GENETICS AND GENODERMATOSES

# DIAGNOSIS AND TREATMENT OF GORLIN-GOLTZ SYNDROME : ABOUT 12 CASES.

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Introduction: Gorlin-Goltz syndrome (GGS) was more clearly defined in 1960 by Gorlin and Goltz, is a genetic syndrome characterized by the eruption of multiple and early onset basal cell carcinomas (BCC).It is inherited in an autosomal dominant pattern.

Objective: From 12 observations, we specify the clinical and radiological characteristics of this syndrome and discuss the therapeutic means and their evolutionary aspects.

Materials and methods: This is a retrospective study of 12 observations collected in the department of oral and maxillo-facial surgery at CHU Sahloul, between 1993 and 2016.

Results: A total of 12 patients aged 4 to 64 years, were referred to our department .The sex ratio (F/H) was 1.In our study, the presence of multiple naevi of the face and trunk, maxillary keratocysts, and falx cerebral calcification led to the diagnosis of GGS for all the patients. Six family cases were found in our series. The principal cause preceding the GGS is the germline mutations of the PTCH gene. Therefore we choose a stepwise approach to manage such patients which include a preoperative biopsy to establish a definitive diagnosis and complete removal of all the lesions to prevent recurrence. A close clinical follow-up was recommended to detect any newly developed or recurrent lesions.

Conclusion: GGS is associated with multiple BCC and odontogenic keratocysts of the jaw at an early age as well as various anomalies such as bifid ribs, palmar and plantar pits, and ectopic calcification of falx cerebri. A multidisciplinary approach becomes necessary for the diagnosis and follow-up of patients with GGS, considering the complexity of the clinical manifestations. A family survey as well as a clinical and radiological assessment should be performed in search of other manifestations.