

Ameloblastic Carcinoma

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Abstract

Ameloblastic carcinoma is an extremely rare, malignant epithelial tumor of the jaws with a poor prognosis. The most common site of occurrence is the posterior mandible. Clinically, it is very aggressive and has potential for extensive local destruction. Majority of the cases arise de novo (primary type), but a few cases arise from a pre-existing ameloblastoma (secondary type). Early diagnosis and wide local excision on the primary site is the treatment of choice. The risk of malignant transformation should always be considered when a classic ameloblastoma is diagnosed and the prompt definitive management of an ameloblastoma is essential to eliminate this risk. Finally, meticulous, long-term follow-up is mandatory because recurrence and metastasis in the lung and regional lymph nodes have been reported.

Keywords: ameloblastic carcinoma, tumor

Introduction

Ameloblastic carcinoma is an extremely rare, aggressive, malignant neoplasm of the jaws with a poor prognosis. It belongs to the family of malignant epithelial odontogenic tumors [1]. It combines the histological features of ameloblastoma with features of cytological atypia regardless of whether it has metastasized [2, 3]. In contrast to ameloblastoma, ameloblastic carcinoma exhibits more aggressive clinical behaviour, such as rapid growth, perforation of the cortex and painful swelling [3, 4]. It may metastasize to the regional lymph nodes or lung [3]. In rare cases, brain or multiple bone metastases have been reported [3]. Ameloblastic carcinoma occurs in a wide age range of 15–84 years, but the mean age is approximately 30 years [5, 6]. There is no apparent sex predilection [5]. The most commonly involved area is the posterior portion of the mandible [5, 6]. Majority of the cases of ameloblastic carcinoma appear to arise de novo, and are termed primary ameloblastic carcinoma. Secondary ameloblastic carcinoma is defined as a tumor with malignant transformation within a pre-existing benign ameloblastoma, regardless of the presence of metastasis [4]. Secondary ameloblastic carcinoma is extremely rare.

Discussion

Ameloblastic carcinoma is a rare odontogenic tumor that poses a real challenge to the clinician in diagnosis, treatment planning and prognosis [7]. It is a neoplasm demonstrating histological evidence of malignant transformation of the ameloblastoma-like epithelial component in the primary tumor whether or not it has metastasized [8]. WHO defined ameloblastic carcinoma as a rare odontogenic malignancy that combines the histological features of ameloblastoma with cytological atypia even in the absence of metastases [7]. In the updated histologic classification of the WHO in 2005 [7], ameloblastic carcinoma is classified as primary type and secondary type. Primary ameloblastic carcinomas are those arising de novo whereas the secondary type arises from a pre-existing ameloblastoma. Ameloblastic carcinoma, secondary

type is extremely rare. The exact mechanism for the malignant transformation of ameloblastoma is currently unknown because of the limited number of cases [4]. Clinically, ameloblastic carcinoma is well known for its aggressiveness and extensive destruction of local structures [8]. The most common sign described has been swelling, although others include associated pain, rapid growth, trismus and dysphonia [5]. In most cases, radiographic findings show ill-defined radiolucency, however, focal radiopacity may be detected in radiolucent lesions [3]. Microscopically, it resembles the features of conventional ameloblastoma, except for the epithelium which shows various cytological features of malignancy [8]. Histologically, ameloblastic carcinomas have characters of both a benign ameloblastoma and carcinoma [9]. A palisade arrangement of epithelial cells with nuclei away from the basement membrane (reverse polarity) is a common feature of benign ameloblastomas [9]. The epithelial cells of ameloblastic carcinomas have features of hyperchromatism, a high mitotic rate, and a high nuclear-to-cytoplasmic ratio [9]. Recommended surgical treatment is jaw resection with 2–3 cm bony margin [5]. Ameloblastic carcinoma can recur locally 0.5–11 years after definitive therapy. Distant metastasis may occur as early as 4 months or as late as 12 years postoperatively and it is usually fatal [10]. Metastatic deposits of ameloblastic carcinoma are usually found in the lung, followed by bone, liver and brain [10]. Metastasis can occur even without any evidence of local recurrence [10]. The prognosis is much worse when distant metastasis occurs [9]. Cervical lymph node dissection should be considered when there is obvious lymphadenopathy [7]. The efficacy of adjuvant radiation or chemotherapy as a post-surgical treatment is not clear, because there is insufficient evidence that either provides a significant advantage [3]. However, radiotherapy and chemotherapy need to be considered in locally advanced cases and metastatic lesions not amenable to surgical resection [3]. The prognosis of ameloblastic carcinomas is poor [9]. The fatal factor includes the rapid growth of the primary tumor and distant metastasis [9]. Close periodic reassessment with a long-period of follow-

up (at least 10 years) is mandatory ^[7]. Radical surgical resection, strict screening for and early detection of metastatic lesions, and periodic follow-up after surgery are needed to improve patient prognosis ^[3].

Conclusion

Ameloblastic carcinoma is a rare entity of jaw neoplasm that combines the histological features of an ameloblastoma with features of cytological atypia, with a poor prognosis.

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