Case Report

Adenoid cystic carcinoma of jugular foramen

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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.

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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 arises 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 preoperative 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.

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A 65-year-old woman was referred for left retroauricular 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. Nasofibroscopy 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 infiltration 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 microbiopsy 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. 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-DOTATOM PET-CT, arrow: nonspecific mild uptake. (D) Axial 18-FDG PET-CT, arrow: an uptake adenopathy of II B left sector.
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-operative 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.

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