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Case Report
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Microcystic adnexal carcinoma—diagnostic criteria and therapeutic methods: case report and review of the literature

M. Mamic[1], L. Manojlovic[2], P. Suton[3], I. Luksic[1]

[1]Department of Maxillofacial Surgery, University Hospital Dubrava, Zagreb, Croatia; [2]Department of Pathology, University Hospital Dubrava, Zagreb, Croatia; [3]Division of Radiation Oncology, Department of Radiotherapy and Medical Oncology, University Hospital for Tumours, University Hospital Centre "Sisters of Mercy", Zagreb, Croatia

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Abstract. Microcystic adnexal carcinoma (MAC) is a rare, infiltrating, locally aggressive cutaneous neoplasm of combined follicular and eccrine/apocrine histogenesis, usually presenting on the upper lip or face. Differentiation from other adnexal tumours is very important because the clinical management of these tumours is radically different, and misdiagnosis may lead to incorrect treatment. A case of recurrent MAC in the upper lip, treated with multiple excisions and postoperative radiation therapy (PORT), is presented herein. There have been no signs or symptoms of recurrence since the subsequent reconstructive surgery and PORT. Based on reports in the literature it appears that although immunohistochemistry can be helpful in distinguishing between MAC and other adnexal tumours, careful histopathological examination is essential for an accurate diagnosis. Perineural and intramuscular invasion strongly suggest the diagnosis of MAC. Its predilection for the facial area often limits the width of surgical excision. In such cases, PORT may be considered.

Key words: microcystic adnexal carcinoma; differential diagnosis; radiation therapy; reconstructive surgery; skin carcinoma.

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Microcystic adnexal carcinoma (MAC) is an uncommon, malignant adnexal tumour, which was first reported as a distinct pathological entity by Goldstein et al. in 1982[1]. Local recurrence has been reported to occur in 40–60% of patients after standard wide local excision[2–5], but this is much less likely if the excision margins are free of tumour in the initial excision. Therefore, accurate diagnosis is essential. Histological overlap with other benign and malignant cutaneous tumours presents the primary diagnostic challenge.

Surgical approaches including standard excision and Mohs micrographic surgery technique may require more extensive excision, resulting in a worse functional and/or aesthetic outcome. Possible disfiguring procedures require subsequent reconstructive surgery. Recent reports have discussed

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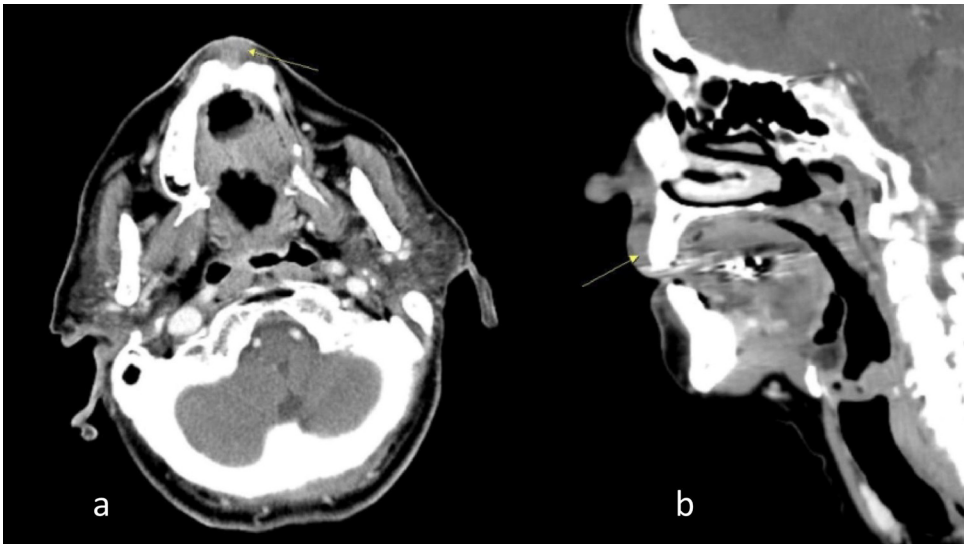

Fig. 1. Multi-slice computed tomography axial (a) and sagittal (b) reconstructions. Arrows point to the lesion.

the role of radiation therapy, either as an adjunct to primary surgical treatment or as monotherapy[6–10]. The results are controversial due to the rarity of the tumour. Reliable studies with a larger number of samples have not been reported.

Case report

A 74-year-old Caucasian woman with a 10-year history of recurrent desmoplastic trichoepithelioma (DTE) in the upper lip, presented to the University Hospital Dubrava, Zagreb with a newly formed tumour in the scar area. Clinically, there was a diffuse infiltration measuring 35 mm × 20 mm, encompassing the left side of the upper lip. The tumour was painless, but she reported a burning sensation. The overlying skin was hyperpigmented. Examination of the oral cavity, pharynx, and larynx was unremarkable and there was no lymphadenopathy. Multi-slice computed tomography (MSCT) scans revealed a soft tissue thickening 19 mm × 8 mm, without visible destruction of the maxilla (Fig. 1). Initial surgical excision was followed by reconstruction with transposable bilateral local lobes. Histopathological findings were consistent with recurrent DTE. After the surgery, the patient recovered with good functional and aesthetic results and was discharged to follow-up.

After a 36-month disease-free period, the patient presented with an upper lip nodule that had appeared suddenly 2 weeks earlier. Fine needle aspiration biopsy also hinted at a possible recurrence of DTE. Cuneiform excision of the upper lip was performed.

Histopathological analysis revealed tumour tissue composed of numerous small clusters, as well as tubular and cribriform structures of atypical epithelial cells with low mitotic activity, surrounded by abundant hyaline material. Perineural infiltration was prominent. Some of the tubular structures contained an eosinophilic material. MAC, morpheaform basal cell carcinoma (mBCC), and malignant cylindroma of the skin were considered in the differential diagnosis.

Histopathological re-evaluation and immunohistochemical analysis of all surgical specimens were performed. A similar histopathological pattern presented in all specimens. The poorly circumscribed tumour deeply invading the dermis and subcutis was composed of nests of atypical basaloid cells, embedded in the desmoplastic stroma. Some keratinous cysts and cystic glands were visible in the upper dermis (Fig. 2). Small ductal or glandular structures within a hyalinized stroma were visible in the deep dermis, accompanied by perineural and intramuscular invasion (Fig. 3). The tumour cells had bland histological features with little cytological atypia or mitotic activity. Immunohistochemical analysis revealed a strong positive reaction with various cytokeratins (CK7, CK5/6, CKAE1/AE3, CK19) and myoepithelial markers (p63, p40, CD10), and mild positive CEA reaction within keratin-filled cysts and duct lumina (Table 1). Negative reactions were observed with BerEP4 and androgen and progesterone receptors, as well as with Merkel cell marker CK20 (Fig. 4). Proliferative activity (Ki67 expression) was less than 5%. The histological and immunohistochemical pattern was consistent with MAC.

Due to the positive resection margin and the necessity for a more radical intervention, surgical revision treatment was indicated. Resection of the upper

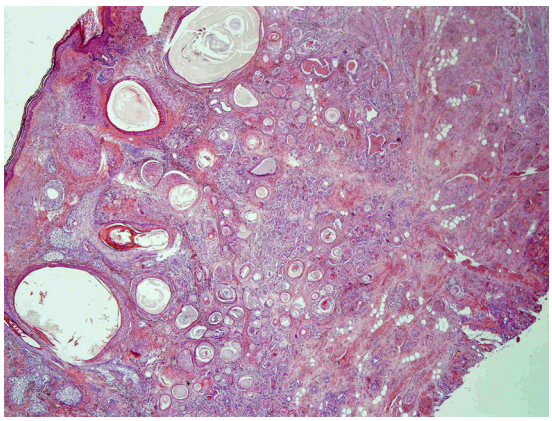

Fig. 2. The primary microcystic adnexal carcinoma misdiagnosed as a desmoplastic trichoepithelioma (haematoxylin–eosin ×40).

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