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

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

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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Discussion

Intracranial epidermoid cysts are congenital lesions that develop from ectodermal remnants during neuroembryogenesis. They mostly occur in the basal subarachnoid cisterns and ventricles, especially in the cerebellopontine angle (CP angle), fourth ventricle, and parasellar regions. About 10% of all intracranial epidermoid cysts are extradural. Malignant transformation of epidermoid cysts into SCC is very rare. 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 are reported. In addition, silent behavior of the tumor and detection of the lesion in follow‑up brain MRI were described in the literature. 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. 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. Gamma knife neurosurgery has recently been reported as a useful adjuvant therapy in these cases.

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.

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Conflicts of interest

There are no conflicts of interest.

References