Aortic bicuspidia (BA) is characterized by the presence of two functional sigmoids instead of three. It is the most common congenital heart defect [1], its prevalence is estimated between 0.5 and 2% according to the studies [1,2]. It is also the most frequent cause of aortic valvulopathies in subjects under 65 years of age [3].

METHODS

We carried out a retrospective study of 50 patients undergoing bicuspid aortic valvulopathy, collected in the Cardiology A department of the Ibn Sina University Hospital Center in Rabat, over a period of 5 years between January 2012 and December 2016. The census of these patients and the collection of the intraoperative data were carried out on the basis of the operative reports. Pre- and post-operative data were collected from the archived medical records.

RESULTS

Over a period of 5 years we have identified 50 cases of aortic bicuspidia, corresponding to a frequency of 10 cases per year. The average age of our patients was 56.02 years with extremes ranging from 7 years to 77 years? The male sex was predominant, with 37 men (74%) and 13 women (26%) with a sex ratio of 2.8. Among cardio-vascular risk factors, smoking was more commonly associated with aortic bicuspidia (40%), followed by hypertension (28%). Dyslipidemia was found in 16% of cases, 14% of our patients were diabetic, whereas the obesity of android was found only in 4 patients (8%).The most frequent reason for consultation was dyspnea, found in 45 patients, ie 90% of the cases. We evaluated the dyspnea of these patients according to the functional classes of the NYHA.

Other signs were exercise angina (42%), syncope (10%) and lipothymia (12%), palpitations (6%), and 4 patients (8%). in overall cardiac decompensation. Aortic bicuspidia following a febrile syndrome, associated with infectious endocarditis, was found in two patients (10%). The etiological assessment of malignant hypertension in a 20-year-old presence of aortic bicuspidia associated with coarctation of the aorta. One patient was diagnosed with acute coronary syndrome and one with cyanosis in relation to a Fallot tetralogy. In 2 patients, the discovery of cardiopathy was fortuitous during a consultation for bronchitis.

Cardiac auscultation revealed the presence of a rough rough systolic murmur in 86% of the cases with aortic B2 ablative in 16 patients (37%), decreased in 5 patients (12%), preserved in 5 patients patients (12%) and unspecified in 17 patients (39%). A diastolic blast at the same focus was found in 2 patients (4%) and a systolic-diastolic murmur in 6% of the cases. On the other hand, cardiac auscultation was normal in 2 patients (4%).A 12-lead electrocardiogram was performed in all of our patients: 44 patients had a regular sinus rhythm (88%) and 6 patients were ACFA (12%).

All patients in our series received an echocardiographic examination, which led to the diagnosis of BA in 24 patients (48%). The aortic sigmoids were calcified and reshaped in 76% of the cases and dysplastic in 4% of the cases. Aortic stenosis

was predominant (74%), followed by aortic disease (18%), whereas pure aortic insufficiency was only 8%.

Aortic bicuspidia was associated with dilatation of the ascending aorta in 18% of cases (Figure 1), interventricular communication in one patient, and Fallot tetralogy in another patient. The left ventricle was hypertrophied in 82% of cases and dilated in 18% of cases. Overall left ventricular function failure was found in 30% of patients with 2 patients with severely impaired LVEF, while the vast majority (70%) had LVEF> 50%.

In our work, the determination of the different types of valve according to the classification proposed by Sievers and Schmidtke was only made for 23 patients, with the majority of type 1 (70%) followed by type 0 valves (30%), whereas no type 2 patients are present. Thoracic angiography was performed in two patients following the discovery of an ascending aortic aneurysm on transthoracic ultrasound (Figure 2).

Surgical management was performed in all our patients; 96% of our patients had aortic valvular replacement in 73% of the cases and a bioprosthesis in 27% of the cases. Open heart commissurotomy was performed in 1 patient (2%). And an aortic annuloplasty was performed in one patient (2%). The result was found to be satisfactory in the vast majority of cases (98%), whereas the aortic prosthesis was stenosing in a single patient. The evaluation of the aortic leak found 2 patients in grade I, and only one patient in grade II.

In our series, there was only one death in a 77-year-old patient who underwent combined surgery: double aorto-coronary bypass surgery with aortic valvular replacement. The total postoperative hospital stay was 15 days on average with a minimum duration of 10 days and a maximum of 44 days.

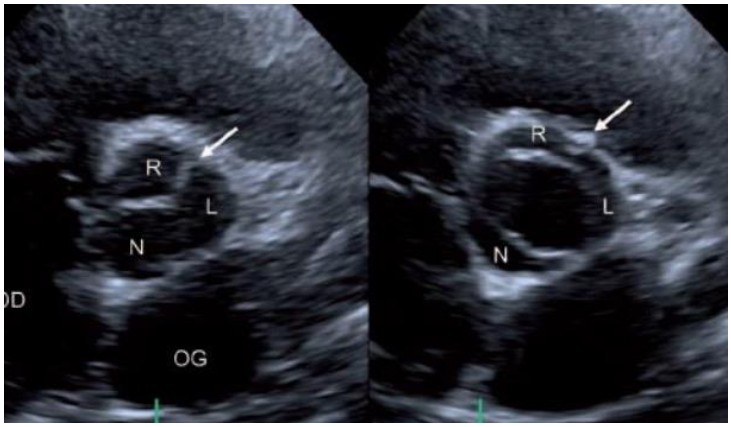

Fig-1: bicuspidia type 1 (1 raphe) "fusing" the coronary sigmoids (right and left).

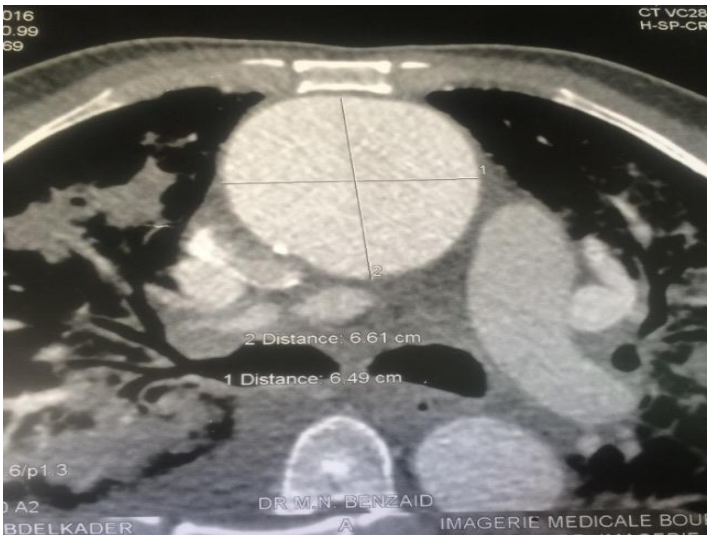

Fig-2: Scannographic image showing a fusiform ectasia of the ascending aorta measuring 66x65 mm.

DISCUSSION

The method of retrospective recruitment of our patients described above does not make it possible to evaluate the prevalence of this anomaly in our population. However, many autopsy studies from the years 1920 to 2000 estimated that 0.5-2.2% of the world population has BVA [4]. Our work found a clear male predominance of 74%, which agrees with the literature [5-8].

The mode of discovery of BA is variable; it can be discovered in a fortuitous way by the discovery of a breath on clinical examination during a routine consultation, which was the case of 2 patients in our series, or then diagnosed at the screening based on a family history. It may also be revealed by complications, or by associated malformations, or during an echocardiography performed for any other reason [9].

The cardiac auscultation, can perceive a protoseosystolic blast quite gentle ejection at the base, often as well audible in the second left parasternal intercostal space as in the aortic focus. This breath has an early peak and an intensity of 1-3 / 6th; it irradiates towards the vessels of the neck, but also frequently towards the point, in a scarf, as in the real aortic constrictions. There is never any shivering at the sternal range. Sometimes there is no breath [10], and a protosystolic ejection is very common.

Transthoracic echocardiography represents the essential examination for the diagnosis of aortic bicuspidia and the detection of ascending aortic anomalies [53]. However, although its specificity is excellent (close to 95%), its sensitivity appears much worse (about 80%) [11].

Transesophageal ultrasound provides a more precise and therefore more reliable imaging, particularly in the analysis of commissures, valves (size, orientation) and raphe, but its realization must not be systematic [12,13]. As regards therapeutic management, the use of pharmacological agents is limited because they rarely prevent the progression of most complications. A surgical procedure involving the aortic valve and, if necessary, the aorta is necessary in the majority of patients when the complications become too severe and their quality of life is affected.

CONCLUSION

This descriptive study of aortic bicuspidia showed that most patients did not develop symptoms before adulthood and did not require surgical management before advanced age. There are nevertheless early forms which have required care from early childhood. The risk of complication is real, whether at the level of the valve (leakage, stenosis, endocarditis) or the aorta (aneurysm). The association with other risk factors must be sought and treated (including hypertension and dyslipidemias). The different anatomical forms must be identified in view of a variable evolutionary potential depending on the anatomy. Finally, the management of narrowing or aortic insufficiency does not differ from that of patients without valvular congenital anomaly.

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