GENETICS AND GENODERMATOSES

GORLIN-GOLTZ SYNDROME: ODONTOGENIC KERATOCYSTS REVEALING AN OFTEN MISSED DIAGNOSIS

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Introduction: Multiple basal cell carcinomas (BCCs) are often linked to some genodermatosis, including Gorlin-Goltz syndrome (GGS). We report a series of 4 patients.

Objective: The importance of an early diagnosis of GGS is based on the initiation of a multidisciplinary follow-up in order to detect the complications such as multiple BCC and maxillofacial deformities.

Materials and methods: Of 33 consecutive patients with BCCs who consulted our department, between 2010 and 2018, 4 patients were diagnosed with GGS by correlating the clinical findings, histological findings,and evaluating the radiological exploration.

Results: A total of 4 male patients were referred to our department for BCCs on the face. The mean age at the detection of this syndrome was 51 years.All the patients had a history of sun exposure. In one case a history of tar exposure and a basal cell carcinoma of the penis, and the back, was noted. The clinical examination revealed palmar pits in one case. The most common subtypes were nodular followed by ulcerated and sclerodermaous lesions.Further investigations were carried out and showed in four patient mandibular keratocysts and calcifications of falx cerebri in one patient. All the lesions were surgically removed. The evolution was marked by the recurrence of the lesions and the appearance of new lesions in one case. BCCs are often a feature of genetic syndromes including GGS, xeroderma pigmentosum,Bazex syndrome, Muir-Torre syndrome and Rombo syndrome. GGS is a rare autosomal dominant disorder caused by mutations of the PTCH gene, that involves multiple organ systems including the skin. It is characterized by the eruption of multiple and early onset BCCs, multiple odontogenic keratocysts, bifid ribs, palmar and plantar pits and other abnormalities. It was first reported in 1894 by Jarisch and White. The treatment may be complicated due to the large number of basal cell carcinomas.

Conclusion: Considering the complexity of the clinical manifestations, a multidisciplinary approach becomes necessary for the diagnosis of patients with GGS.