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Ameloblastic Carcinoma of the Mandible: Case Report and Literature Review

Ankur Rathaur, Prasanna Kumar P, Ankita Raj, Akash Tiwari, Rupesh Srivastav , Ayushi Agarwal

Rama Dental College Hospital & Research Centre, Rama University, Mandhana, Kanpur, Uttar Pradesh- India 209217

Abstract-Aim : This article aims to document an additional case of ameloblastic carcinoma affecting the left hemi-mandible, and to conduct a comprehensive review of its clinical presentations, radiologic and histopathological characteristics, various treatment strategies, and overall prognosis.

Case Presentation: We report the case of a 60-year-old female who arrived at the Maxillofacial Department complaining of painful swelling on her left cheek. After thorough diagnostic procedures, she was diagnosed with ameloblastic carcinoma of the left hemi-mandible. The patient underwent extensive treatment including hemimandibulectomy, supraomohyoid neck dissection, and reconstruction using a fibula free flap. Postoperative management included radiotherapy.

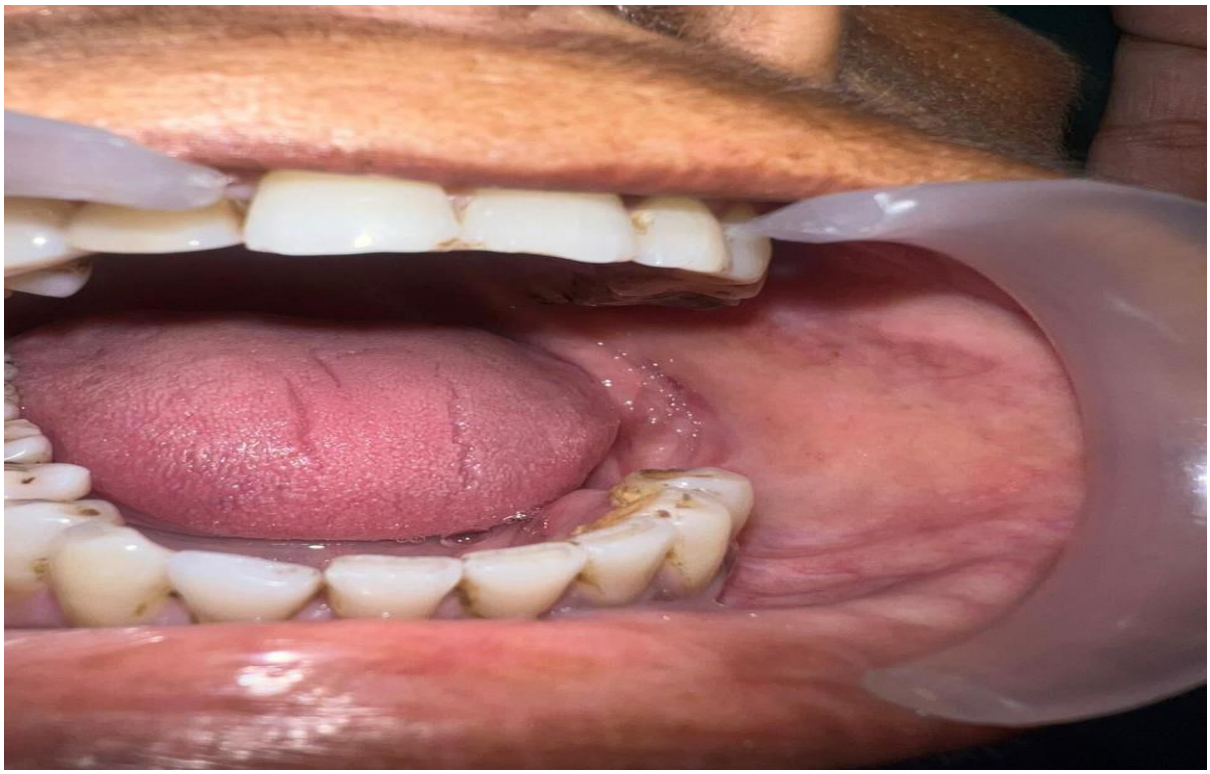
Discussion: Ameloblastic carcinoma in the mandible is a rare and highly aggressive tumor known for its poor prognosis. It manifests across a wide age range and is typically characterized by a rapidly enlarging painful swelling. Histologically, it is an epithelial odontogenic malignancy that shows both features of ameloblastic differentiation and malignant cytological characteristics. There is no established consensus on treatment, but the approach generally involves extensive surgical excision accompanied by radiotherapy.

Conclusion: Early diagnosis and regular monitoring for metastases are critical in enhancing the prognosis of patients with ameloblastic carcinoma. This case contributes to the scarce literature and emphasizes the need for awareness and prompt action in managing such aggressive tumors.

Introduction

Ameloblastic carcinoma (AC) is a profoundly rare and aggressive malignant epithelial odontogenic tumor characterized by poor prognosis. This cancer may develop de novo or from an existing benign ameloblastoma. Distinguishing AC from ameloblastoma and malignant ameloblastoma presents significant clinical challenges due to their similar manifestations, the potential for inconclusive cytology or biopsy results, and the need for different management strategies. Metastasis most frequently targets the lungs. Due to its rarity, there are no established treatment guidelines for AC.

We report a case involving a 60-year-old patient with AC of the mandible, highlighting the complexities of differential diagnosis and presenting an unusual outcome.



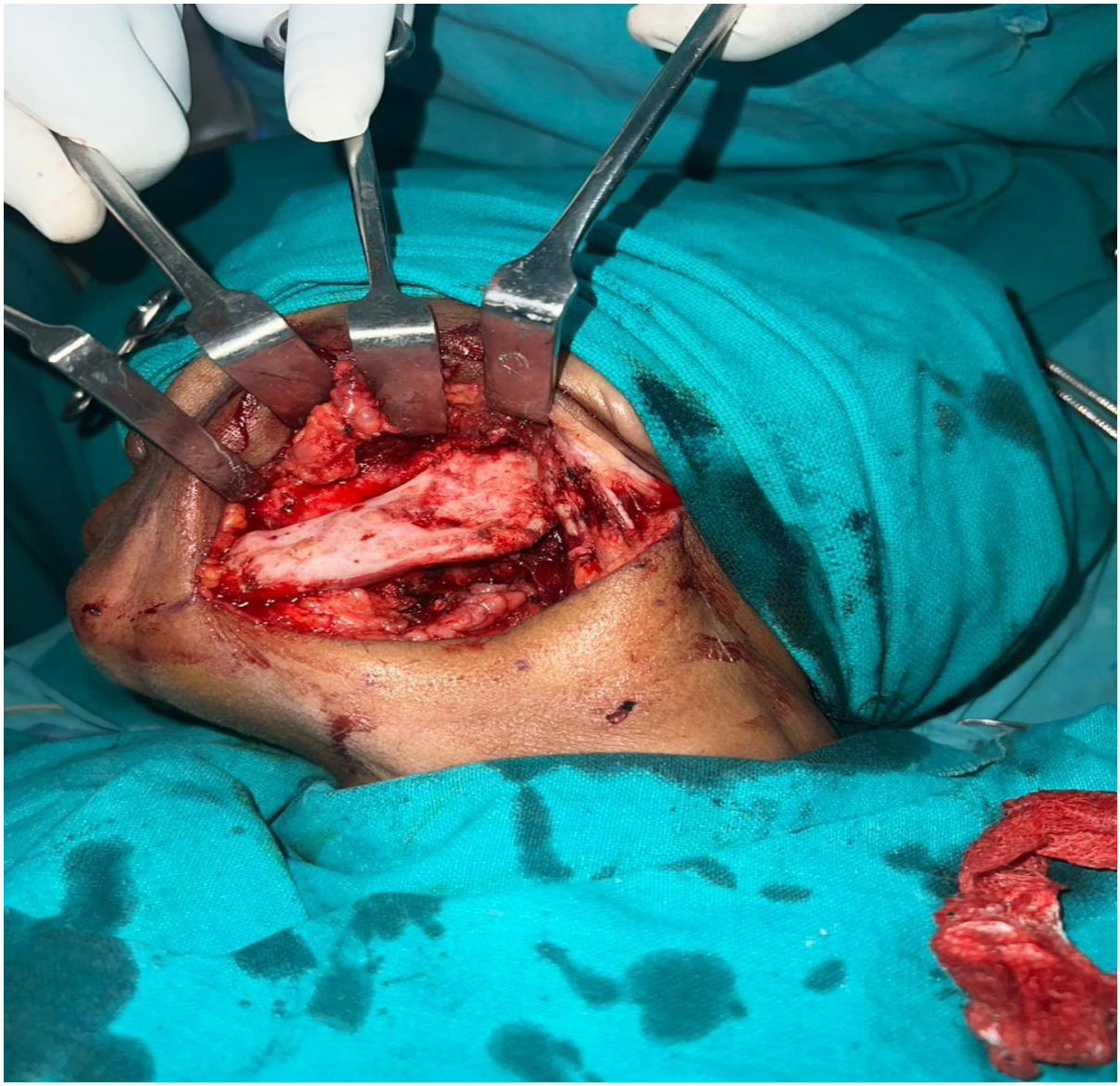
Observation

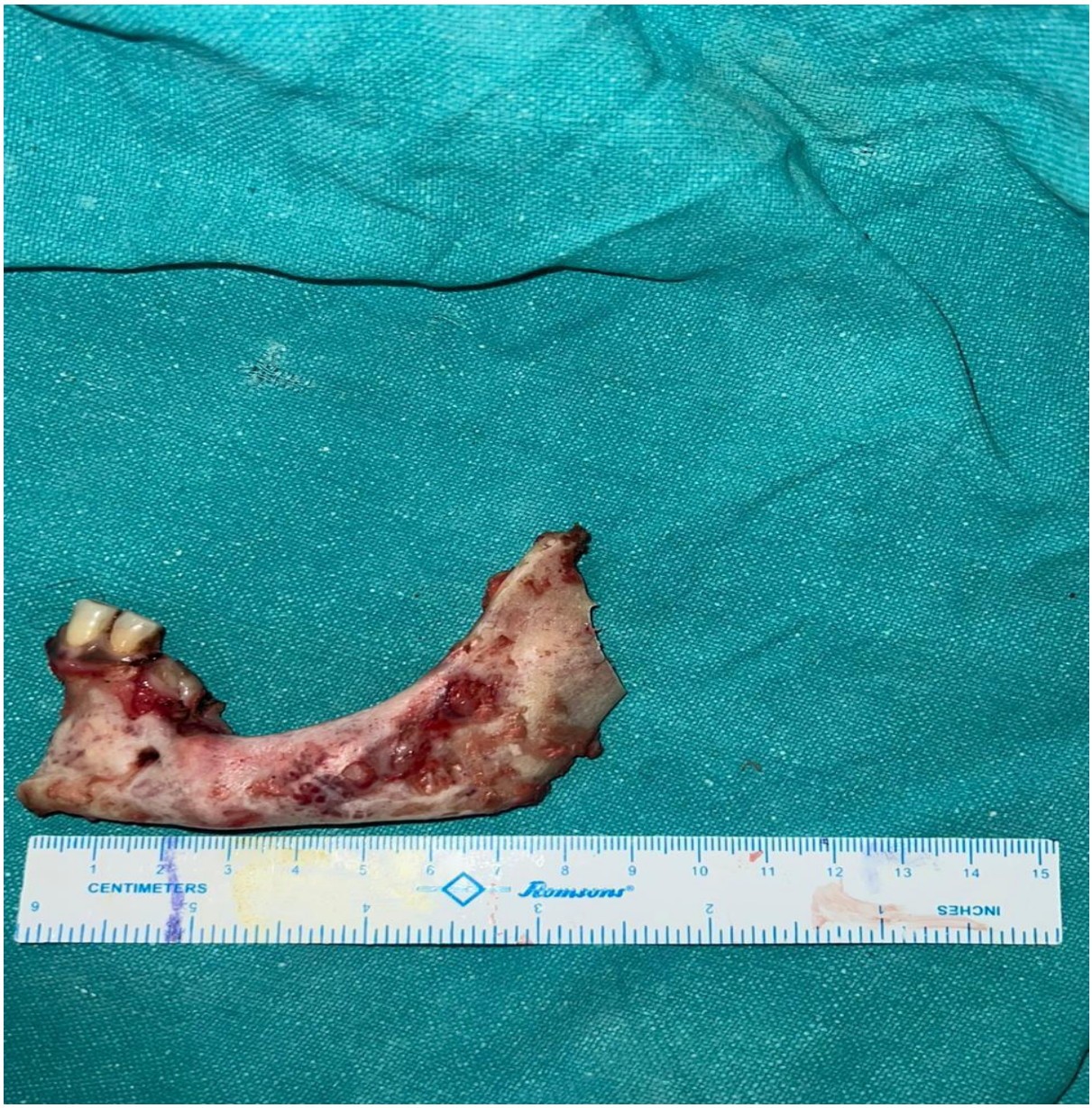
A 60-year-old female presented at the Department of Maxillofacial Surgery with a two-month history of swelling in the left cheek. Initial clinical examinations revealed a painless mass causing asymmetry in the left mandibular body, with no signs of trismus or anesthesia. The intra-oral examination indicated a partially edentulous state with vital and immobile teeth numbered 31, 32, 33, and 34. An ulcerated lesion was noted in the left retromolar area. Neck examinations showed no lymphadenopathy.

No Panorex imaging was conducted. A CT scan displayed an expansive mixed radiolucency, 60 x 60 mm, affecting the left ramus, mandibular angle, and body, perforating both buccal and lingual cortices with soft tissue extension. A PET scan was not available.

An incisional biopsy showed a tumor mass with islands and cords of odontogenic epithelium, featuring palisading columnar cells at the island peripheries. Histopathological and immunohistochemical analyses confirmed a diagnosis of ameloblastic carcinoma.

The patient underwent radical left hemimandibulectomy extending to the parasymphseal region and a prophylactic supraomohyoid neck dissection to mitigate the risk of delayed metastasis. Surgical margins were clear, and the defect was immediately reconstructed using a fibula osteocutaneous free flap. Postoperative treatment included 60 Gray of radiotherapy. Four years post-operation, the patient exhibited no signs of local or distant metastasis.

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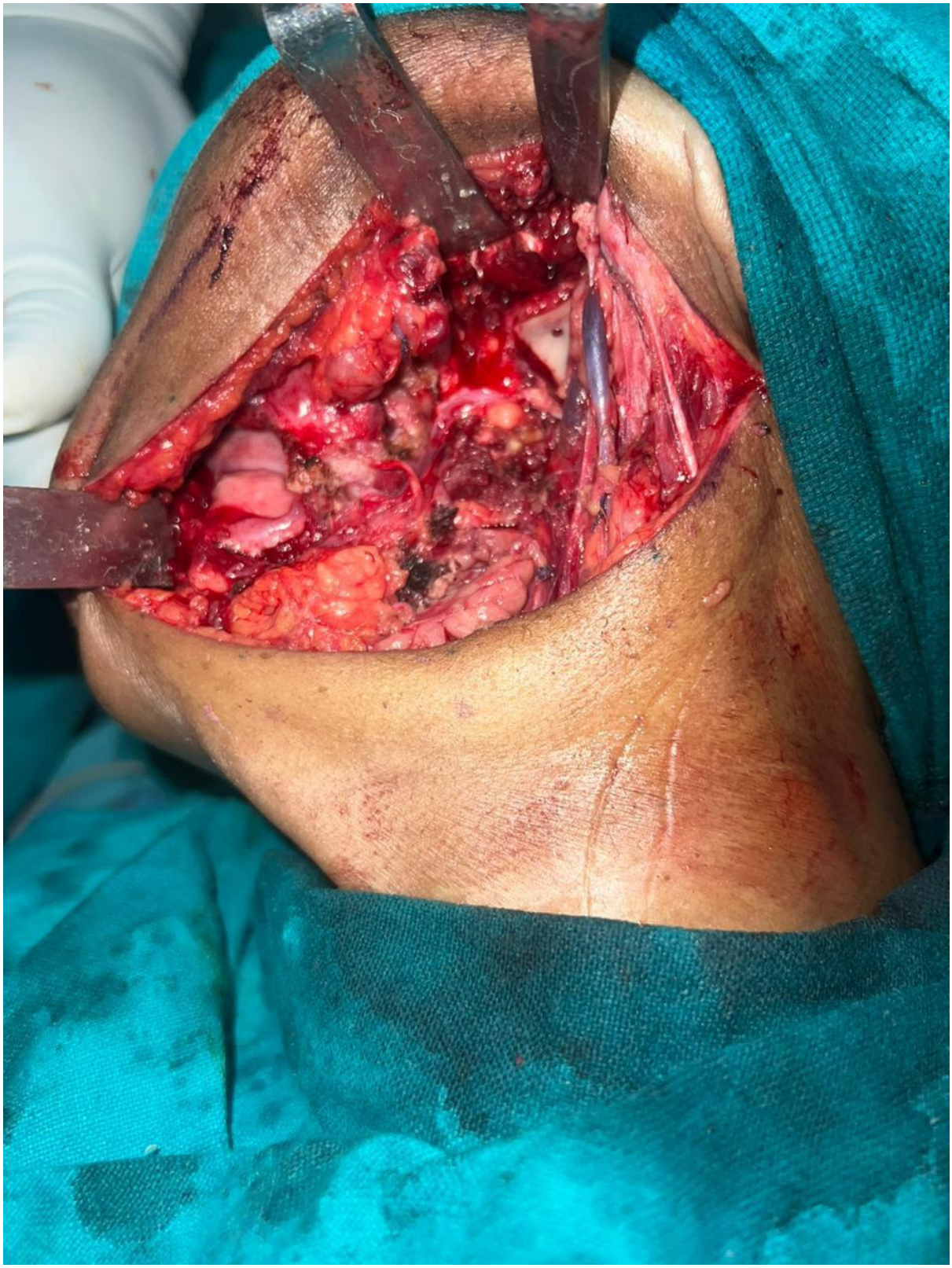

Discussion

Carcinomas derived from ameloblastomas are variably described as malignant ameloblastoma, metastatic carcinoma, and primary intra-alveolar epidermoid carcinoma, reflecting the nomenclature confusion. Slootweg and Muller categorized odontogenic carcinomas into three types to highlight their distinct biological behaviors and histomorphological characteristics:

Primary intraosseous carcinoma originating from an odontogenic cyst.
Malignant ameloblastoma or ameloblastic carcinoma, which may arise de novo, from existing ameloblastoma, or an odontogenic cyst.
Primary intraosseous carcinoma, which can be nonkeratinizing or keratinizing.
According to the World Health Organization's classification, ameloblastic carcinoma falls into group C of odontogenic carcinomas. The consensus is to use "ameloblastic carcinoma" for tumors showing histological malignancy signs in the primary, recurrent, or metastatic tumor, regardless of metastasis presence. In contrast, "malignant ameloblastoma" refers to metastasizing ameloblastomas with benign histological features in both primary and metastatic lesions.

A review by Benlyazid et al. in 2007 noted 67 reported cases of ameloblastic carcinoma, including one of their own cases. According to Sciubba et al., the mean age of onset is 30.5 years with a male to female ratio of 1.5:1, predominantly affecting the posterior mandible.

This expanded and detailed introduction, observation, and discussion provide a comprehensive overview of ameloblastic carcinoma, emphasizing its rarity, diagnostic challenges, and the clinical implications of its management.
Further Considerations on Ameloblastic Carcinoma

The majority of ameloblastic carcinomas (AC) originate de novo, as is the case with our patient, who had no significant medical history to suggest a precursor lesion such as ameloblastoma. The remainder of these cancers develop through malignant transformation from a preexisting ameloblastoma.

Clinically, ameloblastic carcinoma presents more aggressively than its benign counterpart, ameloblastoma. Distinguishing features of AC include rapid growth, painful swelling, cortical perforation, tooth mobility, non-healing extraction sites, ulceration or fistula formation, facial asymmetry, trismus, and paresthesia. These symptoms underscore its aggressive nature compared to typical ameloblastomas.

Diagnosing and determining the extent of AC are primarily aided by panoramic radiography and computed tomography (CT). Radiographically, AC often appears as an ill-defined destructive radiolucency with occasional focal radiopacities, perforation of buccal and lingual plates, evidence of root resorption, and aggressive extension into adjacent soft tissues.

Histologically, while these lesions retain some architectural features of conventional ameloblastoma, they exhibit significant cytologic atypia including increased mitotic activity, hyperchromatism, significant cellular pleomorphism, areas of necrosis, and evidence of neural and vascular invasion. Distinguishing de novo ameloblastic carcinoma from other malignancies such as primary intraosseous squamous cell carcinoma, metastatic carcinoma to the jaw, high-grade mucoepidermoid carcinoma, and invasive carcinoma from adjacent soft tissues can be challenging due to overlapping histologic features.

Ameloblastic carcinoma is typically marked by aggressive local behavior, potential regional nodal involvement, and hematogenous spread, with the lungs being the most common site for distant metastases. Once metastasized, the median survival is approximately two years.

Due to the rarity of ameloblastic carcinoma, there is no consensus on its treatment. However, wide local excision, including monobloc resection with 1 to 2 cm of healthy bone margin, remains the preferred surgical approach, yielding disease-free survival rates with local recurrence rates under 15%. Prophylactic cervical lymph node dissection is advised even in the absence of apparent lymphadenopathy.

Reconstructive strategies using osseous free flaps, especially from the fibula, are highly effective for primary mandible reconstruction, providing excellent functional and aesthetic outcomes. Alternative donor sites like the radius or scapula may be used depending on the extent of soft tissue and bone requirements.

Radiotherapy is recommended to reduce tumor size preoperatively or to improve local control when surgical margins are compromised. Chemotherapy has seen limited application, primarily in cases with metastatic disease, where regimens including cisplatin, adriamycin, and cyclophosphamide have shown some benefit.

Long-term follow-up is critical to monitor for late recurrence, metastasis, or regional lymph node involvement. Early diagnosis, regular screening for metastasis, and adequate treatment are essential to improve the prognosis for patients with ameloblastic carcinoma.

Conflicts of interest

The authors declare that they have no conflicts of interest in relation to this article.

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