Keratocystic odontogenic tumors as first clinical manifestation of nevoid basal cell carcinoma syndrome in pediatric age: our microinvasive surgical approach.

Aim. Nevoid Basal Cell Carcinoma Syndrome (NBCCS) is a rare genetic condition involving multiple organs; Keratocystic Odontogenic Tumors (KCOTs) are often the first clinical manifestation in pediatric age. The aim of this study was to describe the clinical and histopathological features of KCOTs as first clinical sign of NBCCS in pediatric patients allowing an early diagnosis, and their treatment with conservative microinvasive piezosurgery. Methods. Twenty pediatric patients affected by NBCCS showing 60 KCOTs came to our attention at the Complex Operating Unit of Odontostomatology, Policlinic of Bari, from 1996 to 2014. After clinical examination, OPT and computed tomography analysis, all patients underwent conservative micro-invasive surgical treatment under generalanesthesia, consisting in enucleation of KCOTs, cavity ostectomy and osteoplasty with conventional rotative instruments and piezoelectric tools in order to remove damaged bone, epithelial remnants and satellite cystswith maximal teeth preservation in consideration of the young age of the patients, and to possibly minimize therecurrence risk. A sterile gel formulation of sodium hyaluronate and amino acids (Gly-Pro-Leu-Lys) was put into the bone defect, allowing for faster bone regeneration and healing of the surgical site. All the surgical specimens were sent for histopathological examination. The patients underwent clinical and radiological (OPT) follow-up after 7, 15 and 30 days, 2, 3, 6 and 12 months, and, then, once a year. The followup time ranged from 18 months to 8 years. Results. There were 10 males and 10 females, with an average age of 10,6 years. At the initial evaluation, 13 patients showed swelling, teeth agenesis, and dental inclusions or dislocations as first clinical manifestation of their disease; in the remaining cases, KCOTs wereasymptomatic. Among the 20 patients, 11 had been previously diagnosed with NBCCS because of familiarityor presence of characteristic features of the syndrome, while KCOTs were the first clinical sign of NBCCS for the remaining 9 patients without familial history of the syndrome; subsequently, the genetic analysis showing PTCH1 mutations confirmed the diagnosis. 60 KCOTs were totally identified: 39 lesions were located in the mandible and 21 lesions were located on the upper jaw. The histopathological analyses highlighted thin connective tissue walls covered by para- and orthokeratotic stratified squamous epithelium, usually about 5-8 cell layers thick, around cystic lumens filled with desquamated keratin, thus confirming the diagnosis ofKCOT. The epithelial lining included a well-defined, palisading basal layer of cuboidal to small columnar cells and a superficial layer with corrugated appearance. Satellite cysts could also be seen due to tissue budding of the basal cell layers into the adjacent connective tissue. The clinico-radiological follow-up of 53 lesions showed progressively decreased radiolucent areas up to complete healing within 12 months from the surgical treatment. Differently, OPT disclosed lack of decreasing radiolucency of 7 lesions after 12 months suggestingKCOTs recurrence, that is more frequent in syndromic KCOTs than in sporadic ones due to their higher dimensions and to the involvement of nobile structures such as teeth which should be preserved in these young patients as much as possible.

Conclusion. KCOTs in pediatric patients require conservativeapproaches for permanent teeth preservation. While conventional enucleation leads up to 60% recurrence rates, cavity ostectomy with piezoelectric tools significantly reduced the recurrence risk allowing thepreservation of permanent teeth.