Case Report

Primary Intracranial Squamous Cell Carcinoma Arising in Dermoid Cyst

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Abstract

Primary intracranial squamous cell carcinoma represents a rare entity. However, few cases have been reported in the literature so far. We report the case of a 32-year-old male who presented with a history of severe headache and vertigo for 3 weeks. Magnetic resonance imaging brain with contrast was done that showed a lesion measuring 5 cm × 4.8 cm in the left cerebellum near the midline which was hyperintense on T2 and hypointense on T1. It also showed ring-like heterogeneous contrast enhancement causing midline shift as well as pressure over the brainstem causing severe obstructive hydrocephalus. After emergency ventriculoperitoneal shunt placement, the patient subsequently underwent left paramedian suboccipital craniotomy and excision of space-occupying lesion. Intraoperatively, a well-demarcated capsule was identified. The entire lesion consisting of areas of calcification and hair particles was removed except the part of capsule which was adherent to the brainstem. Histopathology of the lesion showed infiltrating malignant neoplasm exhibiting papillary architecture with thick fibrovascular cores lined by stratified squamous cells. In order to rule out the possibility of metastases, systemic workup was done which yielded negative results.

Keywords: *Squamous cell carcinoma, dermoid cyst, radiotherapy*

Introduction

Primary intracranial squamous cell carcinoma represents a rare entity. However, few cases have been reported in literature in which they may arise in a background of dysembryogenetic lesions such as dermoid or epidermoid cysts.[1] Intracranial dermoid cysts account for ~0.5% of all brain tumors and are usually located in the posterior fossa or cerebellopontine angle with slight female predominance.[2] Typically, dermoid cysts present in the first three decades of life. There is a remarkable difference in the outcome of such patients who present with malignant transformation, hence emphasizing the importance of diagnosing and treating these lesions aggressively. However, despite all measures, prognosis for squamous cell carcinoma remains poor, with surgery followed by radiotherapy being considered the best treatment option.[3] We present the case of a 32-year-old male who presented with primary squamous cell carcinoma arising in dermoid cyst in posterior fossa.

Case Report

A 32-year-old male presented to our neurosurgery clinic in February 2018 with complaints of severe unbearable headache associated with vertigo for 3 weeks. He had a magnetic resonance imaging brain with contrast done that showed a lesion measuring 5 cm × 4.8 cm in the left cerebellum, near the midline which was hyperintense on T2 and hypointense on T1. It also showed ring-like heterogeneous contrast enhancement causing midline shift and pressure over the brainstem causing severe hydrocephalus. The presence of atypical contrast enhancement made radiological diagnosis of lesion a bit difficult. However, due to severe headache, the patient underwent emergency placement of ventriculoperitoneal shunt. Postprocedure, the headache improved considerably, but he continued to have severe vertigo. The patient subsequently underwent left paramedian suboccipital craniotomy and excision of space-occupying lesion. Intraoperatively, a well-demarcated capsule was identified. The entire lesion consisting of areas of calcification and hair particles was removed except the part of capsule which was adherent to the brainstem. At this stage, a presumptive diagnosis of dermoid cyst was made, but frozen section was not sent. Histopathology of the lesion showed infiltrating malignant neoplasm exhibiting papillary architecture with thick fibrovascular cores lined by

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stratified squamous cells. Tumor also showed marked atypia and pleomorphism. This represents a moderately differentiated infiltrating squamous cell carcinoma arising in a background of dermoid cyst. In order to rule out the possibility of metastases, systemic workup was done which yielded negative results. The patient was later referred to an oncologist for further management [Figure 1a-d].

Discussion

A dermoid tumor is a congenital lesion that originates from ectodermal cells abnormally trapped during neural tube closure. These are slow-growing, benign tumors roughly accounting for 0.5% of all intracranial tumors and usually present during the third to fourth decades of life.[2] These lesions usually have a well-demarcated boundary with a true capsule primarily consisting of squamous epithelium without vascularity.[4] The occurrence of squamous cell carcinoma in these lesions represents a rare entity but has been well described in the literature. Although the exact mechanism leading to such transformation has not been identified yet, a theory has been proposed by Hamlat *et al*.[3] which states that a chronic inflammatory reaction triggered by cyst rupture eventually leads to such transformation. Other authors suggest the inflammatory nature of cystic contents that cause chronic irritation of epithelial cells leading to a malignant change.[5] When malignant transformation occurs, the clinical course is much more aggressive with significantly reduced survival.[6] Since intracranial squamous cell carcinoma can also occur as a result of metastases, it is important to perform a thorough systemic examination and workup to rule out any potential primary source.

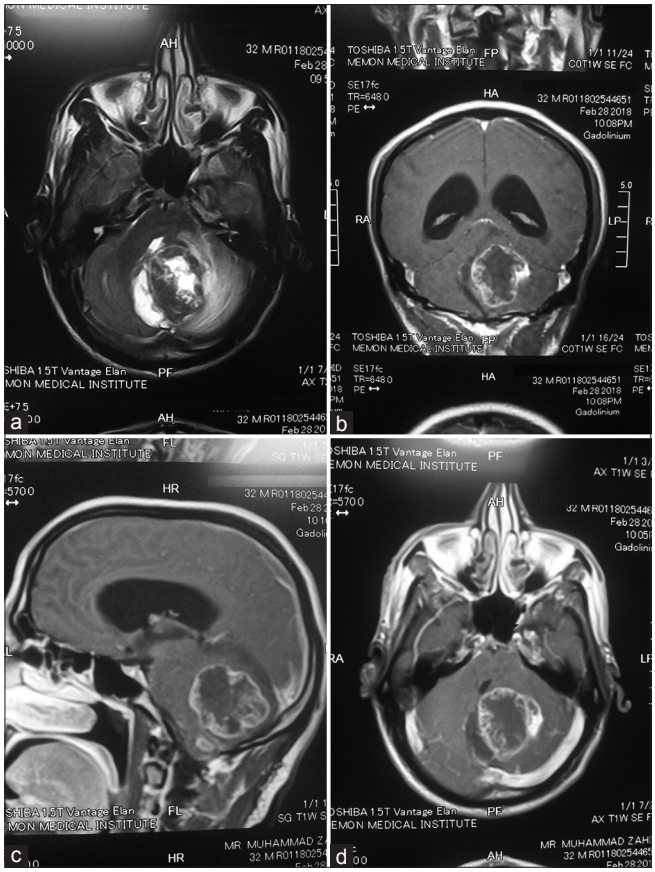

Figure 1: (a) T2-weighted axial cuts of the lesion, (b) T1 contrast coronal cuts of the lesion, (c) T1 contrast sagittal cuts of the lesion, (d) T1 contrast axial cuts of the lesion

Hamlat *et al*.[3] have proposed definitive criteria that must be fulfilled in order for a lesion to be classified as primary intracranial squamous cell carcinoma. These include the confinement of tumor intradurally with no extradural extension or communication through any of the orifices, no communication with middle ear, sinuses and sella turcica, no evidence of nasopharyngeal tumor, and absence of tumor elsewhere in the body. Our case meets the inclusion criteria and is therefore reported.

Radiological features of a typical dermoid cyst include iso- to hyper-intense on T1 and hyperintense on T2 with thin periphery contrast enhancement. In rare cases of malignancy, the enhancement becomes even more pronounced. Diffusion-weighted imaging (DWI) is an important modality to differentiate these lesions from simple arachnoid cysts that show enhancement on DWI.[7] The diagnosis of malignant lesion is still difficult due to variable degrees of cellular atypia,[2] but in our case, there was heterogeneous enhancement with surrounding edema in the cerebellum. Due to the rare nature of this pathology, the diagnosis was not suspected by our radiologist or primary neurosurgeon.

The recommended treatment option is to excise benign dermoid cysts completely including their wall,[8] but occasionally, this may not be possible if the lesion is adherent to the brainstem. Similar is the case with squamous cell carcinoma. Maximum safe resection is recommended; else complete removal may lead to significant morbidity.[9]

Radiotherapy has a significant role in reducing postoperative recurrence, particularly in cases where subtotal excision is carried out. Due to the rare nature of this pathology, no study has been carried out to date to assess the long-term outcome of dermoid cysts with malignant transformation. However, a study conducted by Nagasawa *et al*. revealed a significant increase in postoperative survival by comparing surgery versus surgery combined with radiotherapy in a total of 36 patients with malignant epidermoid cysts. Patients treated with surgery alone had a mean survival of 6.6 months, which was almost doubled to 12.7 months in patients who received postoperative radiotherapy.[10]

Malignant dermoid cysts have a poor prognosis. However, Tsugu *et al*. reported one patient with malignant dermoid cyst who survived for 6 years following complete surgical excision and local radiotherapy.[11] Such relatively long survival is extremely rare even after total removal of tumor and subsequent radiotherapy. We recommend maximum safe resection of lesion followed by local radiotherapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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