

Available online at www.sciencedirect.com

ScienceDirect

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

Case Report

Adenoid cystic carcinoma of jugular foramen[☆]

Clémentine Maheo*, Ronan Abgrall, Virginie Conan, Julien Ognard, Rémi Marianowski, Jean-Christophe Leclere

Resident Clémentine MAHEO, Hopital Morvan, CHRU de Brest, 2 Avenue Foch, Brest, Bretagne 29200, France

ARTICLE INFO

Article history:

Received 31 January 2022

Revised 21 April 2022

Accepted 24 April 2022

Available online 29 May 2022

Keywords:

Adenoid cystic carcinoma

Parotid tumor

Jugular foramen

Proton beam therapy

ABSTRACT

Adenoid cystic carcinoma is a slowly growing malignant tumor with high local recurrence, perineural and vascular invasion. This tumor might arise from the glands of upper respiratory tract and oral cavity (eg, salivary or serous or mucous). Here we report the case of a 65-year-old woman who was referred to our unit for left retro-auricular radiating pain with trigger points and frontal headache since 6 months. There was no involvement of cranial nerves. Imaging screening using MRI, Positron emission tomography with 2-[fluorine-18] fluoro-2-deoxy-D-glucose, Gallium-68 DOTA-Phe1-Tyr3-Octreotide (68Ga DOTATOC) Positron emission tomography-CT suggested a suspicion of schwannoma or paraganglioma of the jugular foramen. However, the CT-guided biopsy revealed presence of adenoid cystic carcinoma. These warrants performing mandatory histological analysis combined with imaging screening suspicion of schwannoma or paraganglioma.

© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Introduction

ACC is a rare, malignant neoplasm of the head and neck comprising approximately 10% of epithelial salivary gland neoplasms, 7%–18% of parotid malignancies and 20% of all malignant salivary gland tumors (mostly submandibular and sublingual vs 35%–80% of minor salivary glands) [1]. It rises from salivary glands and grows slowly to other tissues and structures, for example, glands sites, oral cavity, throat, paranasal sinuses, larynx, and trachea. This slow growing disease, is often diagnosed at advanced stages (eg, skull base infiltration) concomitant with remote metastasis. ACC can involve

nerves, perineural structures and vessels. Such ACC presents poor prognosis for remote disease. Other significant prognostic factors are tumor's size, primary site, histological structure, level of differentiation, and growth pattern and post-operative histological margin. A <3 cm solid tumor, negative histological margin, lack of perineural and invasion and low level of organ involvement (eg, lack of metastasis) are positive prognostic factors. Consequently, ACC treatment depends on the above prognostic factors assessed through preoperational pathomorphological and histological findings for diagnosis accuracy. First-line treatment is tumor resection with negative histological margin and preservation of organ's functions.

Abbreviations: ACC, Adenoid Cystic Carcinoma.

[☆] Competing Interests: The authors declared that they have no competing interests.

* Corresponding author.

E-mail address: clementine.maheo@chu-brest.fr (C. Maheo).<https://doi.org/10.1016/j.radcr.2022.04.046>1930-0433/© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

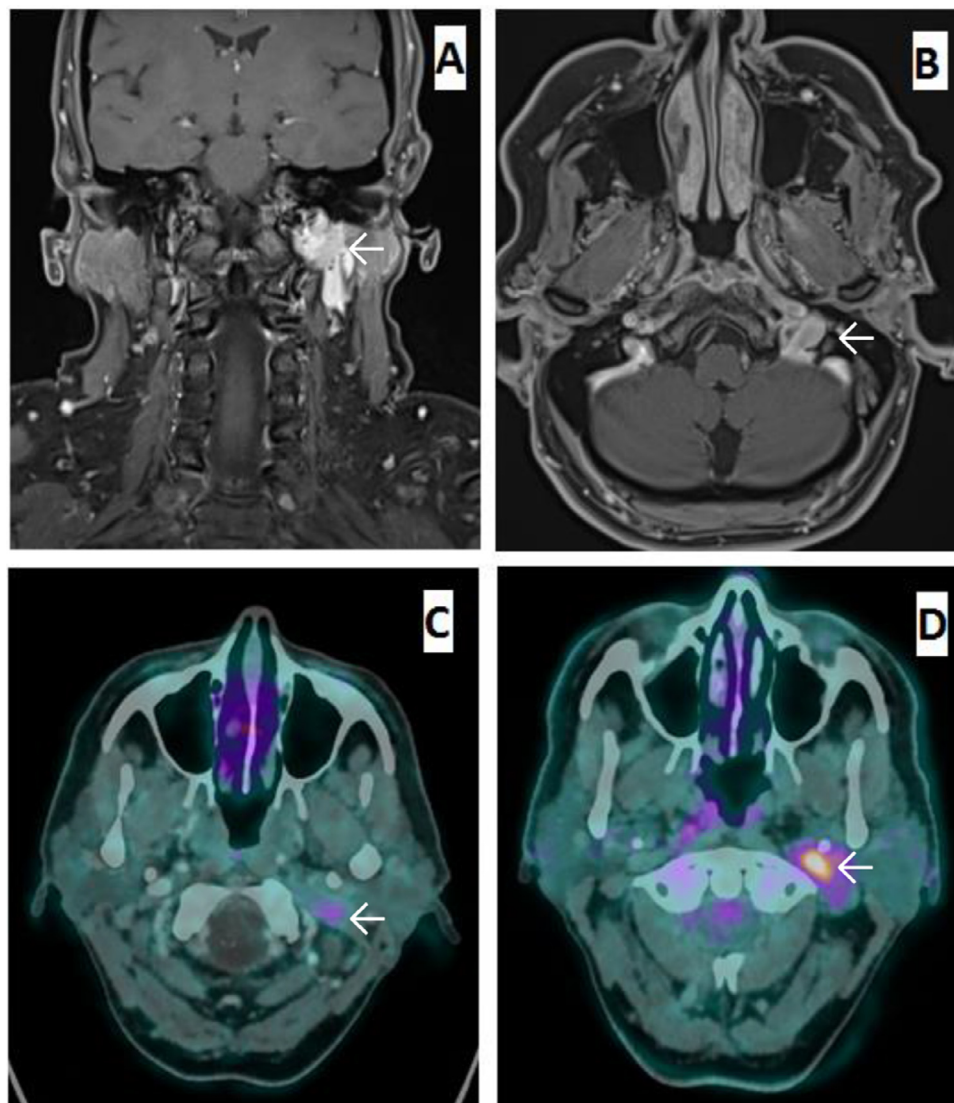


Fig. 1 – (A) Coronal MRI T1, arrow: contrast enhanced showing tumor invading left jugular foramen. (B) Axial MRI T1, arrow: contrast enhanced showing left jugular thrombosis. (C) Axial 68 Ga-DOTATOC PET-CT, arrow: nonspecific mild uptake. (D) Axial 18-FDG PET-CT, arrow: an uptake adenopathy of II B left sector.

Case report

A 65-year-old woman was referred for left retro auricular pain with trigger points and frontal headache since 6 months. She had neuropathic pain, cervical stiffness and pain in the left shoulder.

On physical examination, she presented with a left cervical swelling without adenopathy. Nasofibroscope results were normal. There was no cranial nerves involvement.

The initial diagnostic workup was magnetic resonance imaging (MRI) showing an oval tumor with contrast enhancement measuring 32 × 19 mm and reaching the left jugular foramen (Fig. 1). On the one hand, the oval shape suggested a suspicion of schwannoma. On the other hand, the contrast enhancement was in favor of paraganglioma. However, the above suspicions were inconsistent with the observed infil-

tration of parotid gland, sternocleidomastoid muscle, facial and mixed nerves. Positron emission tomography (PET) with 2-[fluorine-18] fluoro-2-deoxy-D-glucose (FDG) showed an isolated and nonspecific left jugular foramen uptake. Given the suspicion of paraganglioma a Gallium-68 DOTA-Phe1-Tyr3-Octreotide (68Ga DOTATOC) PET-CT was performed. It showed nonspecific mild foramen jugular uptake. Possible pheochromocytoma associated with paraganglioma was ruled out after obtaining a normal urinary catecholamine dosage. The results of ultrasound guided puncture were not conclusive. A CT-guided micro biopsy under general anesthesia was done. The histopathological and immunohistochemistry findings were conclusive of ACC. The tumor proliferation was arranged in trabeculae with hyaline cylinders (Fig. 2). The cells had a scarce and poorly bounded cytoplasm. The nuclei were angular, irregular with small amounts of chromatin. There was tumor proliferation around nerve branches (Fig. 2).

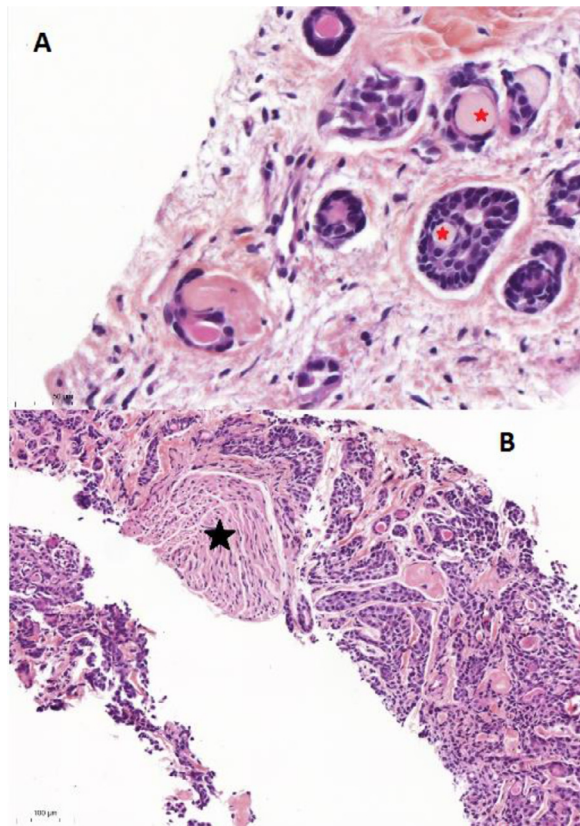


Fig. 2 – (A) Red star: Hyalin cylindroma (x40 HES). (B) Tumor proliferation around nerve (x10 HES) Black star: Nerve branches.

The case was discussed at a multi-disciplinary committee of experts. Radical tumor resection was ruled out due to the infiltration of internal carotid and vertebral artery. A proton beam therapy was performed as first-line treatment. The post-radiation cervical MRI showed tumor regression (39 × 13 mm), volume reduction without contrast enhancement and radiation-induced fibrotic tissue scar, at month 1, month 3 and year 1, respectively.

Discussion

Clinicians and ENT surgeons are faced with the challenges of diagnosis and management of the ACC of the head and neck region due to the neoplasms' (eg, tumor) inaccessibility and most often advanced stage. To date, ACC of the jugular foramen has not been reported in English literature. The most frequently described jugular foramen tumors are paraganglioma, lower cranial nerve schwannomas and meningioma [2]. The least described ones are chordomas, chondrocytomas, endolymphatic sac tumor and metastatic carcinoma [3,4]. The most affected nerves are the trigeminal and facial nerves in ACC of major and minor salivary glands [10].

The most frequent early symptoms are pulsatile tinnitus, hearing loss and cranial nerve involvement among different tumor types [2,3].

This tumor was well circumscribed and followed the course of cranial nerve thus suggesting a schwannoma. However, infiltration of both facial and mixed nerves was not often described. These tumors rise from glossopharyngeal or vagus nerve in more than 90% of the cases [4].

The usual treatment is complete tumor resection to negative histological margins combined with postoperative radiotherapy. Such therapy allows a 10-year local regional disease control rate of 88.2% compared with 36% using radiotherapy alone [8,9].

Proton Beam Therapy shows encouraging results with an 8-year and 2-year local regional disease control rate of 82% and 92%, respectively [7,6].

Consistent with the literature, optimal treatment for our case of non-operable tumor was radiotherapy [5]. Due to the tumor's location and inaccessibility, proton beam therapy was performed in line with the literature for the treatment of skull base involvement to avoid damaging critical adjacent tissues and structures [6,7].

Conclusion

Initial imaging work-ups of our rare case of jugular foramen ACC showed suspicions of schwannoma and paraganglioma. For diagnostic accuracy of such rare cases, preoperative imaging screening (eg, tumor's size, primary site, staging) using PET-FDG and ⁶⁸Ga DOTATOC PET-CT may not be conclusive and should be combined with preoperative pathomorphological data (e.g. histological structure, level of differentiation, and growth pattern).

Patient consent

We obtained the patient consent on January 25, 2022.

REFERENCES

- [1] Bradley PJ. Adenoid cystic carcinoma evaluation and management. *Curr Opin Otolaryngol Head Neck Surg* 2017;25(2):147–53.
- [2] Ramina R, Maniglia JJ, Fernandez YB, Paschoal JR, Pfeilsticker LN, Neto MC, et al. Jugular foramen tumors: diagnosis and treatment. *Neurosurg Focus* 2004;17(2):31–40.
- [3] Guinto G, Kageyama M, Trujillo-Luarca VH, Abdo M, Ruiz-Than A, Romero-Rangel A. Nonglomic tumors of the jugular foramen: differential diagnosis and pronostic implications. *World Neurosurg* 2014;82(6):1283–90.
- [4] 3rd Thomas AJ, Wiggins RH, Gurgel RK. Non paraganglioma jugular foramen tumors. *Otolaryngol Clin North Am* 2015;48(2):343–59.
- [5] Guazzo E, Panizza B. Management of advance adenoid cystic carcinoma infiltrating the skull base: a contemporary review. *J Neuro Oncol* 2020;150(3):419–27.

- [6] Linton OR, Moore MG, Brigance JS, Summerlin D-J, McDonald MW. Proton therapy for head and neck adenoid cystic carcinoma: initial clinical outcomes. *Head Neck* 2014;37(1):117–24.
- [7] Pommier P, Liebsch NJ, Deschler DG, Lin DT, McIntyre JF, Barker II FG, et al. Proton beam radiation therapy for skull base adenoid cystic carcinoma. *Arch Otolaryngol Head Neck Surg* 2006;132:1242–9.
- [8] Guazzo E, Bowman J, Porceddu S, Webb L, Panizza B, et al. Advanced adenoid cystic carcinoma of the skull base – the role of surgery. *Oral Oncol* 2019;99:104466.
- [9] Balamucki CJ, Amdur RJ, Werning JW, Vaysberg M, Morris CG, Kirwan JM, et al. Adenoid cystic carcinoma of the head and neck. *Am J Otolaryngol* 2012;33(5):510–18.
- [10] Frunza A, Slavescu D, Lascar I. Perineural invasion in head and neck cancer. *J Med Life* 2019;7(2):121–3.