EYELID TUMORS

Desmoplastic trichilemmoma: a rare tumor of the eyelid

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PURPOSE: To report an upper eyelid mass which proved to be a desmoplastic trichilemmoma.

METHODS: A 60-year-old man had a slowly enlarging upper eyelid mass. The tumor was excised. The pathologic evaluation of the tumor was centered on the differential diagnosis.

RESULTS: The clinical appearance of this lesion is nonspecific and can simulate a verruca, follicular keratosis, or basal cell carcinoma. Central desmoplasia, outer root sheath differentiation of the tumor cells, and CD34 positivity are the main characteristics that allow differentiation from basal cell carcinoma.

CONCLUSIONS: Proper recognition of a benign neoplasm that may be misdiagnosed as basal cell cancer can prevent aggressive surgical treatment.

Mantle cell lymphoma in the ocular adnexal region

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PURPOSE: To study the clinicopathologic features of mantle cell lymphoma (MCL) in the ocular adnexal region.

DESIGN: Retrospective review.

METHODS: The slides of 23 suspect patients were reevaluated with a panel of monoclonal antibodies, including anti-CD20, cyclin-D1, CD5, CD3, and p53 immunostains. Patients confirmed to have MCL were examined retrospectively on the basis of chart review.

RESULTS: Ten patients with periocular MCL were included in the study on the basis of characteristic histopathologic features and coexpression of nuclear cyclin-D1. This included 1 female and 9 male patients, with an age range of 32 to 84 years (median, 73.5 years). Median follow-up was 20 months (range, 5-172 months). Six of the 10 patients died, all of lymphoma. The orbit (90%) was most commonly involved followed by the lacrimal gland (50%) and lid (50%), with 90% of cases having lymphoma present at 2 or more periocular sites. Most had a primary periocular presentation (80%) that was associated with stage III/IV disease (80%), including atypical cells in the peripheral blood smear (60%) and bone marrow involvement (70%) at presentation. Three cases were CD5-negative, and 2 other cases showed composite histologic findings (MCL and follicular lymphoma and MCL and a plasma cell neoplasm).

Fluorescent in situ hybridization performed in these 2 cases demonstrated t(11;14) in the MCL component. Actuarial survivals were median progression-free (PFS) survival, 12 months; median overall survival (OS), 57 months; 5-year PFS, 0; 5-year OS, 39%.

CONCLUSIONS: Mantle cell lymphoma presenting in the ocular adnexal region has a male predominance and tends to affect an elderly age group, as is typical of MCL involving nodal sites. A higher frequency of these tumors fail to co-express CD5, and composite lymphomas were observed in 20% of patients. Mantle cell lymphoma presenting in the ocular adnexal region is associated with advanced-stage disease and short PFS but an OS similar to MCL at other sites.

Lymphoproliferative disorders of the ocular adnexa

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PURPOSE OF REVIEW: To describe recent advances in clinical features, diagnostic tools, and treatment modalities for lymphoproliferative diseases affecting the ocular adnexa.

RECENT FINDINGS: A close relationship of non-Hodgkin lymphoma with long-term use of aspirin and nonaspirin nonsteroidal antiinflammatory drugs has recently been documented. Some infectious agents have also been implicated in the pathogenesis of lymphomas. The advent of immunohistochemistry and polymerase chain reaction have helped to distinguish between benign lymphoproliferation and malignant lymphoma. New techniques for radiation therapy are proposed to decrease complication rates associated to this treatment.

SUMMARY: Recent studies demonstrated that ocular adnexal lymphomas may be masked by a number of different diseases and clinical features. Prognosis depends on the stage and histology of lymphoma as well as the location.

Immunohistochemical diagnosis of a rare case of epithelioid malignant peripheral nerve sheath tumor with multiple metastases

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BACKGROUND: The epithelioid variant of malignant peripheral nerve sheath tumor (MPNST) is a rare tumor with poor prognosis that sometimes involves the head and neck. The diagnosis is based principally on the histological examination, and it is generally very difficult to reach the correct diagnosis.

CASE: An 84-year-old Japanese woman presented with a tumor mass of 2 week's duration in the right medial canthal region.

OBSERVATIONS: Although the tumor was excised surgically, metastases occurred three times on her face and head, and the patient died of distant systemic multiple metastases. In the histopathological analysis, the tumor showed a composite pattern comprising spindle or polygonal cells arranged in irregular bands, and a population of larger epithelioid cells in solid sheets and nests. In the immunohistochemical analysis, the tumor cells were positive for S-100 protein, vimentin, and nerve growth factor receptor (NGFR), and negative for cytokeratin and HMB 45 (melanoma-associated

antigen). These findings confirmed the diagnosis of MPNST.

CONCLUSIONS: Epithelioid MPNST has complex histopathological findings and histopathological features similar to other epithelioid tumors, especially malignant melanoma. Immunohistochemical examination using NGFR and HMB-45 is important in the differential diagnosis.

Sebaceous carcinoma of the eyelids: personal experience with 60 cases

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OBJECTIVE: To describe clinical features, management, and prognosis of sebaceous carcinoma of the eyelid and adjacent structures.

DESIGN: Single-center retrospective interventional case series.

PARTICIPANTS: Sixty consecutive patients with sebaceous carcinoma of the eyelid and adjacent structures.

METHODS: Retrospective chart review and literature review.

MAIN OUTCOME MEASURES: Presenting features, sites of origin, location, growth patterns, management, histopathologic findings, incidence of recurrence, metastasis, and mortality.

RESULTS: The median age at referral was 72 years, with 73% female. Four patients had prior irradiation to the area where the sebaceous carcinoma developed. Initial clinical diagnoses elsewhere were sebaceous carcinoma (32%), blepharoconjunctivitis (25%), chalazion (20%), basal cell carcinoma (13%), and squamous cell carcinoma (10%). Initial histopathologic diagnoses elsewhere were sebaceous carcinoma (50%), squamous cell carcinoma (18%), basal cell carcinoma (8%), and others or not available (24%). Initial anatomic sites were upper eyelid (75%), lower eyelid (22%), caruncle (2%), and bulbar conjunctiva (2% [1 case]). Orbital exenteration was necessary in 13%. Recently introduced techniques of posterior lamellar resection of the eyelids with reconstruction (7%) hopefully will decrease this incidence in the future. Pathologically, 47% showed intraepithelial (pagetoid) involvement, 27% of sebaceous carcinomas arose from the meibomian glands, and 18% arose from both meibomian and Zeis glands. Local recurrence developed in 18%, metastasis in 8%, and death from metastasis in 6%.

CONCLUSIONS: Despite the fact that the clinical features of sebaceous carcinoma have been widely reported, the diagnosis was suspected initially in only 32% of patients at first examination elsewhere and in only 50% at histopathologic examination elsewhere. Orbital exenteration was necessary in 13%, mostly patients seen in the earlier years of the study. With more recently employed treatment methods, there is a tendency to avoid exenteration and to use more conservative methods of treatment. It is hoped that these modern therapeutic approaches will result in fewer cases of recurrence and metastasis.

Patterns of regional and distant metastasis in patients with eyelid and periocular squamous cell carcinoma

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PURPOSE: To determine the frequency and location of regional lymph node metastasis in patients with squamous cell carcinoma (SCC) of the eyelid and periocular skin. Patterns of distant metastasis were also investigated.

DESIGN: Retrospective case series.

PATIENTS: One hundred eleven patients treated at The University of Texas M. D. Anderson Cancer Center for SCC of the eyelid and periocular skin between 1952 and 2000.

METHODS: The clinical records of the 111 patients were retrospectively reviewed. The clinical characteristics analyzed included age, gender, location of lesion, treatment modalities, patterns of regional nodal and distant metastasis, and perineural invasion. Follow-up time ranged from 6 to 484 months (median, 76.6).

MAIN OUTCOME MEASURE: Incidence of regional lymph node metastasis.

RESULTS: The most common sites of SCC were the lower eyelid (54 patients [48.6%]), the medial canthus (40 patients [36.0%]), and the upper eyelid (25 patients [22.5%]). Local treatment of SCC consisted of wide local excision with frozen section analysis to ensure negative margins in 96 patients (86.4%), radiotherapy (without surgery) in 7 patients, and primary exenteration because of extensive tumor in 7 patients. Local recurrence occurred in 41 patients (36.9%). Twenty-seven patients (24.3%) had regional nodal metastasis during the study period. Sixteen of these patients had regional lymph node metastasis at the time of the initial presentation to M. D. Anderson Cancer Center. Eleven developed regional nodal disease later. Seven patients (6.2%) had distant metastasis during the study period. Nine patients (8.1%) had perineural invasion.

CONCLUSION: This study indicates that the overall rate of regional lymph node metastasis in patients with SCC of the eyelid or periocular skin may be as high as 24%. Thus, careful surveillance of the regional lymph nodes is an important aspect of the initial management of eyelid or periocular skin SCC. Consideration could be given to studying sentinel lymph node biopsy as a technique to stage SCC of the eyelid or periocular skin more accurately, especially in patients with recurrent, large, or highly invasive lesions or with perineural invasion.