Journal of Clinical Neuroscience 33 (2016) 59–62

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Review

Malignant transformation of a residual cerebellopontine angle epidermoid cyst

Stylianos Pikis [a], Emil Margolin [b,*]

[a] Department of Neurosurgery, "Korgialenio Benakio" Red Cross Hospital of Athens, Athens, Greece
[b] Department of Neurosurgery, Hadassah University Medical Center, P.O. Box 12000, Jerusalem 91120, Israel

ARTICLE INFO

Article history:
Received 3 April 2016
Accepted 7 April 2016

Keywords:
Epidermoid cyst
Intracranial
Malignant transformation
Squamous cell carcinoma
Treatment

ABSTRACT

Malignant transformation is a rare but devastating complication following partial resection of an intracranial epidermoid cyst (EC). Time to malignant transformation is highly variable and optimal management is unclear. A literature search from 1965 to January 2016 identified manuscripts discussing clinical presentation, management, and outcome of malignant transformation of a remnant intracranial EC. One male patient diagnosed with malignant transformation of a remnant intracranial EC in our institution was also included in the study. There were 21 patients with malignant transformation of a remnant intracranial EC, including the current patient. Mean age was 51.4 years (range 36 to 77) and there was a female predominance (12 women, 9 men, ratio 1.33:1). The mean time interval from partial resection of a benign intracranial EC to malignant transformation was 7.74 years (range from 3 months to 33 years). Surgical resection of the tumor alone was the treatment of choice in 10 patients with one of them requiring a second operation and radiotherapy 2 months following the first operation. Adjuvant treatment modalities were employed in 11 patients and included radiotherapy (n = 4), stereotactic radiosurgery (SRS) (n = 3), chemotherapy (n = 1), chemotherapy combined with SRS (n = 1) and with radiotherapy (n = 1) and radiotherapy combined with SRS and followed by a second tumor resection (n = 1). Follow-up period ranged from 1 day to 5 years and 11/19 patients (57.8%) were reported dead on follow-up. Prospective studies are required to define the optimal management of malignant transformation of remnant intracranial EC.

1. Introduction

Intracranial epidermoid cysts (EC) account for 0.2%–1.8% of all intracranial neoplasms [1–6] and for 7% of cerebellopontine angle (CPA) tumors. They are histologically benign, slow growing tumors, believed to derive from aberrant ectodermal cells that become trapped during neurulation between the third and fifth week of intrauterine development [1]. Gross total resection is the treatment of choice of intracranial EC. However, when only subtotal tumor resection is possible, malignant transformation to squamous cell carcinoma (SCC) may occur rarely. Optimal management of such malignancies is unclear and prognosis is particularly poor.

We report on a 77-year-old man who presented with rapid neurologic deterioration 6 months after subtotal resection of a CPA EC due to malignant transformation of the remnant EC. The relevant literature for this unusual condition is also reviewed.

2. Methods

Relevant articles published from 1965 to January 2016 were identified using PubMed and Google Scholar. The search terms included "intracranial", "epidermoid cyst", "epidermoid" "malignant transformation", "malignant degeneration", "squamous cell carcinoma", "treatment", "surgery", "surgical outcome", "radiotherapy", "radiation" "chemotherapy", "radiosurgery", "prognosis". Additional reports were identified through a review of citations in papers identified in the search. We included studies on patients with remnant ECs reported to have undergone malignant transformation to SCC. Remnant EC was defined as a partially resected, histologically proven, benign, intracranial, EC. Interval to malignancy was defined as the time from tissue diagnosis of the initial benign EC to tissue diagnosis of SCC malignant transformation. Patient outcome was defined as the time from diagnosis of SCC malignant transformation to death or last reported follow-up.

The electronic file and imaging data for one patient who presented with malignant transformation of a remnant EC were retrospectively reviewed.

* Corresponding author. Tel.: +972 2 677 7092; fax: +972 2 643 1740.
E-mail address: emilm@hadassah.org.il (E. Margolin).

http://dx.doi.org/10.1016/j.jocn.2016.04.008
0967-5868/

3. Illustrative patient

A 77-year-old man was initially referred to our outpatient clinic in June 2013 with a 2-month history of unsteadiness and repeated falls. Brain MRI was significant for an extra-axial, cystic, non-enhancing, space occupying lesion in the left CPA (Fig. 1a). The patient underwent a left retrosigmoid sub-occipital craniotomy, and subtotal resection of the tumor. Intra-operatively a large cystic tumor was identified. The content of the cyst appeared grossly as a yellow-white, soft, cheesy material consisted with an EC. The patient had an uneventful post-operative course with complete resolution of his symptoms noted. Histological examination findings were consistent with a benign EC.

The patient remained asymptomatic until March 2014 when he was re-admitted because of a 2-week history of symptom recurrence. Brain MRI showed a cystic left CPA space occupying lesion. Gadolinium-enhanced T1-weighted brain MRI was significant for lesion enhancement (Fig. 1b). Partial resection of the tumor was achieved via a retrosigmoid sub-occipital approach. Histological evaluation of the tumor was compatible with malignant transformation of a benign EC to SCC. Post-operatively his neurologic status remained unchanged. He undergone a full systemic work-up and intracranial metastasis was ruled out. The patient was subsequently referred to the oncology department for adjuvant radiotherapy and chemotherapy. However, the patient did not consent to the chemotherapy regimen offered and he received adjuvant local radiotherapy (55 Gy) only. He continued to deteriorate and he died 6 months after the second operation.

4. Literature review

Including the patient presented here there were 21 patients with malignant transformation of a remnant EC to SCC reported in the literature. Data on patient age at the time of malignant transformation were available for all but one patient [2]. Mean age was 51.4 years (range 36 to 77) and there was a female predominance (13 women, 8 men, ratio 1.62:1). The mean time interval from partial resection of a benign intracranial EC to SCC malignant transformation was 7.74 years (range from 3 months to 33 years) with 42, 85% (9/21) of the patients presenting within 2 years from diagnosis of EC.

Tumor location included the CPA (n = 15), the temporal lobe (n = 2), the fronto-temporal region (n = 1), the left para-pontine and medial temporal region (n = 1), the base of the brain (n = 1), and the ventricles (n = 1).

Surgical resection of the tumor alone was the initial treatment preferred in ten patients with one of them requiring second operation and radiotherapy 2 months following the first operation [11]. Adjuvant treatment was employed in 11 patients and included radiotherapy only (n = 4), stereotactic radiosurgery only (n = 3), and chemotherapy only (n = 1). Multimodality adjuvant treatment was employed in three patients and included chemotherapy combined with SRS (n = 1) and with radiotherapy (n = 1). Adjuvant radiotherapy combined with SRS and followed by a second tumor resection was reported in one patient (Link).

Outcomes were provided for 19 of the 21 patients. Follow-up period ranged from 1 day to 5 years and 11/19 patients (57.8%) were reported dead on follow-up. For the 10 (43.2%) patients reported to be alive, the follow-up period ranged from 1 month to 60 months. Outcome data were available for seven out of nine patients managed with surgical resection of the tumor only [2,6,13–16,18–20]. All patients were reported dead. Survival of these patients ranged from 0.14 weeks (1 day) to 20 weeks. Including the patient reported here, adjuvant radiotherapy was reported in five patients [7,8,17,21]. Our patient despite aggressive radiotherapy died 6 months after diagnosis of EC to SCC malignant transformation while Kano et al., reported on a patient that died 25 months following diagnosis of malignant transformation [8]. Three patients treated with adjuvant radiotherapy were reported to be alive on follow-up ranging from 1 month [7] to 36 months [17]. Knorr et al., reported a 15 week survival in a patient initially managed with surgical resection of the tumor only which was

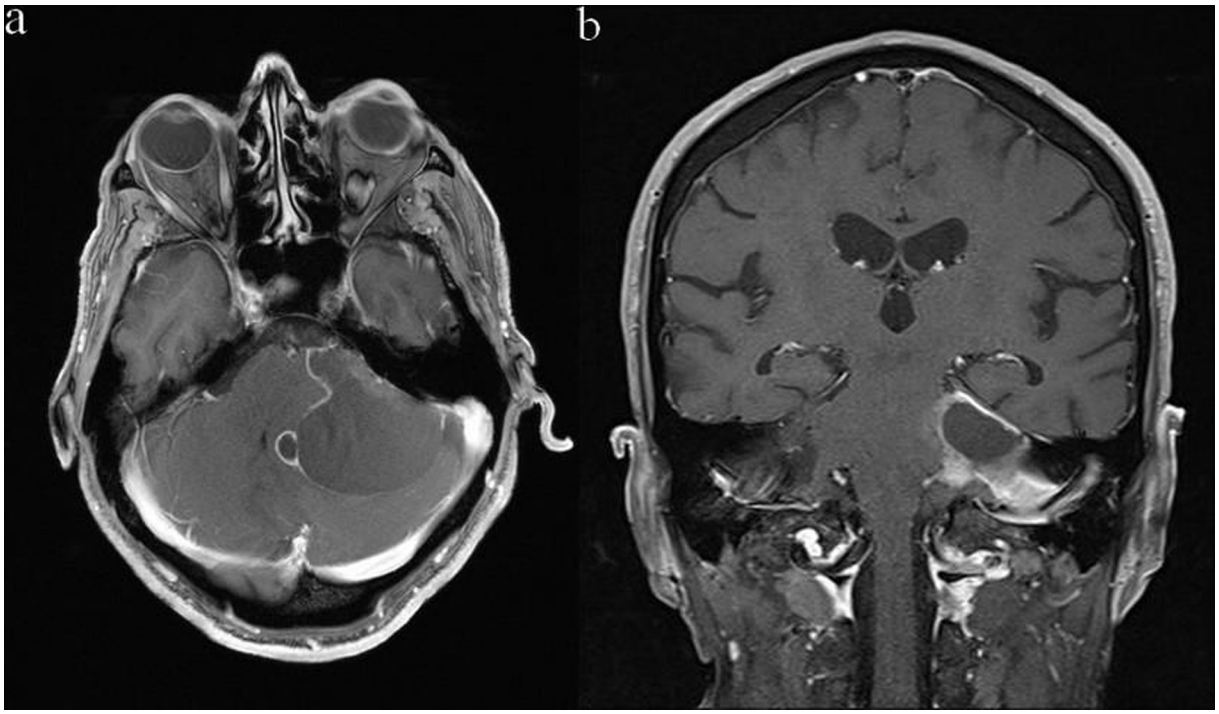

Fig. 1. (a) Axial T1-weighted brain MRI demonstrating a hypo-intense, cystic, space occupying lesion in the left cerebellopontine angle and (b) post-operative coronal, brain T1-weighted MRI significant for new contrast enhancement of the residual tumor.