The etiology of MCC is unknown, but some risk factors have been postulated, including increased age, Caucasian race, sunlight exposure, immunocompromised status, and polyomavirus infection.[1] More recently, an association with other prior cancers has emerged.[11] A significantly increased risk of MCC was observed among patients with a history of previous hematologic[11–14] and cutaneous malignancies,[11,12,14] in particular. Here, we report 2 cases of MCC of the eyelid after a primary breast tumour. MCC of unspecified anatomic location was observed in a total of 40 patients with history of breast cancer, but a significant association between the 2 entities was not established.[11,13–15] Although more evidence is required to support such an association, a link between MCC and breast cancer cannot be excluded.

Conclusions

We report 3 new cases of MCC of the eyelid—a very rare neuroendocrine tumour associated with poor prognosis, in part due to its often-delayed diagnosis. MCC commonly mimics benign and other malignant lesions, and recognition of MCC based on clinical grounds alone is difficult. Immunohistochemistry with positivity for CK20, expression of neuroendocrine markers, and negativity for thyroid transcription factor-1 allows its differentiation from its histologic imitators.

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References

1. Czapiewski P, Biernat W. Merkel cell carcinoma—recent advances in the biology, diagnostics and treatment. *Int J Biochem Cell Biol*. 2014;53:536-46.
2. Smith VA, Camp ER, Lentsch EJ. Merkel cell carcinoma: identification of prognostic factors unique to tumors located in the head and neck based on analysis of SEER data. *Laryngoscope*. 2012;122:1283-90.
3. Missotten GS, de Wolff-Rouendaal D, de Keizer RJ. Merkel cell carcinoma of the eyelid review of the literature and report of patients with Merkel cell carcinoma showing spontaneous regression. *Ophthalmology*. 2008;115:195-201.
4. Radovanovic AB, Latkovie Z, Borovie ML, Rašić DM. Primary Merkel cell carcinoma of the eyelid—clinical, histopathological, immunohistochemical and electron microscopical features: a case report. *Srp Ark Celok Lek*. 2015;143:309-13.
5. Cook BE Jr, Bartley GB. Epidemiologic characteristics and clinical course of patients with malignant eyelid tumors in an incidence cohort in Olmsted County, Minnesota. *Ophthalmology*. 1999;106:746-50.
6. Takamura H, Yamashita H. Clinicopathological analysis of malignant eyelid tumor cases at Yamagata university hospital: statistical comparison of tumor incidence in Japan and in other countries. *Jpn J Ophthalmol*. 2005;49:349-54.
7. Deprez M, Uffer S. Clinicopathological features of eyelid skin tumors. A retrospective study of 5504 cases and review of literature. *Am J Dermatopathol*. 2009;31:256-62.
8. Peters GB, Meyer DR, Shields JA, et al. Management and prognosis of Merkel cell carcinoma of the eyelid. *Ophthalmology*. 2001;108:1575-9.
9. Dekmezian MS, Cohen PR, Sami M, Tschen JA. Malignancies of the eyelid: a review of primary and metastatic cancers. *Int J Dermatol*. 2013;52:903-26; quiz 22-3, 26
10. Colombo F, Holbach LM, Junemann AG, Schlötzer-Schrehardt U, Naumann GO. Merkel cell carcinoma: clinicopathologic correlation, management, and follow-up in five patients. *Ophthal Plast Reconstr Surg*. 2000;16:453-8.
11. Howard RA, Dores GM, Curtis RE, Anderson WF, Travis LB. Merkel cell carcinoma and multiple primary cancers. *Cancer Epidemiol Biomarkers Prev*. 2006;15:1545-9.
12. Gass JK, Chan SK, Rytina E, Greenberg DC, Burrows NP. Multiple primary malignancies in patients with Merkel cell carcinoma. *J Eur Acad Dermatol Venereol*. 2010;24:601-3.
13. Youlden DR, Youl PH, Peter Soyer H, Fritschi L, Baade PD. Multiple primary cancers associated with Merkel cell carcinoma in Queensland, Australia, 1982-2011. *J Invest Dermatol*. 2014;134:2883-9.
14. Kaae J, Hansen AV, Biggar RJ, et al. Merkel cell carcinoma: incidence, mortality, and risk of other cancers. *J Natl Cancer Inst*. 2010;102:793-801.
15. Brenner B, Sulkes A, Rakowsky E, et al. Second neoplasms in patients with Merkel cell carcinoma. *Cancer*. 2001;91:1358-62.

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Haller cells: a risk factor for spontaneous orbital floor fracture?

Orbital floor fractures most commonly occur as a consequence of trauma through either buckling or hydraulic forces acting on the orbit. Spontaneous orbital floor fractures have been described in the literature usually in the setting of thyroid eye disease (TED). We present the case of a patient with a history of a spontaneous floor fracture associated with anteriorly displaced ethmoid air cells. We propose a pneumatic mechanism of spontaneous floor fracture in patients with Haller cells.

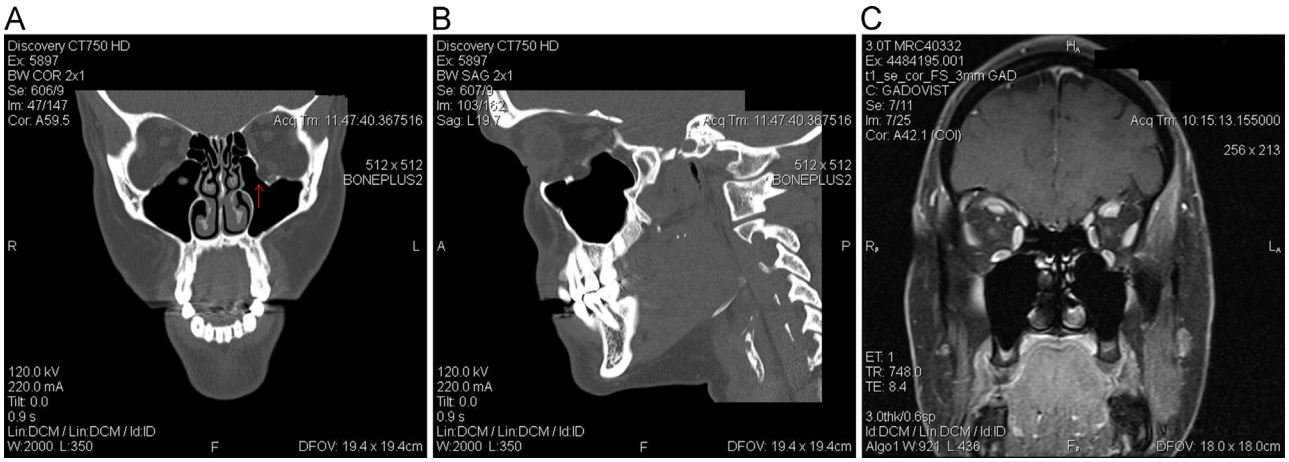

Fig. 1—(A–C) Computed tomography coronal and sagittal views, and magnetic resonance imaging coronal view of orbits. Site of fracture is shown (arrow) with entrapment of inferior rectus and soft tissue on magnetic resonance imaging.

Case Report

A 40-year-old otherwise healthy female patient presented with gaze-evoked diplopia that she had first noted 2 weeks previously. She described awakening with a sudden onset of double vision and initially having nausea and vomiting on upgaze. She denied any history of trauma and had an uneventful sleep. Seventeen years previously, the patient had been involved in a minor motor vehicle accident and had not sustained any injuries. On examination in clinic, she had normal visual acuity with no external evidence of trauma. Her examination was satisfactory except for marked limitation (75%) of monocular elevation of the left eye. She described vertical diplopia present mainly in upgaze. A forced duction test under topical local anaesthesia confirmed restriction and suspected entrapment of the inferior rectus muscle. A white blowout fracture was suspected.

Computed tomography imaging confirmed a defect in the left medial orbital floor with inferior herniation of orbital contents (Fig. 1A and 1B). Magnetic resonance imaging of orbits demonstrated displacement of the inferior rectus into the site of the fracture within a trapdoor deformity (Fig. 1C). On discussion with the patient, she elected to have exploratory surgery to repair the fracture. An anterior orbitotomy was performed via a swinging lower eyelid approach to the orbital floor. A posteromedial fracture was found with entrapment of soft tissue. The tissue was released and a 0.85-mm Medpor implant was fashioned and placed as a barrier over the fracture. There were no complications. Re-examination of her initial imaging highlighted the presence of anterior ethmoid air cells near and over the site of the fracture. These cells were continuous with the ethmoid capsule and more obviously noted on the contralateral side[1] (Fig. 2A–C). Unfortunately, the patient's symptoms persisted, suggesting ischemia and fibrosis of the muscle.

Discussion

The orbital floor is the most commonly fractured wall of the orbit.[2] This is in part related to the configuration of the

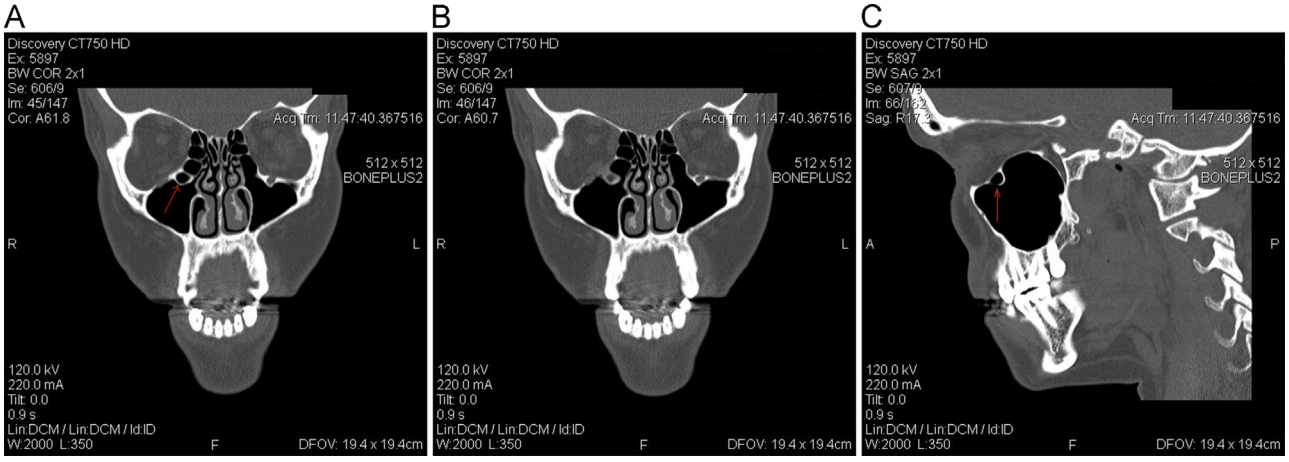

Fig. 2—(A–C) Computed tomography coronal and sagittal views of orbits demonstrating contralateral capsule of ethmoid cells with anterior air cell (Haller cell) at medial orbital floor (arrow). Sagittal view demonstrates presence of Haller cell at right orbital floor (arrow).