Extraskeletal myxoid chondrosarcoma of the external auditory meatus

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Abstract

Extraskeletal myxoid chondrosarcoma (EMC) is an uncommon but distinct entity with clearly different clinicopathological, immunohistochemical, cytogenetical and histogenetical features from conventional skeletal chondrosarcoma. Because of its better prognosis in comparison to conventional skeletal chondrosarcoma, the right diagnosis is essential. A case of EMC occuring in the external auditory meatus, coexistent with in situ squamous carcinoma of the covering epithelium is being presented. This is the first report of this tumor coexistence.

INTRODUCTION

Extraskeletal myxoid chondrosarcoma is an unusual but distinct entity characterized by special clinicopathological, immunohistochemical, and cytogenetical features, first described in 1953 by Stout and Verner [1]. It is a low-grade soft tissue sarcoma, locally aggressive and potentially metastasizing [2]. EMC is a tumor mostly affecting the deep soft tissues adjacent to the bones of the extremities, especially the lower one, and is rarely seen in other sites, such as pelvis, shoulder, chest wall, sinonasal tract [3], nasopharynx [4], jugular foramen, clavicle [5] and orbit. We are going to present a case of an EMC occurring in the external auditory meatus, which is the second one referred in the literature, located in this site [6] and, to the best of our knowledge, the first one coexistent with in situ squamous carcinoma of the covering epithelium.

CASE REPORT

A 74-year-old man admitted to the Hospital with a mass in the left external auditory meatus, causing poor hearing. CT scan showed a soft tissue mass filling left external meatus. The patient underwent resection of this mass, which was measuring 4.5 cm in maximum diameter.

PATHOLOGY

Gross examination revealed a whitish lobulated mass with some hemorrhagic areas. The cut surface displayed a partly myxoid appearance.

Sections showed dysplasia in the covering squamous epithelium of the external auditory meatus, focally evolving to in situ squamous carcinoma . In the dermis, there was invasion of round to oval-shaped cells arranged in nests and strands within a myxoid, Alcian-blue-positive, stroma. The nuclei were hyperchromatic and the cytoplasm eosinophilic. Focally, the cells displayed epithelioid features with vesicular nuclei containing nucleoli . The mitotic activity was low.

Immunohistochemically, the tumor cells were positive for vimentin and neuron-specific-enolase, but negative for epithelial membrane antigen, cytokeratin, S-100 protein, synaptophysin, and chromogranin. Our findings were consistent with the immunophenotype of EMC, as this has been documented in previous studies [7]. Differential diagnosis includes conventional skeletal chondrosarcoma, chordoma, parachordoma, myxoid chordoid meningioma, mixed myoepithelial tumor and other soft tissue lesions with myxoid appearance.

DISCUSSION

The histogenesis of EMC remains unclear and controversial. In a recent study [8], it is suggested that the basic cellular phenotype of EMC is not chondrocytic or prechondrocytic, thus excluding its chondrosarcomatous nature. It is more likely consisted of primitive mesenchymal cells with focal, multidirectional differentiation. Previously, immunohistochemical and ultrastructural features of neural or neuroendocrine differentiation had been reported by some authors [79101112], arising a new insight into the histogenesis of

this tumor. Consequently, according to the revised version of the World Health Organization classification of tumors of soft tissue and bone, EMC is classified as a tumor of uncertain differentiation [13]. Cytogenetically, the translocation t(9;22) (q22-31)(q11-12) has been found in most EMC cases. Cranial cartilage is considered to be derived from pluripotential precursor cells of the neural crest; on the other hand, chondrocytes found elsewhere in the body are believed to be derived from mesoderm. Soft tissue tumors with cartilaginous differentiation may thus be related either to neural crest or mesoderm, depending on their anatomic location. The presumed origin of EMC is cartilage, and thus of mesodermal origin when it occurs in locations other than the head and neck; actually, S-100 immunoreactivity in such cartilaginous neoplasms has been attributed to their chondroid nature and hence mesodermal derivation [14]. The present case was located in the external auditory meatus and was S-100 negative; so, its mesodermal derivation is ambiguous. Peripherin is an intermediate filament apparently expressed exclusively in cells derived from the neural crest and neural tube. The peripherin immunostaining in a considerable portion of malignant cells implies that EMC of the head and neck may contain a subpopulation of cells with the ability to demonstrate neural differentiation in addition to chondroid differentiation, as suggested Cummings et al [14].

As far as the present case is concerned, it is noteworthy that in a non-sun exposed site two distinct morphological types of malignancy appeared in close proximity; the first one was in situ carcinoma and originated from squamous cells and the other was invasive and probably derived from primitive cells of uncertain origin.

EMC, which occurs in almost the same incidence in both sexes, with a peak in the fifth to seventh decades, seems to be a quite indolent but potentially metastasizing tumor, especially to the lung. Additionally, recurrences are frequent. However, its high rate of metastasis is not accompanied by decreased patient's survival. Studies have shown [7], that the 5-year survival rate is 91%, while the 10-year overall survival rate is 78%. The presence of some adverse prognostic factors such as tumor size ≥ 10 cm, high cellularity, anaplasia or rhabdoid features, mitotic activity more than two per 10 high-power fields, and Ki-67 ≥ 10% indicates a possible more aggressive behavior, requiring a closer follow-up of the patient.

In summary, EMC is an uncommon but distinct entity, clearly different from conventional skeletal chondrosarcoma. Its location at the external auditory meatus is extremely rare, as until now, there has been only one such report in the literature.

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