Mixed Tumor of the Choroid

Mixed tumors, often referred to as pleomorphic adenomas, are common tumors of salivary1 and lacrimal2 glands. Rarely these tumors occur elsewhere in the body, most commonly in the soft tissue.3,4 I describe the first reported case, to my knowledge, of an intraocular mixed tumor arising in the choroid.

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Figure 2. Pathological findings of ciliary body medulloepithelioma. A, Low-magnification view of the tumor close to the sclera (hematoxylin-eosin, original magnification ×25). B, Tumor cells forming cords in a loose stroma resembling primitive vitreous (hematoxylin-eosin, original magnification ×200). C, Retinoblastoma-like areas with Flexner-Wintersteiner rosettes (hematoxylin-eosin, original magnification ×400). D, Neoplastic cells exhibiting nuclear retinoblastoma protein on immunostaining (anti-retinoblastoma protein antibody, avidin-biotin complex method, original magnification ×400).

Figure 3. Clinical evaluation 2 years after iridocyclectomy and brachytherapy.

Report of a Case. A 37-year-old woman underwent magnetic resonance imaging and was found to have diffuse intracranial leptomeningeal thickening and an expansive intra-axial mass in the distal thoracic spinal cord and conus medullaris during evaluation of persistent headaches and neck pain following a motor vehicle crash. She initially visited Duke Hospital after a witnessed seizure and was found to have extremely elevated intracranial
pressure that was refractory to aggressive medical management. Three days later, a perfusion scan revealed no evidence of intracerebral perfusion, she was unresponsive to stimuli, and she died.

Autopsy revealed diffuse leptomeningeal oligodendrogliomatosis, including the leptomeninges of the right optic nerve, with an intramedullary spinal cord component. The left eye had an amelanotic 3.0 x 2.5 x 1.0-mm mass in the choroid posteriorly (Figure 1A). The tumor cells had lightly eosinophilic cytoplasm and were organized as cords, nests, and ducts in a myxoid stroma containing spindle cells (Figure 1B). Ducts were best seen in the sections immunostained for cytokeratin and S-100 protein (Figure 2A and B). Some of the tumor cells had clear cytoplasm. Approximately 95% of the cells were immunoreactive with the pancytokeratin antibody AE1/AE3 (Figure 2A), and all of the cells expressed cytokeratin 7 and S-100 protein (Figure 2B). Vimentin (Figure 2C) and calponin (Figure 2D) were expressed by about 60% of the tumor cells, mostly in the central region of the tumor; smooth muscle actin expression had a similar distribution. Near the center of the tumor there was expression of CD57 between tumor cells, but the tumor peripherally was devoid of staining. There was only focal staining for epithelial membrane antigen (1% of cells), and only 1 tumor cell was undergoing DNA synthesis as evidenced by nuclear staining for Ki-67 antigen. The tumor cells did not express cytokeratin 20, neuroendocrine markers (chromogranin, synaptophysin, and neuron-specific enolase), p63 protein, gross cystic disease fluid protein 15, CD10, CD30, CD56, renal cell carcinoma antigen, CDX2, thyroid transcription factor 1, glial fibrillary acidic protein, WT1 protein, placental alkaline phosphatase, or α-fetoprotein, and they did not react with a cocktail of HMB-45/MART-1 antibodies.

Comment. Mixed tumors, myoepitheliomas, and parachordomas are extremely rare soft-tissue neoplasms thought to represent a continuum of myoepithelial tumors distinguishable based on their morphology. The tumor in this patient was classified as a mixed tumor owing to the presence of ducts as a minor component of the tumor. Immunohistochemical analysis confirmed the identity of this choroidal neoplasm as a mixed tumor expressing cytokeratin, S-100 protein, vimentin, and calponin by tumor cells. The tumor was deemed benign based on the lack of cytological atypia and the negligible proliferation rate.

Myoepithelial cells are normal constituents of salivary, lacrimal, mammary, and sweat glands. They are located within the epithelial basement membrane of secretory and terminal ductules and serve a contractile function. Myoepithelial cells have not been reported within the normal human eye to my knowledge, although ectopic lacrimal tissue containing myoepithelial cells occurs rarely within the eye. Based on the hypotheses surrounding the origin of intraocular ectopic lacrimal tissue, I postulate that this mixed tumor of the choroid arose from aberrant implantation of embryonic cells that were destined to become lacrimal gland myoepithelium.

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Socioeconomic Status and Choroidal Melanoma in Scotland

The adverse effect of socioeconomic deprivation on health and mortality is well recognized in the United Kingdom, particularly for health inequalities in Scotland. We evaluated the demographic characteristics and audited the management of our patients with choroidal melanoma at the only ocular oncology tertiary referral center in Scotland.

Methods. A retrospective record review was performed for clinical details at the first visit and subsequent treatments since January 1, 1994, for all patients with choroidal melanoma examined at the National Ocular Oncology Service at Tennent Institute of Ophthalmology, Glasgow, Scotland. All cases of choroidal melanoma in Scotland are referred from local ophthalmologists, general practitioners, and primary eye services to this center for initial evaluation. Choroidal melanoma was diagnosed via clinical appearance and ultrasonographic findings. Home address postcode was used to determine the Scottish Index of Multiple Deprivation score and correlated with the distribution of incidence and treatment modality. The Scottish Index of Multiple Deprivation records 7 domains (current income, employment, health, education skills and training, geographic access to services, housing, and crime). The data zones are ranked from most deprived (score of 1) to least deprived (score of 6505), leading to a picture of relative area deprivation across Scotland (http://www.scotland.gov.uk/Topics/Statistics/SIMD).

Results. A total of 536 patients were identified from January 1, 1994, to December 31, 2008, equating to 35.7 cases/year with an annual incidence of 7 per 1 million population. Mean (SD) age at the initial visit was 63.8 (13.7) years; 54.1% of the patients were male; and 50.4% of the lesions occurred in the right eye. All patients had unilateral pathological findings. The average wait from re-