Malignant Transformation of an Intracranial Extradural Epidermoid Cyst into Squamous Cell Carcinoma Presented with Cerebrospinal Fluid Leakage

Abstract

We report a case of malignant transformation of an intracranial extradural epidermoid cyst into squamous cell carcinoma (SCC), that presented with cerebrospinal fluid (CSF) leakage at the time of recurrence. Intracranial epidermoid cysts are histologically benign and slow-growing neoplasms. They are congenital lesions that develop from ectodermal remnants during neuroembryogenesis. Malignant transformation of epidermoid cysts into SCC is very rare. Various clinical presentations of these tumors after malignant transformation are mentioned in the literature. None of the previous cases, presented with CSF leakage as the recent case did. In cases of malignant transformation, surgical resection and then adjuvant radiation therapy are highly recommended.

Keywords: *Intracranial epidermoid cyst, intracranial squamous cell carcinoma, malignant transformation*

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Introduction

Intracranial epidermoid cysts are histologically benign and slow-growing neoplasms containing 0.2–1.8% of all intracranial tumors.[1-5] Malignant transformation of epidermoid cysts into SCC is very rare.[5] We present a case of malignant transformation of an intracranial extradural epidermoid cyst into SCC that presented with CSF leakage at the time of recurrence.

Case Report

An 83-year-old man with headache was referred to our clinic. He had a history of a chronic headache with different degrees of severity and changing patterns that continued for 2 weeks before his admission. The headache was global. Besides, it sometimes localized to the back of his head and neck. There was no association with nausea, vomiting, seizure, or fever. Physical examination did not demonstrate any neurological deficit. The patient was also suffering from a severe renal disease.

Brain magnetic resonance imaging (MRI) showed a well-defined epidural mass lesion in the left side of posterior fossa, with severe compression of the cerebellum. Opacification of adjacent mastoid air cells was seen. Craniocervical junction and upper cervical cord signal and thickness were normal [Figure 1]. These findings considered with an extradural posterior fossa mass lesion. Gadolinium images were not performed because of the presence of severe renal disease.

Gross total resection of the tumor was performed via the left retrosigmoid craniectomy. The tumor was a well-defined, large intracranial and completely extradural cystic lesion with a thin layer of a white capsule, containing yellow cheesy material. The cyst including the capsule and its contents was totally resected. The tumor eroded the inner surface of the skull at that site. Gross involvement of dura mater was not seen. The surface of the dura was shaved and coagulated, and eroded bones were removed. Microscopic examination showed interosseous epidermal inclusion cysts, filled with keratinous needle-like material [Figure 2]. The patient had a satisfactory recovery after the surgery.

Two months after the operation, the patient came back with a chief complaint of watery discharge from the surgery site. He also suffered from increased

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severity of headache, neck pain, and bulging site of surgery. Physical examination showed that the bulging was very loose. Fluid collection and leakage of clear to xanthochrome liquid via an orifice on the surgery wound were seen. No ulcerative or tumoral lesion was seen on the skin. The patient was not febrile, and his neurological examination was normal.

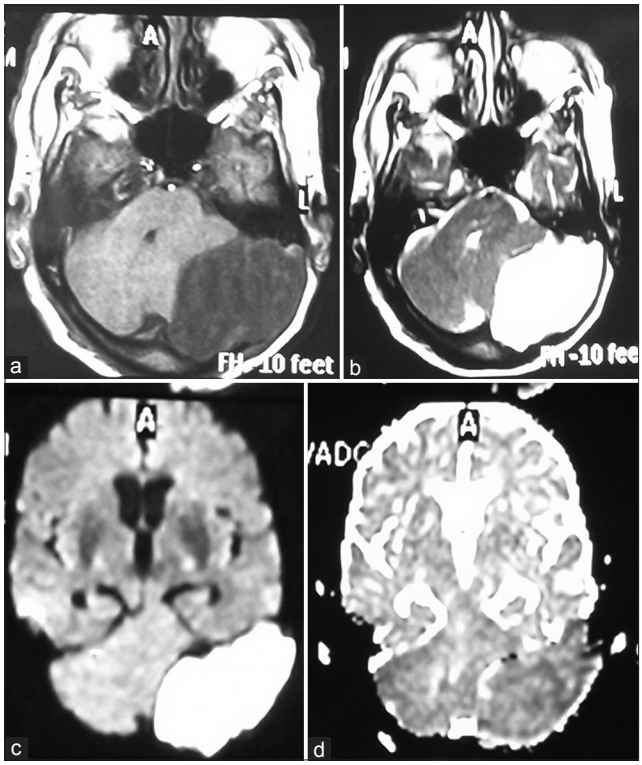

Figure 1: The first brain magnetic resonance imaging performed for the patient revealed an extradural posterior fossa mass lesion that was hypointense in T1 (a) and hyperintense in T2, (b) increased signal intensity in diffusion-weighted imaging, (c) and decreased signal intensity in apparent diffusion coefficient map, (d) suggested the diagnosis of an epidermoid tumor

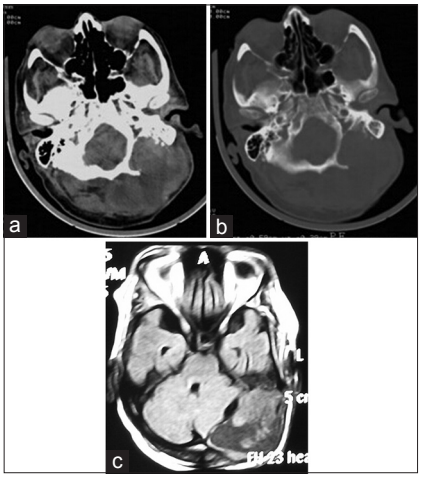

Figure 3: Two months after previous surgery, brain computed tomography scan revealed a large heterogenous dense mass in the left occipital part of the scalp (a), associated with destruction of the occipital bone (b), brain magnetic resonance imaging showed a heterogenous lesion including cystic and solid components in the left side of posterior fossa (c)

Brain imaging was performed. Brain computed tomography scan revealed a large heterogenous dense mass in the left occipital part of the scalp associated with destruction of the occipital bone. The brain MRI revealed a heterogenous lesion including cystic and solid components in the left side of the posterior fossa [Figure 3]. Gadolinium images were not performed due to the patient's severe renal disease.

The second operation was performed. The intraoperative findings were totally different from the previous one. After removal of about 50 ml of xanthochrome subcutaneous fluid, a large gray tumor with irregular borders was revealed. It invaded and destroyed muscles, skull, and dura. The tumor lesion was totally removed. CSF leakage was seen directly from the site of dural invasion to subcutaneous space. There was not grossly invasion to the cerebellar parenchyma, and the subarachnoid space persists between the tumor and the brain parenchyma. The dura was repaired with a fascia lata patch graft. The microscopic examination revealed a moderately differentiated SCC [Figure 4].

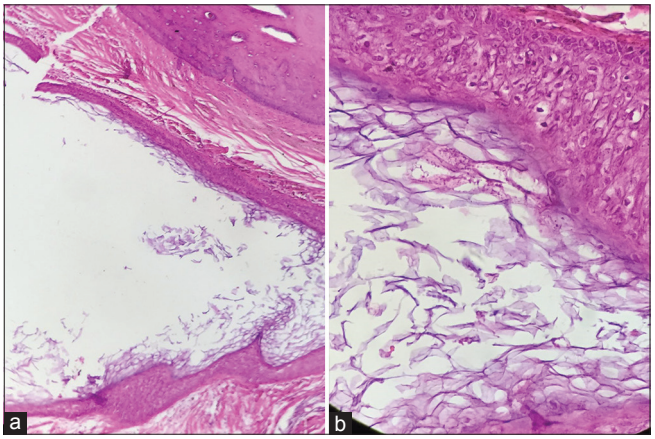

Figure 2: (a and b) Intraosseous epidermoid cyst contains multiple layers of squamous cells with granular layer and keratinous material (H and E, ×100, H and E, ×400)

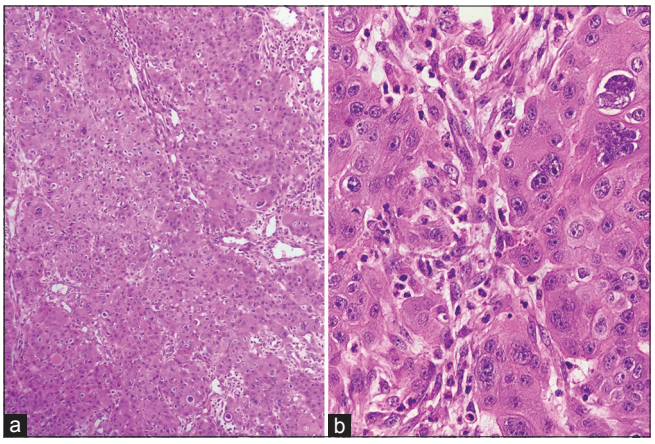

Figure 4: (a and b) Squamous cell carcinoma characteristically contains cohesive nests of cells with vesicular nuclear pattern and prominent nucleoli, in addition to eosinophilic cytoplasm. Nuclear pleomorphism, mitotic figures, accessional squamous pearls, and perineural invasion are identifiable elsewhere within this tumor (H and E, ×100, H and E, ×400)

Discussion

Intracranial epidermoid cysts are congenital lesions that develop from ectodermal remnants during neuroembryogenesis.[2] They mostly occur in the basal subarachnoid cisterns and ventricles, especially in the cerebellopontine angle (CP angle), fourth ventricle, and parasellar regions.[6] About 10% of all intracranial epidermoid cysts are extradural.[7]

Malignant transformation of epidermoid cysts into SCC is very rare.[5] Few cases were presented in the literature. Various clinical presentations of these tumors after malignant transformation are mentioned in the literature. Aggressive neurological symptoms in relation to the location of the tumor such as severe facial paresis, facial numbness, gait disturbance, and raised intracranial pressure in CP angle SCCs[5,8] are reported. In addition, silent behavior of the tumor and detection of the lesion in follow-up brain MRI were described in the literature.[4] Focal-enhanced mass lesion on the site of resection of an epidermoid tumor is a typical finding that highly suggests the occurrence of malignant transformation.[4]

None of the previous cases, presented with CSF leakage, as the present case report did. Treatment options in the case of intracranial SCC include surgery with adjuvant chemotherapy or radiotherapy. Radiation therapy following the surgery has an effective role on the disease-free survival of at least 5–8 years and local tumor control for 29 months.[9,10] Gamma knife neurosurgery has recently been reported as a useful adjuvant therapy in these cases.[9]

Conclusion

We reported a case of malignant transformation of an intracranial extradural epidermoid cyst into SCC that presented with CSF leakage. Intracranial epidermoid cysts are generally benign tumors. After resection of these tumors, following the patient with serial brain MRIs and physical examinations is recommended. Rapid onset of new neurological symptoms and symptoms of tumor recurrence and tumor enhancement in MRI suggests a malignant transformation of the epidermoid cyst. In these cases, surgical resection and then adjuvant radiation therapy are highly recommended.[5]

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Nil.

Conflicts of interest

There are no conflicts of interest.

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