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307 A CARDIAC MASS IN A PATIENT WITH GORLIN-GOLTZ SYNDROME: INCIDENTAL DIAGNOSIS OR SOMETHING TO LOOK FOR?

Roberto Tarantini[a], Stefano Alonge[a], Lorenzo Acone[a], Marco Foti[a], Evelina Toscano[b], Daniele Poggio[b], and Andrea Mortara[b]
[a]Università Di Pavia, Pavia; and [b]Policlinico Di Monza, Dipartimento Di Cardiologia Clinica, Monza

Aims: Gorlin-Goltz syndrome or nevoid basal cell carcinoma syndrome (NBCCS) is a rare autosomal dominant, multisystem, tumor-predisposing disorder. The primary manifestation of NBCCS is the development of multiple basal cell carcinomas (BCCs) but is also associated with a variety of other benign and malignant tumors. Cardiac fibromas are increased in frequency in patients with NBCCS, developing in approximately 3 percent of affected individuals. They typically present in infancy. They are benign growths, and almost all develop within the ventricular myocardium. Although usually asymptomatic, they can result in impaired left ventricular function and conduction defects, necessitating resection.
Methods and results: we presented a case of a 59-years-old woman with a diagnosis of Gorlin-Goltz syndrome and no history of cardiovascular diseases who underwent a right knee arthroplasty in our hospital. After the surgery the patient was admitted to rehabilitation department. Electrocardiogram that showed sinusal rhythm with inverted Twaves in lateral leads and a transthoracic echocardiography examination were performed. Echocardiography showed a large mass within the basal and middle segments of left ventricle lateral wall. Left ventricular ejection fraction was preserved. Cardiac magnetic resonance (CMR) revealed an intramyocardial mass with well-defined borders and several calcifications, hypointense on T1-weighted and T2-weighted images and delayed-contrast hyper-enhancement with hypoenhancing central cores. Based on these imaging features, the mass was suggestive of cardiac fibroma with calcifications. The patients was asymptomatic. Afterwards, a total body CTwas performed to exclude other tumours in other sites. This case was discussed with cardiac surgeons and since the patient was asymptomatic with no conduction abnormalities or heart failure signs, urgent surgical intervention was excluded. The patient was discharged with a 3 months follow-up.
Conclusion: our case highlights the importance of echocardiography evaluation before surgery in patients with genetic syndrome like Gorlin-Goltz syndrome. Last guidelines published in 2021 recommend that all patients with NBCCS should be screened with a cardiac ultrasound and if cardiac symptoms occur in a patient with NBCCS, a cardiac ultrasound should be repeated to exclude a late-onset cardiac tumour. Furthermore, CMR plays an important role in characterizing cardiac masses and determining the management.