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Intracranial poorly differentiated squamous cell carcinoma: case report and literature review

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ABSTRACT

It is extremely uncommon to develop primary intracranial squamous cell carcinoma, which is typically brought on by the malignant transformation of epidermoid cysts or, less frequently, by other non-malignant epithelial cysts. We report a case of squamous cell carcinoma arise in the supra sellar region. She was operated via pterional approach. The patient was plaint of decrease in visual acuity. 6 months follow-up showed visual improvement. Intracranial squamous cell carcinoma is a rare disease whose clinical presentation, treatment, and prognosis are still unclear.

Keywords: squamous cell carcinoma, tumor epidermoid, supra sellar tumor

INTRODUCTION

Epidermoid tumors are congenital lesions that they include only ectodermal elements. They occur intracranially as a result of nests of epithelial cells remaining intracranially during embryogenesis possibly due to a failure of separation between neural and cutaneous ectoderm at the time of the neural groove closure [1]. They are rarely reported, their incidence account for 0.2% to 1.8% of brain tumors [2]. Unusually, they degenerate into squamous cell carcinoma (SCC). Epidermoid cyst mostly found in cerebellopontine angle, infratentorial middle cranial fossa, and suprasellar regions [3]. In 1912, Ernst et al, were the first one who describe SCC [1]. Garcia et al and Hamlat et al, have defined the criteria for such a malignant degeneration as follow [4,5]:

1. The tumor had to be restricted to the intracranial, intradural compartment without invasion of or extension beyond the dura or cranial bones.
2. There must be no extension or invasion through intracranial orifices, no communication or connection with the middle ear, air sinuses, or sella turcica, and no evidence of nasopharyngeal tumor.
3. The presence of a benign squamous cell epithelium within the malignant tumor.
4. Exclusion of carcinoma metastasis.

CASE REPORT

A 62-year-old female patient with no medical history was referred to our department for visual loss that had persisted for 5 years. Physical examination results were normal. Visual acuity was 3/10 in the right eye and light perception in left eye.

Imagery

Brain MRI showed a lesion arising from the pituitary stalk hypo-signal on T1WI with ring contrast enhancement which compress the optic chiasm and; hypersignal on T2WI (Figure 1).

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Treatment and outcome

Initially craniopharingyomas was suspected. The patient underwent pituitary function evaluation and it was in normal range. Patient was operated under general anesthesia with right pterional approach. The selection of the pterional approach was made with the objective of mitigating the potential risk of pituitary injury and not the endonasal approach was chosen nor transcallosal approach. Grosse total tumor resection was achieved. Post-operative brain Ct-scan showed the resection of the tumor with no complications (Figure 2). Histological examination revealed poorly differentiated squamous cell carcinoma (Figure 3). No postoperative complication has been seen. There was no recurrence at her 6-month follow-up. Visual acuity was 6/10 in the right eye and stationary in left eye.

DISCUSSION

Intracranial SCC arising de novo in the absence of a preexisting tumor, as in the present case, is even less common, and only eight cases have been reported to date, none of them in the sellar region.

Physiopathology

Hamlat et al. classified primary intracranial carcinomas into five categories based on clinico-pathological presentation [5]:

1. Initial malignant transformation of epidermoid cyst
2. Malignant transformation of remnant epidermoid cyst
3. Malignant transformation with leptomeningeal carcinomatosis

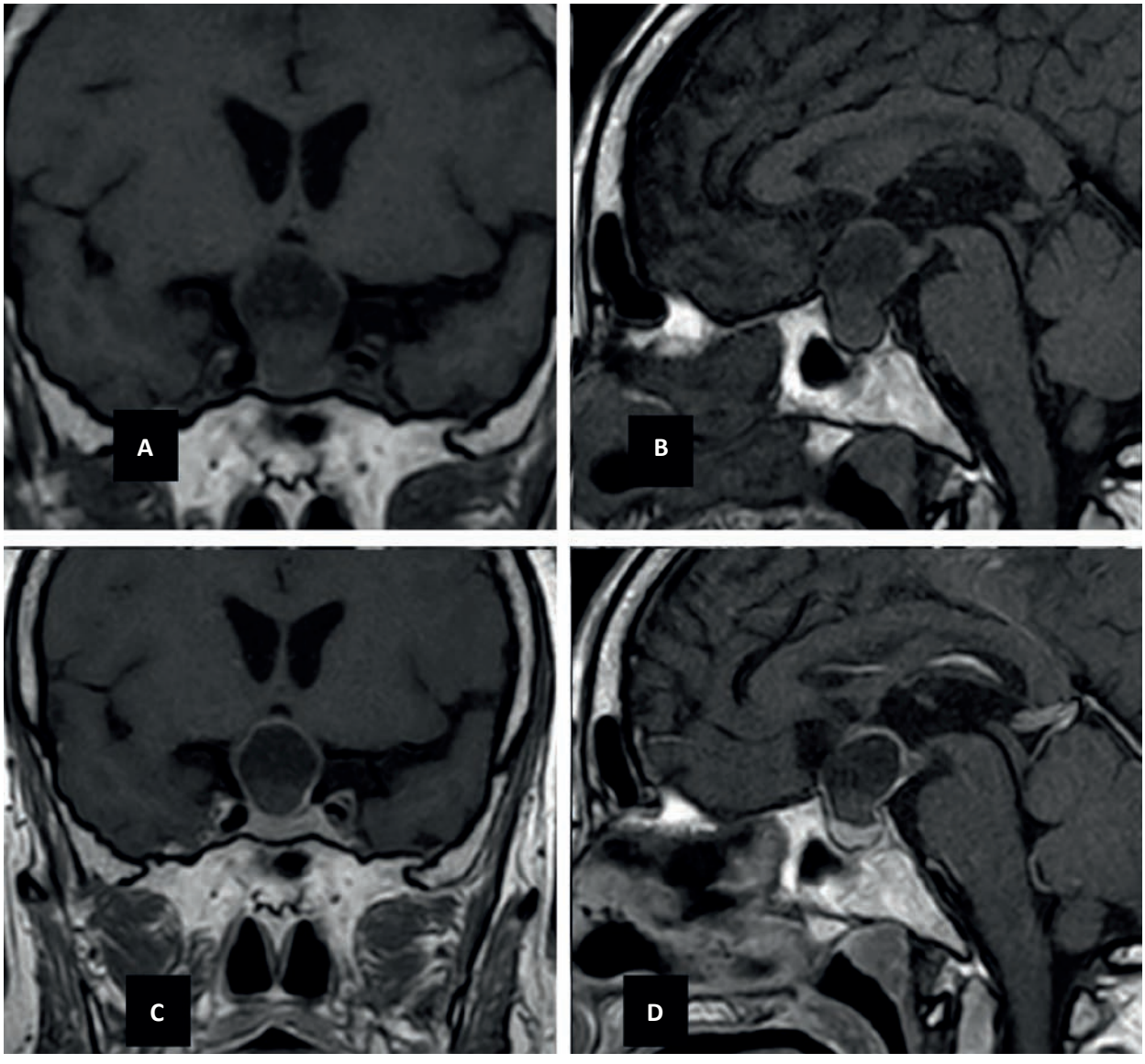

FIGURE 1. *Brain MRI showing groisierly oval intra- and supra-sellar cystic tumoral process, same signal as cerebrospinal fluid, signal suppressed on Flair sequence and unrestricted on diffusion sequences, with ring enhancement after injection of Gadolinium, measuring 30x2 1x22mm It compresses the optic chiasma as well as the floor of V3.*

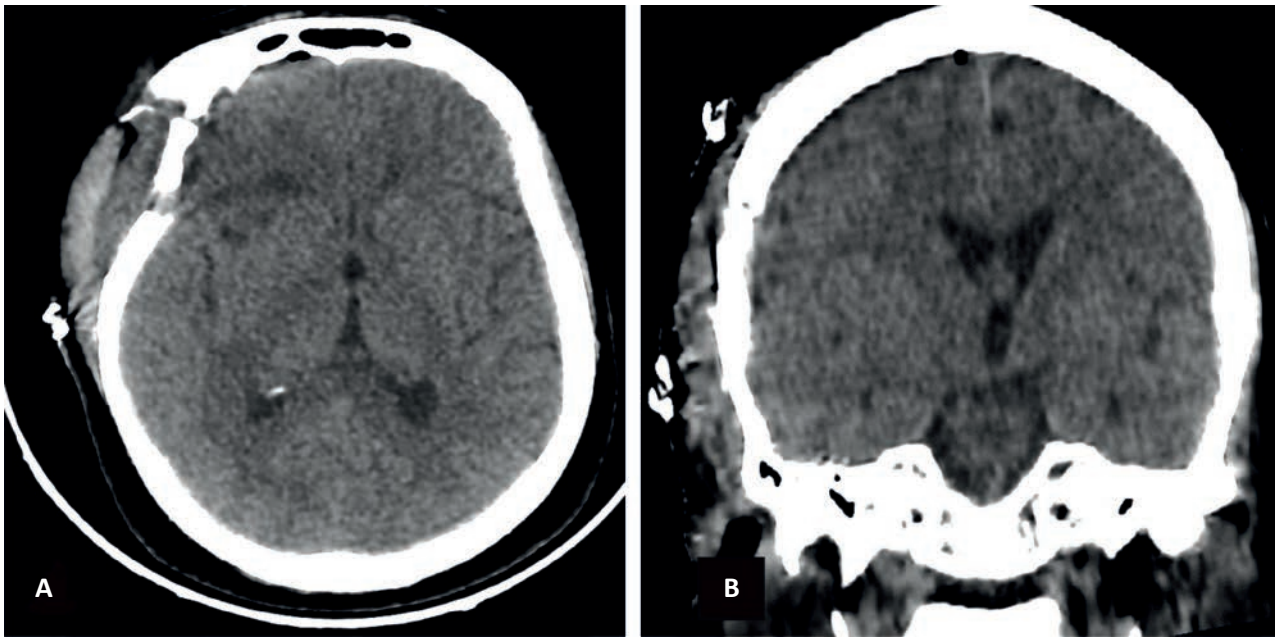

FIGURE 2. *Postoperative CT scan illustrating gross total tumor resection without complications through the right pterional approach*

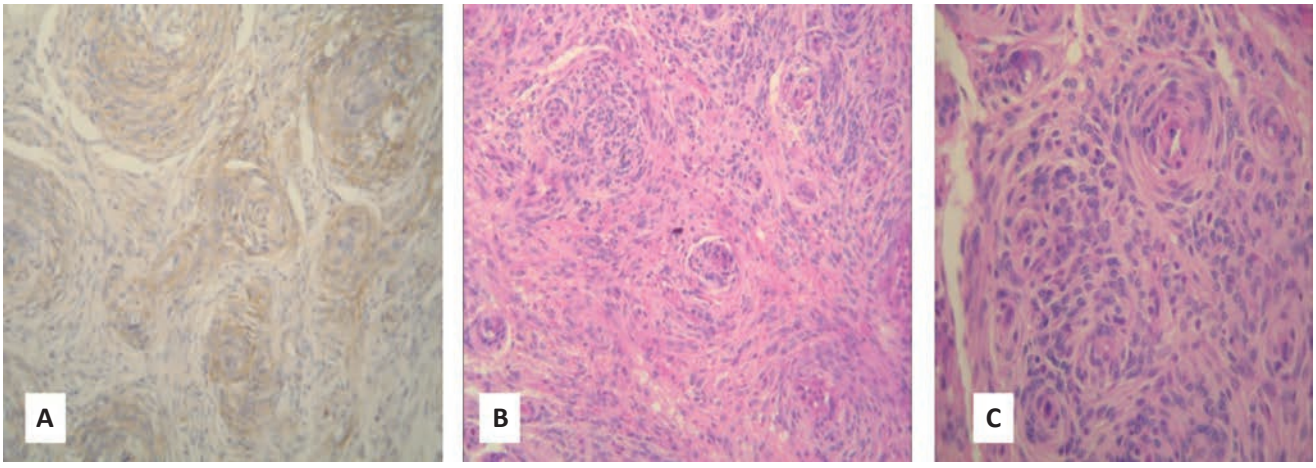

FIGURE 3. *Microscopically, tissue is composed of large, often polyhedral cells with eosinophilic cytoplasm, more or less sharply delineated borders and clearly atypical, enlarged, hyperchromatic, anisokaryotic nuclei, sometimes nucleated and richly mitotic. These cells are often arranged in large clusters within a fibrous, inflammatory stroma*

4. SCC arising from another benign cyst
5. Other malignancies arising from benign cysts including melanoma, adenocarcinoma, sarcomatoid carcinoma, and osteosarcoma.

Recurrent meningitis, intraoperative foreign material introduction, repeated irritation and cyst rupture, subtotal resection of the cyst wall, or chronic inflammation of an epidermoid cyst (EC) can all contribute into intracranial SCC [6].

Epidemiology and clinical presentation

The first case diagnosed with Intracranial SCC arising de novo was presented in 1976 by Wong et al. [7]. To date, 8 cases of that entity were described in world literature, and we present the 9th patient. [4,7-15]. The mean age of occurrence in Intracranial SCC arising de novo was 42.8 years, ranging from 4 to 68 years. Cerebellopontine angle was the most common, found in four patients (50%) (Table 1). Clinical presentation of primary intracranial SCC depends on their location (Table 2), it could be dysfunction or irritation of cranial nerve (V– VII–VIII and X), cerebellar and brain stem dysfunction, as well as hydrocephalus and meningeal irritation [12].

Imagery

Radiologically, ECs are hypodense on Brain CT scan, Hypointense on T1WI with lack contrast enhancement and hyperintense on T2 imaging and diffusion weighted imaging caused by the “T2 shine-through” effect. In malignant changes, they will have irregular margins, lack of diffusion restriction and contrast enhancement [14].

TABLE 1. *General information on patients diagnosed with primary intracranial squamous cell carcinoma arising de novo*

Feature	Number (%)
Demographic features	
Male	6 (75)
Female	2 (25)
Mean age	42.8 (4 years – 68 years)
Location	
Cerebellopontine angle	4 (50)
Para pontine	1 (12.5)
Ventricular	1 (12.5)
Frontal lobe	1 (12.5)
Temporal lobe	1 (12.5)

TABLE 2. *Clinical presentation of intracranial squamous cell carcinoma arising de novo*

Sign and symptom	Number (%)
Vestibular syndrome	3 (37.5)
Cranial nerve palsy	4 (50)
Headache	4 (50)
Hemiparesis	3 (37.5)
Seizure	1 (12.5)
Decreased visual acuity	1 (12.5)

Histology

Histopathologically, the epidermoid cyst wall include benign squamous epithelium, and the cysts have keratin debris and squamous epithelium but lack malignant cells. The primary intracranial squamous cell carcinomas have poorly differentiated epithelial cells with pleomorphic nuclei, and they display stromal invasion [12]. The immunohistochemical findings indicate positive reactivity for prealbumin and cytokeratin. The typical ependymal cyst wall exhibits columnar or cubic epithelium, with positive immunohistochemical results for GFAP, S-100, and other glial cell markers [15]. High Ki67 index indicating a relatively aggressive disease course [14].

Treatment and outcome

Treatment options including surgical management alone; surgery plus radiosurgery; surgery plus stereotactic radiosurgery (SRS); chemotherapy alone or in combination with other treatments; or surgery plus two or more adjuvant therapies. Surgery with adjuvant therapy augment surviving rate [12]. The full surgical resection of the squamous cell carcinoma from the brain is impeded by the tumor's robust adherence to the brain parenchyma and cerebral vessels. Consequently, the pursuit of an aggressive tumor removal approach may lead to severe neurological deficits [16]. The advantageous effects of radiotherapy on squamous cell carcinoma in various systemic organs have been extensively documented. The radiotherapy following surgical intervention may enhance the management of intracranial squamous cell carcinoma [17]. The 5-year survival rate is only 50% after surgery and radiotherapy (Table 3).

TABLE 3. *Treatment options and outcomes in intracranial squamous cell carcinoma arising de novo*

Feature	Number (%)
Treatment	
Gross total resection	4 (50)
Partial resection	2 (25)
Biopsy	2 (25)
Ventriculoperitoneal shunt	3 (37.5)
Radiotherapy	3 (37.5)
Outcome	
Complete neurological recovery	2 (25)
Improved with minor deficits	2 (25)
Died	4 (50)

CONCLUSION

Primary intracranial and sellar squamous cell carcinoma rarely occurs, usually arising from the malignant transformation of epidermoid cysts or, less commonly, other non-malignant epithelial cysts. There is a lack of consensus regarding the best approach to the management. Combination of treatments necessary to obtain the optimal outcome.

Conflict of interest: The authors declare there is no conflict of interest that could be perceived as prejudicing the impartiality of this review.

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