Ventricular Tachycardia in a Patient with Gorlin Syndrome

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A 15-year-old boy with transient syncope was transferred to our hospital. Electrocardiogram showed ventricular tachycardia with right bundle-branch block morphology and right superior axis (Picture 1). The patient’s medical history was remarkable with surgical operations for craniostenosis, hydrocephalus, basal cell carcinoma of the eyelid and

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ameloblastoma performed at another hospital. After hospitalization, a gene examination revealed that he had Gorlin syndrome.

A T2-weighted image of cardiac magnetic resonance showed regional thickness of the posterior wall (Picture 2). There was a high signal mass in that area on a late gadolinium-enhanced image with an appearance and tissue characteristics consistent with fibroma (Picture 3).

Gorlin syndrome is a rare autosomal dominant disorder that is caused by a mutation of the human patched-1 gene. In this syndrome, neoplasms such as basal cell carcinoma, medulloblastoma, odontogenic cysts and ovarian tumor develop with age. Cardiac fibromas are reported to develop in approximately 3% of adults with Gorlin syndrome (1).

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Reference