Radiology Case Reports 18 (2023) 753–756

Available online at www.sciencedirect.com

ScienceDirect

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

Case Report

Intracranial squamous cell carcinoma of the cerebello-pontine angle mimicking a cystic acoustic schwannoma. A case report with discussion of differential diagnosis and review of literature ☆

Lina Mliyh, MD[a], *Dario Di Perri, PhD*[b], *Valeria Onofrj, MD*[a,*]

[a] *Department of Radiology, Cliniques universitaires Saint-Luc, Avenue Hyppocrate 10, 1200 Bruxelles, Belgium*
[b] *Department of Radiotherapy, Cliniques universitaires Saint-Luc, Brussels, Belgium*

ARTICLE INFO

Article history:
Received 24 October 2022
Revised 10 November 2022
Accepted 16 November 2022

Keywords:
Intracranial epidermoid carcinoma
Tumors of the ponto-cerebellar angle
Epidermoid cyst malignant transformation
Cystic schwannoma
Neuroimaging

ABSTRACT

We report a case of a primary intracranial squamous cell carcinoma (SCC) of the cerebello-pontine angle extending through the internal auditory canal, with the unusual presentation of a completely cystic lesion with no diffusion restriction, internal necrotic-hemorrhagic changes and peripheral enhancement, mimicking a cystic acoustic schwannoma. The lack of diffusion restriction and the peripheral enhancement along the lesion, 2 unique findings, supposedly reflected complete cancerization of the epidermoid cyst from which the SCC originated. We discuss the differential diagnosis and review the literature on primary intracranial SCC.

Introduction

Intracranial epidermoid cysts are benign lesions that can very rarely undergo malignant transformation into squamous cell carcinoma (SCC) [1]. Imaging presentation of primary intracranial SCCs arising from an epidermoid cyst can vary: the diffusion restriction usually attributed to the benign epidermoid cyst can be variably present in the malignant component of the lesion [2]. Ring enhancement is a non-specific sign of cancerization of an epidermoid cyst [3].

Abbreviations: SCC, Scuamous Cell Carcinoma; CPA, Cerebello-pontine Angle; IAC, Internal Auditory Canal; DWI, Diffusion Weighted Imaging; ADC, Apparent Diffusion Coefficient.

☆ Competing Interests: The authors have no financial and personal relationships with other people or organizations that could inappropriately influence their work.

* *Corresponding author*.
E-mail address: valeria.onofrj@saintluc.uclouvain.be (V. Onofrj).

https://doi.org/10.1016/j.radcr.2022.11.048

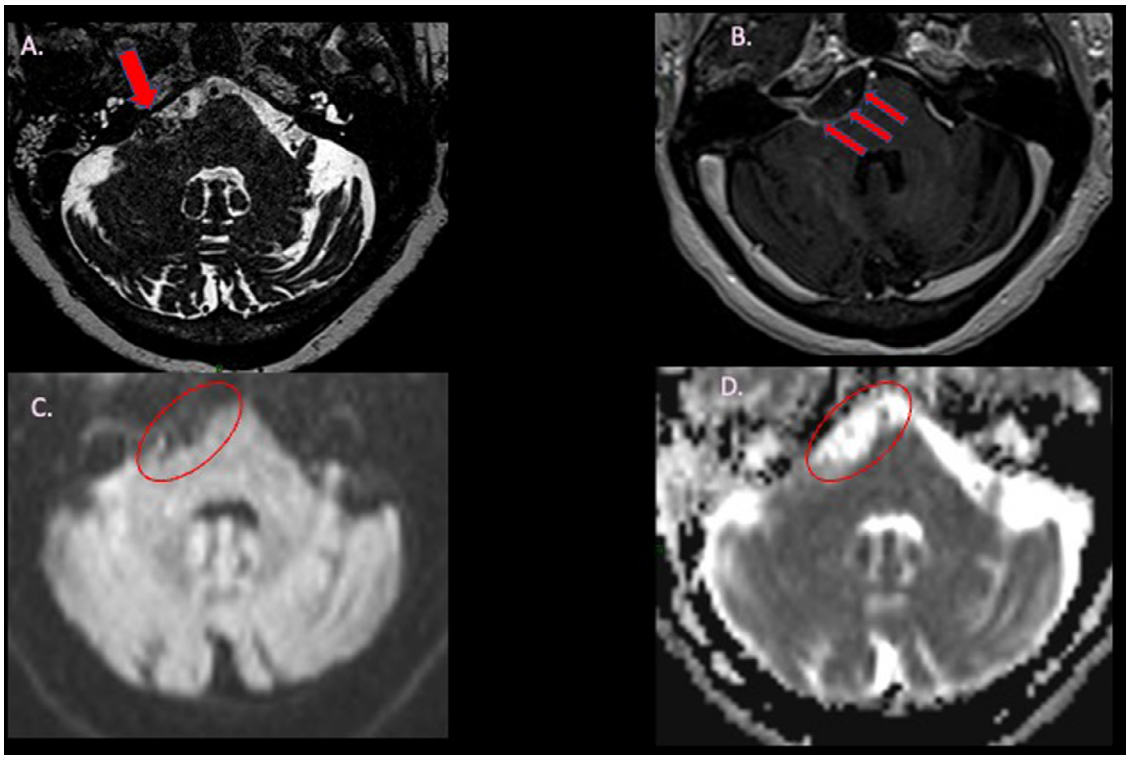

Fig. 1 – (A) Axial balanced steady-state sequence shows a cystic lesion of the right CPA, containing a blood-fluid level (red arrow). (B) T1-weighted post-contrast axial images show peripheral enhancement (red arrows). (C) DWI axial images and (D) the ADC map show absent diffusion restriction (red circles).

Differential diagnosis includes cystic lesions of the cerebellopontine angle (CPA), such as arachnoid cyst, cystic acoustic schwannoma and ependymoma [4].

We report a case of primary intracranial SCC of the CPA, mimicking an acoustic cystic schwannoma and discuss the differential diagnosis with cystic lesions of the CPA.

Case report

All procedures performed in the study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its amendments or comparable ethical standards.

An 80-year-old woman presented with progressive right facial palsy and hypoesthesia of the right hemiface. She underwent a CT scan showing a hypodense extra-axial expansive lesion of the right CPA. MRI with contrast showed a cystic lesion of the right CPA, containing a blood-fluid level and scattered thin septa (Fig. 1A) covered with hemoglobin breakdown products, peripheral enhancement (Fig. 1B) extending along the septa and no diffusion restriction (Figs. 1C and D). The lesion extended through the right internal auditory canal causing moderate mass effect on the right CPA, the cisternal portions of the cranial nerves V, VII, and VIII and on the right posterior communicating arteries without any significant change of the arterial caliber. No previous imaging was available for comparison.

Because of the location of the cystic lesion, its extension through the internal auditory canal and the absence of diffusion restriction, our retained hypothesis was of a cystic schwannoma of the CPA.

The patient underwent partial resection of the lesion about 2 months after the MRI. Pathology showed a keratinizing SCC. Immunohistochemical staining for p16 was negative. A PET/CT was acquired after the surgery and showed a hypermetabolic lesion of the right CPA, but no other lesion potentially representing a primitive tumor. Moreover, clinical examination by the otorhinolaryngologist did not find any lesion compatible with a primary SCC.

The follow-up MRI acquired 5 months later showed a large tumor residual in the right CPA (Figs. 2A and B). The CT scan with contrast performed 1 year later shows tumor progression with bone lysis of the petro-clival junction (Fig. 2C) and invasion of the cavernous sinus (Fig. 2D).

Discussion

Intracranial epidermoid cysts are congenital lesions that develop from ectodermal remnants during neuroembryogenesis representing 0.2%-1.8% of all intracranial tumors and 7% of CPA tumors [5].

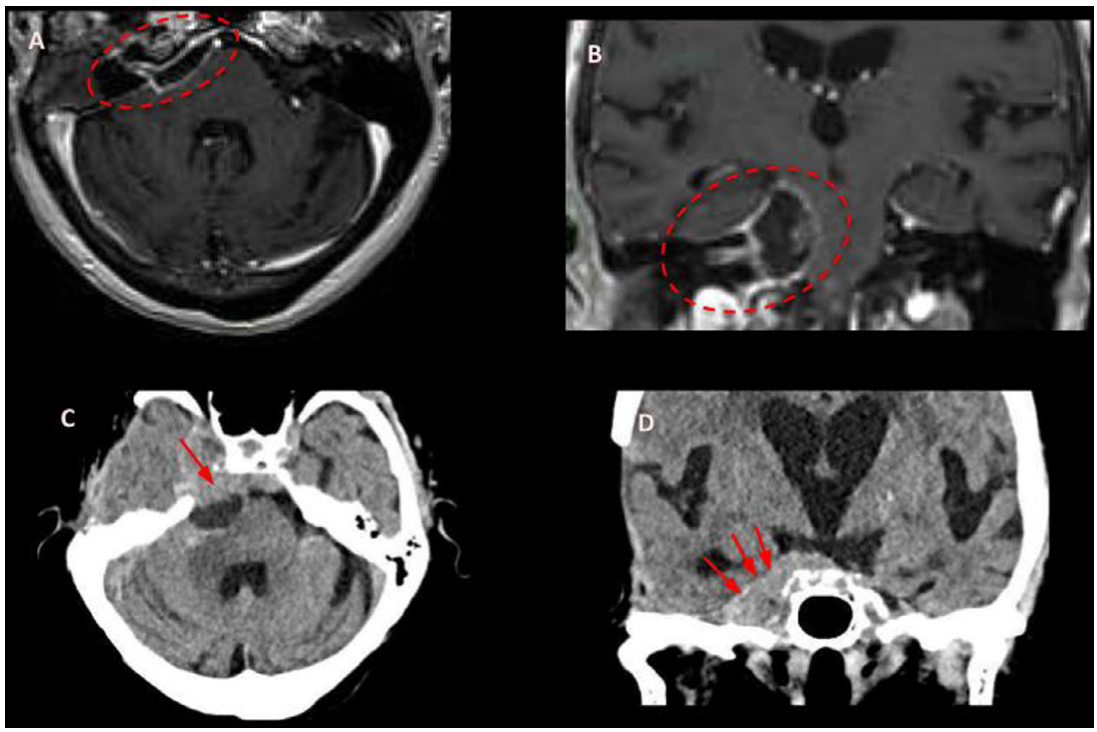

Fig. 2 – The axial (A) and coronal (B) T1-weighted post-contrast images show a large SCC residual in the right CPA after partial resection (red circles). The axial (C) and coronal (D) CT scan images with contrast show marked tumor progression with bone lysis (C, red arrow) of the petro-clival junction. This invades the cavernous sinus (D, red arrows).

Table 1 – Garcia's and Hamlat's criteria for diagnosing primary intracranial SCC.

Garcia's criteria [6]
1. Restriction to intracranial, intradural compartment
2. No invasion or extension beyond the dura, cranial bones, or intracranial orifices
3. No communication with middle ear, air sinuses, or sella turcica
4. No evidence of nasopharyngeal tumor
Hamlat's additional criteria [7]
5. Presence of a benign squamous cell epithelium within the malignant tumor
6. Exclusion of metastatic carcinoma

SCC of the brain is rare and mostly found secondary to metastasis of a primary tumor in the body or direct spread of a head or neck tumor via neural foramens. Malignant transformation of an epidermoid tumor is a very rare entity that occurs in the site of the primary lesion. The underlying mechanism causing malignant transformation is not well known, but chronic inflammation with cystic rupture, subtotal resection of the cyst may be potential factors [3].

According to Garcia et al. and Hamlat et al., true malignant transformation into SCC can only be classified as primary intracranial if it satisfies 6 criteria (Table 1) [6,7].

Diffusion Weighted Imaging (DWI) can usually identify epidermoid cysts from brain parenchyma and surrounding cerebrospinal fluid spaces. Cancerized parts of epidermoid cysts tend to show a lower DWI signal than benign epidermoid cysts; the DWI findings of the carcinomatous mass is, in fact, probably attributable to central necrosis and might be useful to distinguish between benign epidermoid cysts and cancerous part [2,3,9]. In the current case, the cystic lesion of the right CPA did not show any diffusion restriction, whereas it contained a fluid-hemorrhagic level suggesting internal bleeding likely do to necrotic-hemorrhagic changes. We assume that, because of the lack of diffusion restriction, the epidermoid cyst from which the malignant transformation took origin was completely substituted from its cancerized portion.

Enhancement is a sign of malignant transformation of epidermoid cysts which usually present in the form of a ring-enhancing nodule along the residual epidermoid cyst [1,3,8]. However, enhancement is a non-specific sign of malignant transformation because inflammation due to rupture of the epidermoid cysts can manifest with enhancement of the cyst's wall and adjacent edema. In the present case, we observed linear peripheral enhancement along the entire lesion and its internal septa, another finding suggesting possible complete cancerization of an epidermoid cyst.

Lack of diffusion restriction, the peripheral enhancement along the entire lesion and extension into the internal acoustic canal made us initially suspect a cystic acoustic schwannoma with hemorrhagic transformation. Acoustic schwannomas are noncalcifying solid tumors that typically demonstrate high signal on T2-weighted MR images and marked and consistent enhancement after contrast administration. Cystic changes within the tumor and extramural/arachnoid cysts may be associated with acoustic schwannomas. Schwannomas are usually microcystic but may coalesce to form larger cysts. The T1 and T2 signal within the cysts is related to tumoral necrotic material, blood products or colloid fluid. Peripheral enhancement surrounding the intramural cysts after contrast administration may be related to the enhancement of the enveloping tumor and/or the inflammatory reaction to the cyst's content [9].

Other cystic lesions of the CPA without diffusion restriction are arachnoid cysts and ependymoma. Arachnoid cysts are pouchlike intra-arachnoid masses of uncertain origin filled with CSF. Their attenuation and signal intensity match those of CSF almost exactly and show no enhancement [4]. Ependymomas are markedly heterogeneous due to calcification, hemorrhage, cystic components, or necrosis and enhance avidly and irregularly [4]. Ependymomas of the posterior cranial usually arise in the fourth ventricle and its lateral recesses and may extend into the CPA by means of an exophytic component. However, an extra-axial origin directly in the CPA is also possible.

Conclusions

We report a rare case of a primary SCC of the right CPA mimicking a cystic acoustic Schwannoma because of the lesion extending through the internal auditory canal, without diffusion restriction and showing peripheral enhancement surrounding the entire cystic lesion. These characteristics suggest possible complete cancerization of a benign epidermoid cyst and have not been reported in previous studies.

The relevance of this case report lies in the ambiguous presentation of a rare pathology, which must be taken into account in clinical practice in order not to miss the diagnosis of a malignant and rapidly progressive disease.

Declaration of redundant publication

The authors certify that the submitted article will not constitute redundant publication

Submission declaration

The authors declare that this article it is not under consideration for publication elsewhere, that its publication is approved by all authors and tacitly or explicitly by the responsible authorities where the work was carried out, and that, if accepted, it will not be published elsewhere in the same form, in English or in any other language, including electronically without the written consent of the copyright-holder.

Authors' contribution

1. Lina Mliyh: drafting the article and revising it critically
2. Dario Di Perri: acquisition of data, final approval of the version to be submitted.
3. Valeria Onofrj: conception and design of the study, or acquisition of data, interpretation of data, drafting the article or revising it critically for important intellectual content, final approval of the version to be submitted.

Patient consent

This study was approved by our institutional review board after written informed consent was obtained by the patient.

REFERENCES

[1] Zuo P, Sun T, Wang Y, Geng Y, Zhang P, Wu Z, et al. Primary squamous cell carcinomas arising in intracranial epidermoid cysts: a series of nine cases and systematic review. Front Oncol 2021;11:750899. doi:10.3389/fonc.2021.750899.
[2] Kodama H, Maeda M, Hirokawa Y, Suzuki H, Hori K, Taki W, et al. MRI findings of malignant transformation of epidermoid cyst: case report. J Neurooncol 2007;82:171–4.
[3] Ozutemiz C, Ada E, Ersen A, Ozer E. Imaging findings of an epidermoid cyst with malignant transformation to squamous cell carcinoma. Turk Neurosurg 2017;27:312–15.
[4] Bonneville F, Sarrazin JL, Marsot-Dupuch K, et al. Unusual lesions of the cerebellopontine angle: a segmental approach. Radiographics 2001;21:419–38.
[5] Seif B, Pourkhalili R, Shekarchizadeh A, Mahzouni P. Malignant transformation of an intracranial extradural epidermoid cyst into squamous cell carcinoma presented with cerebrospinal fluid leakage. Adv Biomed Res 2017;6:16.
[6] Garcia CA, McGarry PA, Rodriguez F. Primary intracranial squamous cell carcinoma of the right cerebellopontine angle. J Neurosurg 1981;54:824–8.
[7] Hamlat A, Hua ZF, Saikali S, Laurent JF, Gedouin D, Ben-Hassel M, et al. Malignant transformation of intra-cranial epithelial cysts: systematic article review. J Neurooncol 2005;74:187–94.
[8] Cuoco JA, Rogers CM, Busch CM, et al. Intracranial squamous cell carcinoma arising from a cerebellopontine angle epidermoid cyst remnant four decades after partial resection. Front Oncol 2019;9:694.
[9] Tali ET, Yuh WTC, Nguyen HD, Feng G, Koci TM, Jinkins JR, et al. Cystic acoustic schwannomas: MR characteristics. AJNR Am J Neuroradiol 1993;14:1241.