

Gorlin-Goltz Syndrome

Ranjit Divakaran, FDS RCS* Lekshmi Ranjit, MFDS RCS** Bobby Joseph, FDS RCS***

Gorlin-Goltz Syndrome (GGS) is known as nevoid basal cell carcinoma syndrome (NBCCS); it is a rare condition with multi-organ involvement. It has an autosomal dominant trait with complete penetrance and variable expressivity. The condition presents with a wide range of pathological features including malignancy of the skin (basal cell carcinoma); its early diagnosis is vital. Odontogenic Keratocyst (OKC) being one of the prominent features and mostly an early one, the dental specialty most often is in a position to identify this condition first.

We present a case of GGS in a fourteen-year-old male. Enucleation was performed and the postoperative period was uneventful. Histopathologically, the diagnosis was confirmed as multiple Odontogenic Keratocyst. The patient had spina bifida at D2 vertebra, fused anterior end of right 5th and 6th ribs, fused right anterior 2nd and 3rd ribs, Falx and tentorial calcification and Sprengel shoulder. It is essential to emphasize the role of the dental specialty in diagnosing and instituting early treatment of such condition.
