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Case Report

CLEAR CELL VARIANT OF AMELOBLASTIC CARCINOMA OF THE MAXILLA: A CASE REPORT AND REVIEW OF DIFFERENTIAL DIAGNOSTIC AND THERAPEUTIC APPROACH

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SUMMARY – Ameloblastic carcinoma is a rare malignant epithelial tumor, mostly affecting the mandible. Maxillary ameloblastic carcinoma is even more uncommon and its clear cell variant has been exceedingly rarely described. Because clear cell lesions of jaw bones are both rare and diagnostically challenging, the incidence of the tumor, as well as the criteria for classification are not precisely defined. Differential diagnosis includes a variety of benign or malignant tumors of odontogenic and salivary gland origin, as well as metastatic tumors. Diagnostic dilemma extends to therapeutic approach. Treatment modalities are still debated. The present case report discusses the clinical and histologic differential diagnosis of clear cell variant of an aggressive maxillary ameloblastic carcinoma which was successfully treated with preoperative radiation therapy, followed by surgical excision of the residual lesion and reconstruction of the maxilla.

Key words: *Ameloblastic; Clear-cell; Odontogenic; Carcinoma; Pathology; Radiotherapy*

Introduction

Clear cell lesions of jaw bones present a great differential diagnostic dilemma[1-4]. Histologic characterization of such lesions is challenging, and there are several differential diagnoses that must be excluded. Tumors with clear cells may originate from odontogenic and salivary gland tissue. Because metastatic carcinoma is the most common malignancy of the jaw, the diagnosis of a primary intraosseous carcinoma must always be made by exclusion of metastatic disease[5].

The most common malignant epithelial odontogenic tumors with a clear cell histologic pattern are clear cell odontogenic carcinoma and clear cell variant of ameloblastic carcinoma. Clear cell lesions of salivary gland origin are clear cell carcinoma, myoepithelial carcinoma and mucoepidermoid carcinoma. Renal cell carcinoma is the most probable among metastatic

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tumors with such a histologic pattern[2,5]. In the majority of cases affecting jaw bones, a painless swelling and loosening of teeth are the only complaints. Radiographic features reveal a radiolucent intraosseous lesion. Histologically, all these lesions have considerable morphologic and immunophenotypic overlap, which makes distinguishing between them difficult.

Because of the paucity of cases of ameloblastic carcinoma of the maxilla, treatment strategies described in the literature remain divisive. Surgery followed by radiotherapy seems to be the treatment of choice[2,4-6]. Preoperative radiotherapy can be used to treat some rapidly growing tumors before radical surgery[7-9].

This case report presents a patient who underwent preoperative radiation therapy due to rapid and aggressive growth of the tumor, resulting in successful reduction of the tumor mass. Surgical procedure followed.

Case Report

We present a case of an otherwise healthy 56-year-old male patient who was referred to our department due to swelling and loosening of the second right maxillary molar lasting for 5 days. Clinical examination revealed discreet swelling of the upper right vestibulum around the buccal roots of the single remaining right maxillary molar without fluctuation, while the tooth was loosened and painful to palpation and percussion. Routine orthopantomography revealed a relatively well delineated radiolucent lesion measuring 1 cm in diameter, surrounding roots of the second right maxillary molar, suggesting a radicular inflammatory process. Loss of bone next to the radices indicated an advanced periodontal disease. No infiltrative mass was observed in the maxillary sinus.

Extraction of the second right maxillary molar was performed. The apical part of the palatinal root was resorbed and embedded in a soft hemorrhagic mass. Tissue surrounding the roots was excochleated and histopathologically analyzed. Valsalva maneuver was performed after extraction and it was negative. Histologic examination revealed a tumor composed of alveolar nests of round cells with vesicular nuclei, more or less pronounced nucleoli and clear cytoplasm, surrounded by mostly delicate fibrovascular septa. Mitotic activity was high (Fig. 1).

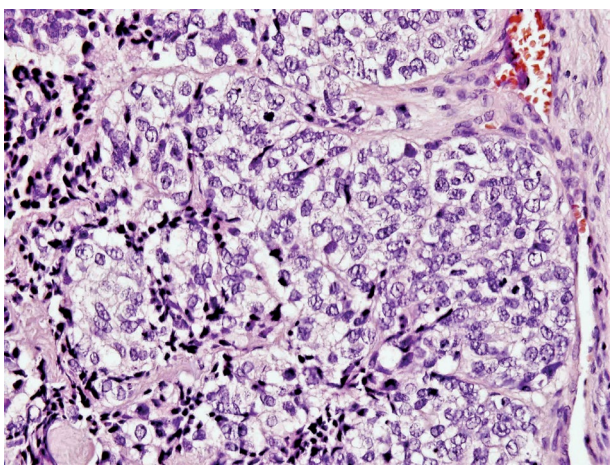

Fig. 1. Tumor composed of alveolar nests of round cells with vesicular nuclei, clear cytoplasm and high mitotic activity, surrounded by mostly delicate fibrovascular septa (hematoxylin-eosin X400).

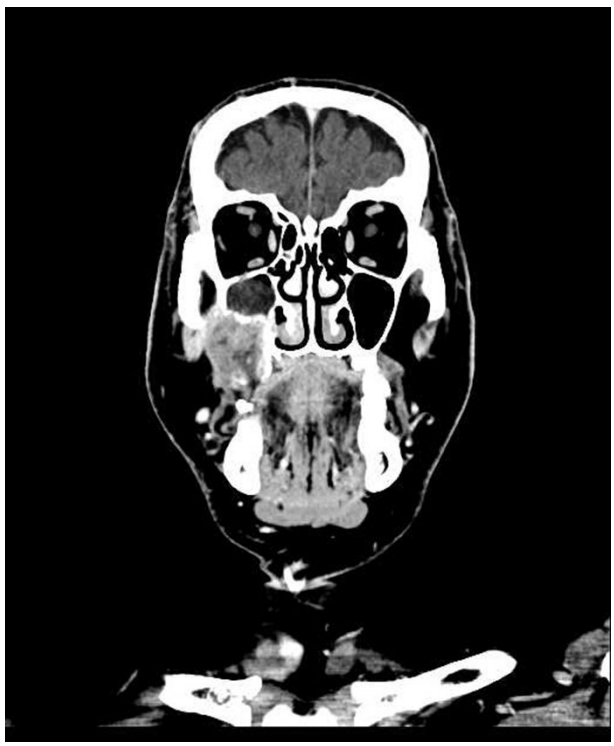

Fig. 2. Coronary multislice computed tomography scan revealing a tumor mass in the right maxilla expanding to the zygomatic bone and maxillary sinus.

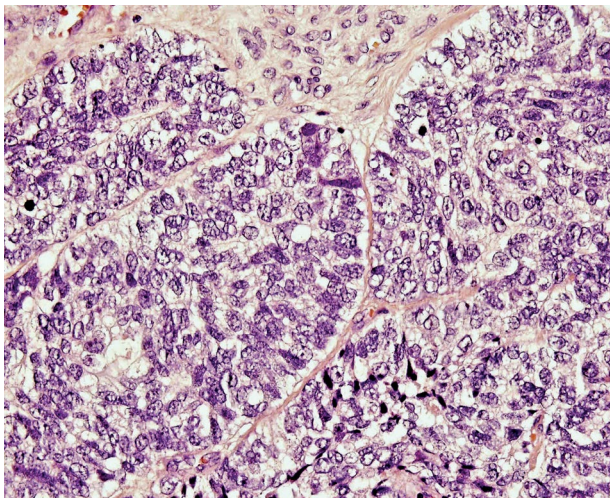

Fig. 3. Tumor nests with peripheral palisading, reverse nuclear polarity and subnuclear vacuolization (hematoxylin-eosin X400).

Immunohistochemical analysis revealed cytokeratin AE1/AE3, vimentin and CD10 positivity, therefore, the first consideration in our differential diagnosis was to exclude the possibility of a metastatic lesion such as renal cell carcinoma. Multislice computed tomography (MSCT) and positron emission tomography (PET) were performed. MSCT revealed a well circumscribed lytic lesion of the right maxilla expanding to the zygomatic bone and maxillary sinus, in a diameter of 3.5 cm (Fig. 2), and PET scan revealed no evidence of any other primary process.

Further material was obtained to more accurately define the exact nature of the lesion. Repeated biopsy revealed the same histologic pattern. The tumor was composed of large, polygonal atypical epithelial cells with mostly clear cytoplasm and brisk mitotic activity, and next to these, there were areas with more densely packed cells with elongated nuclei and less conspicuous cytoplasm. In some nests, there were central necrotic foci and peripheral palisading with reverse polarity and subnuclear vacuolization, suggesting ameloblastic differentiation (Fig. 3). Also, scattered tiny roundish, eosinophilic dentinoid or cement-like globula suggested an odontogenic origin of the tumor.

Additional immunohistochemical analysis revealed cytokeratin (CK) 19, p63 and CK 5/6 positive expression, while CK 7, CK 20, SMA, S100, p16, RCC, PAX 8, androgen receptor and thyroglobulin were all negative. Proliferative index Ki67 was more than 50%. Since metastatic carcinoma was excluded, the diagnosis of a primary intraosseous malignant tumor was confirmed. Molecular analysis was also performed. MAML2 and EWSR1 rearrangement and BRAF mutation were all negative. According to histologic pattern with focal necrosis and peripheral palisading, and immunophenotype of the tumor, definitive diagnosis was clear cell variant of ameloblastic carcinoma.

Due to extensive diagnostics, two months after the first MSCT, follow up MSCT was performed. Compared to the initial MSCT, the tumor was significantly larger, measuring 60 mm in diameter, expansive, osteodestructive, and spreading through the hard and soft palate into the oral cavity, and filled the right maxillary sinus destroying the right zygomatic bone. Because of the rapid, aggressive growth of the tumor, the patient underwent primary radiotherapy with 70 Gy delivered in 35 fractions. During radiation therapy, the tumor showed excellent signs of regression, which was confirmed by follow up MSCT (Fig. 4). The tumor regressed, resulting in a necrotic lesion with calcified and fibrotic borders.

Surgical treatment proceded with subtotal maxillectomy with resection of the maxillary process of the zygomatic bone. Reconstruction of the defect was

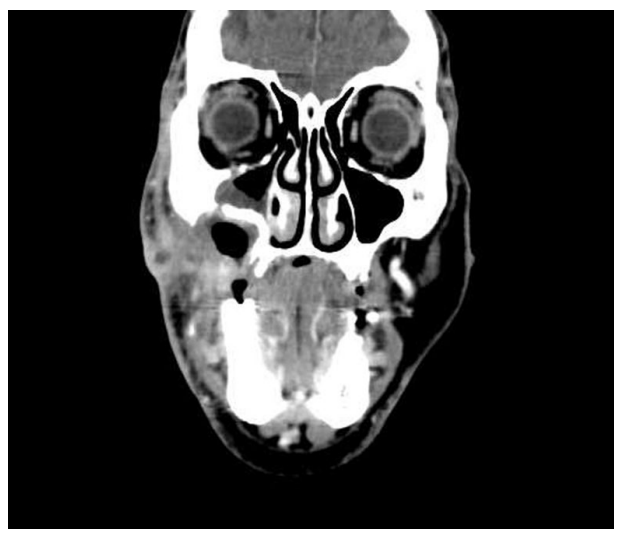

Fig. 4. Multislice computed tomography scan after radiation therapy showing significant tumor regression and necrotic cavitation with calcified and fibrotic borders.

performed with fibula osteocutaneous free flap. Histopathology of the surgical specimen revealed a cavity in the maxilla covered by necrotic debris, and within the fibrous lining of the cavity there were large myxoid pools with several small islands of large polymorphous epithelial cells with squamous and mucinous differentiation. Surgical borders were wide, with no evidence of tumor.

To date, 30 months after radiation therapy and surgical reconstruction, the patient is alive and well, with no evidence of recurrent or metastatic disease.

Discussion

Epithelial tumors affecting jaw bones are extremely rare. They may originate in various odontogenic or salivary gland lesions, whether benign or malignant. Epithelium available for tumorigenesis in the jaws are remnants of dental lamina, reduced enamel epithelium, rests of Malassez, and lining of odontogenic cysts, as well as intraoral minor salivary glands developed from the basal layer of oral mucosa[5-9].

Odontogenic tumors are rare, and malignant tumors of odontogenic origin are even rarer. Malignant intraosseous epithelial tumors are most frequently metastases; therefore, the diagnosis of primary intraosseous carcinoma must be made by exclusion of other tumors[5,10].

Metastatic neoplasms with clear cells most probably originate from renal cell carcinoma and less likely from thyroid carcinoma, sinonasal renal cell-like adenocarcinoma, balloon-cell melanoma, or leukemia[2,5,9,11,12]. Immunohistochemical findings may help distinguish them from primary tumors. In the present case, considering cytokeratin, CD10 and vimentin positivity, our primary concern was exclusion of a metastastic renal cell carcinoma. Radiographic analysis, histologic pattern lacking stromal hyalinization and stromal vascularity, and additional immunohistochemistry ruled out metastasis.

Salivary gland neoplasms with a clear cell appearance would include mucoepidermoid carcinoma, clear cell myoepithelioma, clear cell myoepithelial carcinoma, clear cell oncocytoma, and clear cell acinic cell carcinoma[3,13-15].

Despite immunohistochemical positivity with myoepithelial markers (CD10, p63), histologic pattern (lack of squamous, intermediate and mucinous cells) and other immunohistochemical findings were not consistent with any salivary gland tumor. Molecular analysis of CTRC1/MAML2 translocation suggesting a central mucoepidermoid carcinoma[15], EWSR1/ATF1 translocation suggesting clear cell odontogenic carcinoma (CCOC) of salivary gland origin[16,17], and BRAF V600 mutation suggesting ameloblastic origin of the tumor[18] were all negative.

Histologic pattern revealing conspicuous clearing of cytoplasm, peripheral palisading with reverse polarity and subnuclear vacuolization, as well as dentinoid or cement-like globula strongly suggested an odontogenic lesion.

Malignant epithelial neoplasms of odontogenic origin include ameloblastic carcinoma, primary intraosseous squamous cell carcinoma, CCOC, sclerosing odontogenic carcinoma, and ghost cell odontogenic carcinoma[5,19]. Review of the literature revealed discordant conclusions regarding classification of tumors with ameloblastomatous differentiation and clear cell appearance. Many authors consider CCOC a clear cell subtype of the ameloblastic carcinoma[9,17,29,30], some of the authors agree that these tumors should be called clear cell odontogenic carcinomas, even if they show occasional ameloblastoma-like histologic patterns[6,31-33], whereas others suggest that they should be regarded as different entities[15,28,34]. Since 2005, these tumors have been officially accepted as separate entities in the World Health Organization (WHO) classification of odontogenic tumors. The latest WHO Classification of Head and Neck Tumors[28], edited in 2017, defines ameloblastic carcinoma as a unique odontogenic carcinoma which combines histologic characteristics of ameloblastoma and varying degrees of cytologic atypia. Ameloblastic carcinoma aggressiveness, particularly when arising in the maxilla, is what primarily separates it from CCOC and therefore, ameloblastic carcinoma requires a more aggressive therapeutic approach. Histologic and immunohistochemical differences are minimal, but molecular analysis confirming EWSR1-ATF1 translocation suggests CCOC. Ameloblastic carcinoma may arise *de novo* or by malignant transformation of pre-existing odontogenic lesions[22,24]. In a review of 31 studies published between 2005 and 2011, Casaroto *et al.*[25] found a high incidence of primary ameloblastic carcinoma in the maxilla, while

secondary ameloblastic carcinoma was more frequent in the mandible. The higher frequency of the primary type in the maxilla and the more aggressive behavior of the secondary type are confirmed by other studies[26].

The presence of many clear cells strongly suggests an ameloblastic carcinoma[9]. At present, there is no single definitive microscopic criterion for ameloblastic carcinoma, so the diagnosis is based mainly on anatomic considerations or histomorphologic similarities among some tumors with odontogenic structures.

As ameloblastic carcinomas are rare, locally aggressive neoplasms and inappropriate treatment may result in multiple recurrences and metastatic spread[24,27]. Moro *et al.*[24] report that the majority of recurrences were in cases of posterior tumor location (80%). In 2007, Hall *et al.*[9] revealed that patients with clear cell ameloblastic carcinoma had two or more recurrences of tumor almost twice as often as the non-clear cell group. Each recurrence seemed to decrease the probability that the patient would be cured, even though the tumors did not noticeably change their histologic expression between treatments. The average time between treatment and recurrence was 25 months. Patients who died of the tumor or died with the tumor had multiple recurrences and treatments. Seventy-five percent of the patients who died with tumor in their study had the clear cell type of ameloblastic carcinoma. These data support consideration of early and definitive treatment for ameloblastic carcinoma.

Because of its rarity, there is still no consensus on their treatment. All authors agree that early, aggressive and complete removal of the tumor offers the best chance of survival.

Radiotherapy definitely has a role in the treatment of ameloblastic carcinoma, but indications for its use are disputed[6,8,28]. It has been speculated that neoadjuvant radiotherapy could assist in shrinking the lesion[28]. In a series of four patients with ameloblastic carcinoma treated with radiotherapy, Philip *et al.*[26] found that the disease remained fairly locally contained after treatment without further spreading. Primary radiotherapy was expected to be useful in inoperable cases, primarily in the posterior maxilla, in cases with perineural or massive soft tissue invasion, and in cases with positive surgical margins[7,8,30]. Dhir *et al.*[7] reviewed 18 patients with maxillary ameloblastic carcinoma. In 11 of 18 patients, radiotherapy was used either as primary or secondary treatment in the case of metastasis and/or recurrence. In 2011, Jensen *et al.*[31] introduced carbon ion therapy for multiple recurring ameloblastic carcinoma. Stereotactic radiosurgery (SRS) in the treatment of ameloblastic carcinoma has been recently reported[32]. Takahashi *et al.*[33] report on the efficacy of single fraction helical tomotherapy for the treatment of residual ameloblastic carcinoma following surgical resection, remarking that SRS may be an effective treatment, but only for small volumes due to the high doses used in radiosurgery[26,32,33]. The role of chemotherapy has not yet been proven[17,34,35].

In our case, due to the rapid, aggressive growth of the tumor, primary radiotherapy was performed achieving an excellent result. The tumor regressed, resulting in a necrotic, cystic lesion with calcified and fibrotic borders. Surgical treatment proceded with partial maxillectomy. Regression of the tumor enabled complete surgical resection with free margins.

Conclusion

Ameloblastic carcinoma is a very rare odontogenic neoplasm. Differential diagnostics are demanding, and treatment modalities are still disputed. According to our experience and review of the literature, preoperative radiotherapy of such an aggressive and rapidly spreading tumor greatly increases the chances of a positive outcome.

Because relatively few cases of maxillary ameloblastic carcinoma have been reported, the authors suggest that additional studies and reports of treatment experience with this tumor are necessary to disclose more useful treatment and management strategies in the future.

References

1. Loyola AM, Cardoso SV, Faria PR, *et al.* Clear cell odontogenic carcinoma: report of seven new cases and systematic review of the current knowledge. Oral Surg Oral Med Oral Pathol Oral Radiol. 2015;120:483-96. doi: 10.1016/j.oooo.2015.06.005
2. Maiorano E, Altini M, Viale G, Piattelli A, Favia G. Clear cell odontogenic carcinoma. Report of two cases and review

of the literature. Am J Clin Pathol. 2001;116(1):107-14. doi: 10.1309/WUQ8-D3A8-YV1Y-0VTR

3. Bilodeau EA, Hoschar AP, Barnes EL, Hunt JL, Seethala RR. Clear cell carcinoma and clear cell odontogenic carcinoma: a comparative clinicopathologic and immunohistochemical study. Head Neck Pathol. 2011;5:101-7. doi: 10.1007/s12105-011-0244-4
4. Memtsa PT, Papadopoulou A, Vahtsevanos K, Tzitzikas I. Management of clear cell odontogenic carcinoma: a case report. J Med Cases. 2018;9(7):215-7. doi:https://doi.org/10.14740/jmc3080wM
5. Eversole LR. Malignant epithelial odontogenic tumors. Semin Diagn Pathol. 1999;16:317-24. PMID:10587275
6. Perumal CJ. Ameloblastic carcinoma of the maxilla with extension into the ethmoidal air cells and close proximity to the anterior skull base: a rare case presentation. Craniomaxillofac Trauma Reconstr. 2012;05(03):169-74. doi: 10.1055/s-0032-1322531
7. Dhir K, Sciubba J, Tufano RP. Ameloblastic carcinoma of the maxilla. Oral Oncol. 2003;39:736-41. doi: 10.4103/0975-5950.102169
8. Gardner DG. Radiotherapy in the treatment of ameloblastoma. Int J Oral Maxillofac Surg. 1988;17:201-5. PMID: 3135350
9. Hall JM, Weathers DR, Unni KK. Ameloblastic carcinoma: an analysis of 14 cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2007;103:799-807. doi: 10.1016/j.tripleo.2006.11.048
10. Perić B, Vidaković B, Grgić N, Manojlović S, Čavka H. Angiomyoma of the upper lip – case report and review of the literature. Acta Clin Croat. 2019;58(1):183-6. doi: 10.20471/acc.2019.58.01.24
11. Kopić V, Barbić J, Petrović S, *et al*. Periodontal disease in different stages of chronic kidney disease. Acta Clin Croat. 2019;58(4):709-15. doi: 10.20471/acc.2019.58.04.18
12. Vučićević Boras V, Vidović Juras D, Aurer I, Bašić-Kinda S, Mikulić M. Gingival ulcerations in a patient with acute myeloid leukemia: a case report and literature review. Acta Clin Croat. 2019;58(3):556-60. doi: 10.20471/acc.2019.58.03.23
13. Yamamoto H, Inui M, Mori A, Tagawa T. Clear cell odontogenic carcinoma: a case report and literature review of odontogenic tumors with clear cells. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1998;86:86-9. https://doi.org/10.1016/S1079-2104(98)90155-X
14. Li Y, Li L-J, Huang J, *et al*. Central malignant salivary gland tumors of the jaw: retrospective clinical analysis of 22 cases. J Oral Maxillofac Surg. 2008;66:2247-53. doi: 10.1016/j.joms.2008.01.041
15. Bell D1, Holsinger CF, El-Naggar AK. CRTC1/MAML2 fusion transcript in central mucoepidermoid carcinoma of mandible – diagnostic and histogenetic implications. Ann Diagn Pathol. 2010 Dec;14(6):396-401. doi: 10.1016/j.anndiagpath.2010.05.009
16. Bilodeau EA, Weinreb I, Antonescu CR, Zhang L, Dacic S, Muller S, *et al*. Clear cell odontogenic carcinomas show EWSR1 rearrangements: a novel finding and a biological link to salivary clear cell carcinomas. Am J Surg Pathol. 2013;37:1001-5. doi: 10.1097/PAS.0b013e31828a6727
17. Franca DCC, Moreira JM, de Aguilar SMHCÁ, de Carvalhos AA, Goiato MC. Ameloblastic carcinoma of the maxilla: a case report. Oncol Lett. 2012;4(6):1297-300. doi: 10.3892/ol.2012.937
18. Brown and Betz. Ameloblastoma: A review of recent molecular pathogenetic discoveries. Biomark Cancer. 2015;7(S2):19-24. doi: 10.4137/BIC.S29329
19. El-Naggar AK, Chan JKC, Grandis JR, Takata T, Slootweg PJ. WHO Classification of Head and Neck Tumours, 4th edition. Lyon: IARC, 2007; pp. 204-60.
20. Karakida K, Aoki T, Sakamoto H, *et al*. Ameloblastic carcinoma, secondary type: a case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2010;110:e33-e37. doi: 10.1016/j.tripleo.2010.08.018
21. Jaitley S, Sivapathasundharam B. Ameloblastic carcinoma of the mandible with clear cell changes: a case report. Indian J Cancer. 2013;50:8. doi: 10.4103/0019-509X.112316
22. Fan J, Kubota K, Imamura H, *et al*. Clear cell odontogenic carcinoma: a case report with massive invasion of neighboring organs and lymph node metastases. Oral Surg Oral Med Oral Pathol. 1992;74:768-75. PMID: 1488233
23. Slootweg PJ, Muller H. Malignant ameloblastoma or ameloblastic carcinoma. Oral Surg Oral Med Oral Pathol. 1984;57(2):168-76. PMID: 6366686
24. Moro A, Foresta E, Gasparini G, *et al*. Ameloblastic carcinoma of the maxilla: a case report and an updated review of the literature. Oncol Lett. 2016;12(6):4339-50. doi: 10.3892/ol.2016.5272
25. Casaroto AR, Toledo GL, Filho JL, Soares CT, Capelari MM, Lara VS. Ameloblastic carcinoma, primary type: case report, immunohistochemical analysis and literature review. Anticancer Res. 2012;32:1515. PMID: 22493395
26. Philip M, Morris CG, Werning JW, Mendenhall WM. Radiotherapy in the treatment of ameloblastic carcinoma. H K J Radiol. 2005;8:157-61.

27. Uzawa N, Suzuki M, Miura C, *et al*. Primary ameloblastic carcinoma of the maxilla: a case report and literature review. Oncol Lett. 2015 Jan;9(1):459-67. doi: 10.3892/ol.2014.2654
28. Kar IB, Subramanyam RV, Mishra N, Singh AK. Ameloblastic carcinoma: a clinicopathologic dilemma – report of two cases with total review of literature from 1984 to 2012. Ann Maxillofac Surg. 2014;4:70-7. doi: 10.4103/2231-0746.133070
29. Sargolzaei S, Khaleghjoo A, Taghavi N. Ameloblastic carcinoma of the maxilla: a rare case presentation and a review of new cases. J Solid Tumors. 2018;8(1):15-8. doi: 10.5430/jst.v8n1p15
30. Avon SL, McComb J, Crokie C. Ameloblastic carcinoma: case report and literature review. J Can Dent Assoc. 2003;69:573-6. PMID: 14653932
31. Jensen AD, Ecker S, Ellerbrock M, Nikoghosyan A, Debus J, Münter MW. Carbon ion therapy for ameloblastic carcinoma. Radiat Oncol. 2011;6:13. doi: 10.1186/1748-717X-6-13.
32. Perera E, Lindquist C, Hughes C, Thomas S. The use of gamma knife stereotactic radiosurgery in the treatment of ameloblastic carcinoma. Int J Oral Maxillofac Surg. 2013;42:934-8. doi: 10.1016/j.ijom.2013.04.015
33. Takahashi Y, Bandoh N, Miyamoto A, Kamada H. Single fraction helical tomotherapy for ameloblastic carcinoma. J Oral Maxillofac Surg. 2016;74:302-6. doi: 10.1016/j.joms.2015.07.015
34. Kruse ALD, Zwahlen RA, Grätz KW. New classification of maxillary ameloblastic carcinoma based on an evidence-based literature review over the last 60 years. Head Neck Oncol. 2009;1:31. doi: 10.1186/1758-3284-1-31
35. Giridhar P, Mallick S, Upadhyay AD, Rath GK. Pattern of care and impact of prognostic factors in the outcome of ameloblastic carcinoma: a systematic review and individual patient data analysis of 199 cases. Eur Arch Otorhinolaryngol. 2017 Oct;274(10):3803-10. doi: 10.1007/s00405-017-4631-7

Sažetak

SVJETLOSTANIČNA INAČICA AMELOBLASTIČNOG KARCINOMA MAKSILE: PRIKAZ SLUČAJA I PREGLED DIFERENCIJALNO-DIJAGNOSTIČKOG I TERAPIJSKOG PRISTUPA

*L. Manojlović, D. Jerković, D. Brajdić, P. Kneževi*ć i *S. Manojlović*

Ameloblastični karcinom je rijetko opisan zloćudni epitelni tumor koji najčešće zahvaća mandibulu. Maksilarni ameloblastični karcinom je još rjeđi, a njegova je svjetlostanična inačica izuzetno rijetko opisana. Budući da su svjetlostanične lezije u čeljusnim kostima i rijetke i dijagnostički zahtjevne, pojavnost ovih tumora, kao i kriteriji klasifikacije nisu jasno definirani. Diferencijalna dijagnoza obuhvaća brojne dobroćudne i zloćudne tumore odontogenog podrijetla ili podrijetla iz žlijezda slinovnica te metastaze. Dijagnostička se dvojba ogleda i u terapijskom pristupu, o čemu se u literaturi još uvijek raspravlja. Autori u ovom prikazu slučaja raspravljaju o kliničkoj i histološkoj diferencijalnoj dijagnostici svjetlostanične inačice agresivnog ameloblastičnog karcinoma maksile koji je uspješno liječen prijeoperacijskim zračenjem te kirurškom ekscizijom ostatne lezije i rekonstrukcijom maksile.

Ključne riječi: *Ameloblastični; Svjetlostanični; Odontogeni; Karcinom; Patologija; Radioterapija*