



Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych”

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CASE SUMMARY

The presented case is the young adult with coronary circulation anomaly, that is giant left circumflex artery draining to the coronary sinus. He has positive cardiac family history and was directed for cardiological work-up after new-appearance of left bundle branch block (LBBB) was found on the ECG. The baseline echocardiogram revealed mildly depressed left ventricular systolic function and concomitant mild/moderate mitral regurgitation (MR). Later he underwent symptom-limited dobutamine stress echo that revealed significant improvement of systolic function and no changes in the degree of MR. As a result, the patient was scheduled for planned coronarography that showed aforementioned circumflex artery anomaly. During the same procedure, the right heart catheterization was performed, that demonstrated significant left-to-right shunt due to anomalous artery. To broaden the diagnostic picture, the angio computer tomography was performed, which confirmed the previous findings and nicely demonstrated the pathology.

DISCUSSION

There are three categories of coronary circulation anomalies. Firstly, it is inappropriate origin of the coronary artery. Secondly, the passage of the artery may be abnormal, and lastly the final portion of the artery or draining of the artery can be anomalous. In the presented, it is mostly the draining of the artery to the coronary sinus and eventually to the right atrium that is pathological. Although the dilated or even aneurysmatic arterial wall is also not normal. There are to consequences of this pathology. Firstly, it causes hypoperfusion of the relatively large area of the myocardium that is probably the reason of depressed systolic function at rest and some improvement during adrenergic stimulation. Moreover, it cannot be ruled out the depressed of ventricular function is responsible for concomitant MR. Secondly, the volume-overloaded dilated circumflex artery is a source of significant left-to-right shunt that may have future consequences.

Coronary fistulas are frequent and may vary from very small, multiple to very extensive fistulas (as in this case). Fistulas can cause significant under-perfusion of the large



cardiac territories. The distribution of the fistulas is almost evenly divided between left and right coronary artery. In half of the cases the fistula drains to the right ventricle, one-quarter to the right atrium, followed by the pulmonary artery, left atrium and finally the left ventricle.

EXPERT'S OPINION

Giving the fact that presented coronary anomaly has already caused several problems and there are probably even more to come if the pathology is left unchanged, the decision is relatively straightforward. Not going into the technical aspects of the operation, the surgical operation should be recommended as there is no chance that pathology will reverse by itself and most likely it will progress. Two aspects are in jeopardy, the under-perfused myocardium may dilate and irreversibly lose contractility that will lead to overt systolic heart failure. The other problem is close to significant left-to-right shunt that may cause pulmonary hypertension.

CONCLUSION

The bulk of the evidence strongly suggest the need for surgical correction of the coronary pathology. The open question remains whether concomitant MR should be also corrected by the surgeon. Probably the closure of the fistula will be enough as MR is not an organic valve disease but secondary to under-perfusion of the myocardium, so called "ischemic" MR. Although the applicability of the intra-operative transesophageal echocardiography may be questioned, as it will be probably some time before full restoration of myocardial perfusion will be achieved, nevertheless it may be worth trying.

REFERENCES

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