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Fibrous displasia of craniofacial area in children and adolescents

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Introduction: Fibrous dysplasia (FD) is benign medullary genetically-based sporadic fibro-osseous lesion, which may involve one or more bones and is the most common in children and adolescents, especially in the craniofacial area.
Methods: We analyzed 585 cases of bone tumors and tumor-like lesions in children and adolescents from 2009 to 2014. FD was found in 48 patients (8,2%), craniofacial localization was diagnosed in 30 children (62,5%). The specimens were examined grossly and fixed in 10% formalin. Paraffin embedded sections stained with haematoxylin and eosin and examined histologically. In 18 cases, we have performed immunohistochemical staining with MDM2 and cdk4 in order to differentiate FD from low-grade osteosarcoma. Initial diagnoses of all the cases were based on their clinical, radiological, and histological features.
Results: The age of most of the patients was from 7 to 18 years (25), 5 patients were younger age 7. The male/female ratio was 13/17. Most cases of FD occurred in the maxilla (14), and frontal bone and orbit (8). A few cases were localized in the mandible (3), temporal and occipital bones (3), ethmoid and nasal cavity (2). Two patients (6,6%) had multiple craniofacial lesions. The approximate time to treatment from the first symptoms may be from 1 month to 8 years, the mean time is 24,7 months. The main complaints were a painless swelling often leading to facial asymmetry, displacement of teeth and/or malocclusion. The size of the lesions varies from 13 to 70 mm in diameter. Radiographically FD has well-defined margins, produces ground-glass appearance without prominent periosteal reaction. Histological evaluation revealed extensive osteoblastic rimming of trabeculae and focuses of marked stromal hypercellularity in comparison with FD of other locations. FD has overlapping histological features with juvenile ossifying fibroma and giant cell reparative granuloma. Immunohistochemically all cases were negative for MDM2 and cdk4 except 1 case - slightly positive for cdk4.
Conclusion: The craniofacial FD is most common in children and adolescents. Histologically we revealed extensive osteoblastic rimming of trabeculae and marked stromal cellularity in comparison with FD of other sites. FD should be differentiated with low-grade osteosarcoma, juvenile ossifying fibroma, and giant cell reparative granuloma.