Low-grade, Well-differentiated Fibroblastic Osteosarcoma of the Mandible - A Diagnostic Challenge with Literature Review

Solaleh Shahmirzadi[1] DDS, Makoto Saigusa[2] DMD MD, Yi-Shing L. Cheng[1] DDS MS PhD, Hui Liang[1] DDS MS PhD

[1] Department of Diagnostic Sciences, Texas A&M University College of Dentistry

[2]Private Practice, Tyler Longview Oral Facial Surgery, Texas

Dr. Solaleh Shahmirzadi

s.shahmirzadi@tamu.edu

Telephone: 615-927-9640

Fax: 214-828-8306

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Purpose: Osteosarcoma (OS) of the jaw, in general, is a high-grade lesion. Low-grade osteosarcoma (LGOS), also known as intraosseous well-differentiated osteosarcoma is rare. Diagnostically its clinical and radiographic presentations do not correlate well with its histologic features and is often misinterpreted as a benign lesion. The aim of this study is to report and discuss a case of low-grade fibroblastic osteosarcoma of the mandible along with a review of the literature. The intent of this report is to increase awareness of this condition, which is challenging for diagnosis but essential for proper treatment.

Methods: A 6-year-old otherwise healthy female was referred for evaluation of a swelling in the anterior region of the mandible that had become increasingly larger over a period of 3 months. Clinically, she had significant facial and lingual bone expansion in the left anterior region with a missing left mandibular primary incisor and displaced primary mandibular lateral incisor and canine. No neurological deficit was detected. The initial panoramic radiograph revealed a poorly defined mixed radiopaque/radiolucent lesion with a ground-glass appearance in the median and left anterior regions of the mandible. The lesion displaced permanent tooth buds of the left incisor and canine inferiorly. Multidetector Computed Tomography (MDCT) also demonstrated a heterogeneous density lesion with left mandibular facial-lingual bony expansion and disruption of cortical plates. The aggressive radiographic features of the lesion were suggestive of a malignant disease.

Results: The initial incisional biopsy was made, and the diagnosis by a local pathologist was between benign fibro-osseous lesion and low-grade osteosarcoma. Subsequently a consultation of the biopsy specimen was requested to our institution. Histological examination demonstrated round or irregularly shaped osteoid-like calcifications of varying sizes in a cellular stroma with spindle or stellate shaped cells. These cells showed mild pleomorphism with rare mitoses. A finding of fibro-osseous lesion with features suggestive of well-differentiated, LGOS was made. A second incisional biopsy was submitted directly to our institution for evaluation. The histologic and immunohistochemical features were similar to those seen in the previous biopsy. As a result, an excision of the complete lesion was performed. The final diagnosis was consistent with low-grade, well-differentiated fibroblastic osteosarcoma. The clinical and postoperative imaging follow-up study showed no evidence of recurrence or metastasis.

Conclusions: Reviewing of the literature with ten reported cases, the diagnosis of LGOS remains a challenge because its histologic features are similar to those of benign fibro-osseous lesions such as fibrous dysplasia or cemento-ossifying fibroma. The radiographic appearance of LGOS is variable but demonstrates signs of aggressive behavior including a poorly defined margin, expansile, and cortical bone destruction. These radiographic features were present in this case. Cortical disruption and soft-tissue extension are common in MDCT and MR images for LGOS, which might help in differentiating it from a benign fibro-osseous lesion. Any question about the diagnosis of LGOS should be cause for referral for a second opinion because of the diagnostic challenge. Complete removal of the lesion and continued follow up is necessary because LGOS can recur and undergo transformation into high-grade osteosarcoma. The mean age of LGOS at presentation is in the 3rd or 4th decade of life. This case might be the youngest to be reported.