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RESEARCH ARTICLE

APICAL ANEURYSM OF THE LEFTVENTRICLE: THE HIDDEN FACE OF HYPERTROPHICCARDIOMYOPATHY (THROUGH A SERIES OF 207 CASES)

Fikrat F.Z, Hdioud O, Doghmi N. and Cherti M.

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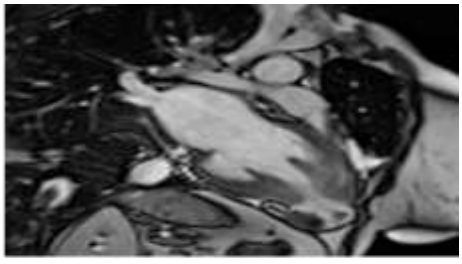
Abstract

Introduction:-
A group of patients withhypertrophiccardiomyopathy (HCM) develop an apical aneurysm of the LV (LV AA), leading to seriouscardiovascular complications. The prevalence of occurrence isunderestimated by echocardiography. Cardiacmagneticresonanceimaging (MRI) has recentlyallowed the individualization of theseaneurysmallesions.

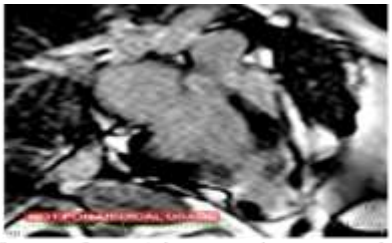
Methods:-
In thisstudy, weevaluated the incidence, clinical course, and MRI semiology of Moroccan HCM patients with LV AA.

Weretrospectivelyanalyzed 207 HCM casesrecruited and evaluatedfrom 2014 to 2021 at 2 centers, 10 of whichwereidentifiedwith LV apical aneurysms.

Results:-
Apical aneurysms of the LV wereidentifiedin 10 patients from a population of 207 CMH of all types: aprevalence of 5%, a meanage of 51.3 years, with a male predominance.Our patients weremostlysymptomatic: palpitations 40%, atypical pain 30%, one patient presentedaloss of consciousnesslike syncope. Three patients out of 10 weredyspneic stage II (NYHA).Electricalabnormalitieswerenoted in all patients: rhythmdisorders, particularlyventricularin 45% of cases, a Q wave of pseudo-necrosisin 30% of cases and an electrical LVH withsecondaryrepolarizationdisorder in the remaining patients.The diagnosis of apical aneurysmwas made by MRI in 8 out of 10 patients. 2 cases of aneurysmsdiscovered on TTE wereconfirmed on MRI.



Coupe 2 cavités montrant une hypertrophie apicale et un AA avec thrombus de la pointe



Coupe 2 cavités en séquences de rehaussement tardif montrant des foyers de fibrose au niveau de l'apex et un AA avec un thrombus apical

Corresponding Author:- Fikrat F.Z

Discussion:-

HCM is a geneticdisordercharacterized by aparticularlyheterogeneousmorphological and clinical expression.

A subgroup of these patients but not negligibledevelopsaleftventricular apical aneurysmleading to seriouscardiovascular complications including: Thromboembolic, arrhythmia, advancedheartfailure, suddendeath.

The mechanismresponsible for the formation of theseaneurysms in HCM patients remainsunexplained, althoughseveralpotentialmechanisms have been suggested, such as increased LV wall stress resultingfrommid-cavity LV obstruction and elevatedsystemic intra-cavity pressure leading to chronicmyocardialischemia, intra myocardial IVA bridging, and geneticpredisposition.

To clarify the clinical and paraclinicalpresentation, especially on cardiac MRI, as well as the prognosticelements of this subgroup of patients, we have compiled a series to gain insight intothisenigmaticentity in a sample of NorthAfrican patients.

Epidemyology

In ourseries the prevalence of LVAA was 5%, with 10 cases diagnosed in a cohort of 207 HCM patients.

This is in line with the literature with a similarprevalence in the mostrecent American series by Rowin et al, and exceedsthat of Maronfrom 2008.

The meanage of our patients at the time of discovery of the leftventricular apical aneurysmwas 51 years, in the literature an agebetween 50 and 60 years has been reported.

Male predominancewas the rule in all studies.

Clinical

The clinical expression of the diseaseis multiple and highly variable from one patient to another.

If the mostrecent studies show that patients are mostoftenasymptomatic or paucisymptomatic in classical HCM, in ourstudy and through a global literaturereview, we note that the majority of patients, arrive at the stage of aneurysm, withalready a symptom, whichis none otherthan the complication of the aneurysm or rarelywithin the framework of the naturalevolution of HCM.

Rhytmdisorders

Rhythmdisorders, especiallyventricularones, are found in more than 40% of our patients, whichis in line with the literature.

It ismainly NSVT, polymorphic VT or even VF that are behind the suddendeath of these patients. Theyfindtheiranatomical substratum in the junction zone between the healthymyocardial tissue and the aneurysmalwallwhere the RT shows transmuralfibrosis on cardiac MRI. Hence the interest of abetter MRI evaluation and thus a possible prophylactic ICD implantation.

In Rowin'sseries, the suddendeath rate was 4.7%/year, including: suddendeath cases, successfullyresuscitatedcardiacarrests.

Thromboembolic complications

The dyskinetic or akineticaneurysmalpocketcanconstitute the bed for thrombus formation with all the thromboembolic complications thatfollow. MRI demonstrated the presence of a thrombus within an aneurysmin 3 patients in ourseries.

In Maron'sseries, 4 thrombiwerefound, 2 of whichpresentedseriousthromboembolicevents; this opens the debate on the need for possible prophylactic anticoagulation in theseaneurysmal patients. Moreover, in Rowin'sseries, 5 patients whowere not anticoagulatedhad a thromboembolicevent,whereas 13 patients whohad an apical thrombus of the LV and were on anticoagulants did not have an embolicevent.

Heartfailure
10 to 15% of HCM patients developheartfailurewithpreserved LVEF. Of these, 3% reach end-stage heartfailurewithsystolicdysfunction and ventricularremodelingthatmayultimatelymimic DCF. These patients maybecome candidates for heart transplantation as a last resort.

Note that, in the literature, this rate issignificantlyhigher in the subpopulation of patients with an LV aneurysm (25%).

This findingsuggeststhat an adverse LV remodelingprocess, responsible for myocardialfibrosis and impairedsystolicfunction, maybecommon to patients with apical aneurysms and thosewith end-stage HCM.

Therefore, these observations underscore the importance of close monitoring in HCM patients with apical LV aneurysms to detectdeterioration of LV functionearly, whichmayallowimplementation of appropriatetargeted management strategies, such as afterload-reducing agents, cardioprotectivetherapies, ICD implantation possibly, and evenheart transplantation.

Transthoracicechocardiography
In our series, echocardiographyallowed the diagnosis of AA in 2 cases/10. It is an essential examination in the overall management of HCM and isrecommended as the initial examination. However, I tmayfail to visualize an apical aneurysmbecause of severalfactors, including size and location, poorquality images of the apical window. Recommendations call for additionalcardiac MRI in the presence of HCM thatisdifficult to analyze on ultrasound, if differential diagnoses are possible (eg, cardiacamyloidosis), or if therapeutic management maybemodified by the MRI findings (beforemyomectomy or septalalcoholization, ICD).

Cardiac MRI
Cardiac MRI providesa lot of information in a patient with or suspected of having HCM, from a precisemorphologicalevaluation to the search for differential diagnoses and the characterization of myocardial tissue. It isbecoming more and more essential in the assessment of HCM.

MRI issuperior to transthoracicechocardiography in detecting and analyzing focal apical and anteroseptalforms and in detecting apical aneurysms.

Moreover, in ourstudy, transthoracicechocardiographyrevealedonly 2 out of 10 AALVs. In Maron'sseries, MRI was more sensitive in detectingtheseaneurysms, and TTE was able to identifythemin 57% of cases.

Study of the Late Rise
Recentstudies have focused on the RT of gadolinium in cardiac MRI as a marker of fibrosis as an independentrisk factor. The latter isconsidered an arrhythmogenicsubstrate and a major player in promoting the development of heartfailure.

A meta-analysissuggestedthat the presence of RT (in 2/3 of cases) isassociatedwithcardiacdeathheartfailure and a tendency for suddendeath A retrospectivestudy of 1293 patients showedthat RT >15% of LV mass wasassociatedwith a twofoldincreasedrisk of suddencardiacdeath, suggestingthat the amount of RT ratherthanitsmerepresencecouldbe an important risk marker.

Electrocardiogram
The ECG is normal in only 5-10% of patients.

The mostfrequentabnormalities in classical HCM are signs of LVH withsecondaryrepolarizationdisorders. Withpathological Q waves of pseudo-necrosis in the inferolateral leads, corresponding to depolarization of the hypertrophicinterventricular septum.

Regardingcomplicatedforms of AA, Masaru in hispaperreleasedin 2014 showed, through a seriesthat (66%) of patients with AA presentedwith ST-segment elevation

Otherpapersdemonstratedthat the ECG of these patients describedconvex ST-segment elevationfrom V1 to V4, T-wave inversion without ST-segment elevationfrom V1 to V4 and loss of giant T-wavenegativity.

The use of ST-segment elevationwould have a sensitivity of 66.7% and a specificity of 98.7%.

Recommandations:-

According to the European Society of Cardiologyrecommendations, routine ICD implantation is not recommended in the absence of clinicalevidence of an increasedrisk of suddendeath. In contrast, the 2020 recommendations of the American Society of Cardiologyrecommend ICD implantation in HCM patients with an LV AA found on ultrasound or MRI, regardless of its size.

Conclusion:-

The presence of apical aneurysms in patients with HCM is not uncommon. Echocardiography most often underestimates their presence, hence the value of MRI coupled with late enhancement.

Diagnosis is important to prevent thromboembolic complications and the risk of sudden arrhythmic death. It must be made early in order to modify the prognosis, through the initiation of anticoagulant treatment or the implantation of an ICD.