Clear Cell Carcinoma of Salivary Gland with Intracranial Extension Through Jugular Foramen

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Abstract Clear cell carcinoma not otherwise specified type affecting the parotid gland is rare and extension of this tumor through the jugular foramen presenting as an intracranial space occupying lesion is extremely uncommon. We report a case of clear cell carcinoma of the parotid gland presenting as a left cerebellopontine angle tumor.

Keywords Parotid · Intracranial invasion · Clear cell carcinoma · Jugular foramen

Introduction

The jugular foramen (JF) is situated in the floor of the posterior fossa posterolateral to the carotid canal, between the petrous temporal bone and the occipital bone with both endocranial and exocranial openings [1]. The relationship of the JF to the deep fascial planes of the neck are important because infections and tumors in these spaces often allow lesions distant from the skull base, like in the lower face and neck to pass through it [1]. We describe such an unusual extension of a malignant parotid gland tumor, presenting as an intracranial primary.

Case Presentation

A 41-year-old female presented with progressive headache localized to back of the head with swaying to one side while walking. On examination there was weakness of left cranial nerves, VIII, IX, X and XI but with no facial palsy. There was no history of vomiting, vision changes, signs of meningeal irritation or intracranial tension. The clinical diagnoses suspected were CP angle neural tumor/lymphoma.

MRI plain and contrast revealed a mass lesion of 6 cm in left CP angle with intra and extra cranial components extending across the expanded JF (Fig. 1). There was thickening and enhancement of left internal carotid—VIII nerve complex that was continuous with CP angle mass. The tumor was removed through high cervical approach, a retro mastoid sub occipital craniotomy was done with excision of intracranial part of tumor was done and sent for histopathology.

The biopsy specimen was multiple grey brown bits together measuring about 0.8 cm. Histology revealed a malignant epithelial tumor with cells in syncytial growth pattern and intervening fine vascular network. The cells were large, polygonal with abundant eosinophilic cytoplasm; some with intra cytoplasmic vacuoles compressing the nucleus. The mitoses were few and there was no necrosis (Fig. 2). A diagnosis of a metastatic carcinoma of clear cell type was given.

Four months later the patient came with an oval swelling below the left ear lobe of 3 months duration 4 × 3 cm in size, slow growing, and horizontally oriented, deep to deep fascia with ill defined margins. It was non tender, non pulsatile, smooth surfaced, firm and with restricted mobility. No lymph nodes were identified. She also complained of dysphagia for both solids and liquids and muffling of voice.

Examination of ear and nose were normal except for a bulge in left lateral wall of oropharynx. Ultra sonogram

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Fig. 1 MRI scan showing a mass lesion with intra and extra cranial components extending across the expanded jugular foramen

Fig. 2 a Photomicrograph of an epithelial tumor showing large polygonal tumor cells in syncytial growth pattern, some with intra cytoplasmic vacuoles (H and E $\times 400$). **b** Pancytokeratin positivity in the tumor cells (IHC $\times 400$)

revealed a hypo echoic mass lesion of 3.6×3 cm, adjacent to left parotid gland indenting it.

CECT neck showed a $6.1 \times 4.6 \times 2.4$ cm well defined, mildly enhancing soft tissue mass lesion in left post styloid compartment in carotid space extending into left CP angle cistern through left JF.

Macroscopically they were multiple grey white tissue bits altogether measuring 3×2 cm. Histopathology revealed a tumor composed of large epithelial like cells in sheets and clusters with rich vascular network between the tumor cells. Some of the cells showed large intracytoplasmic vacuoles. There was no necrosis or prominent mitotic activity (Fig. 3). No normal salivary gland tissue was identified in the biopsy. The histological differential diagnoses considered were epithelial myoepithelial

carcinoma, clear cell carcinoma not otherwise specified (NOS) and Paraganglioma.

Immunohistochemistry (IHC) was positive for both Pancytokeratin and EMA. Negative for GFAP, Chromogranin, S100 and SMA. The final diagnosis was signed out as clear cell carcinoma of parotid, NOS type.

Discussion

The tumors that share a common primary site of development in the JF include glomus tumors, neural sheath tumors such as schwannomas and neurofibromas and meningiomas [1]. Clear cell carcinoma NOS type affecting the parotid gland is rare and usually occurs in the minor salivary

Fig. 3 a Photomicrograph of intracranial tumor composed of large epithelial like cells in sheets and clusters with some of the cells showing large intracytoplasmic vacuoles (H and E $\times 400$). **b** Pancytokeratin positivity in the tumor cells (IHC $\times 400$)

glands [2]. Extension of this tumor through the JF and presentation as an intracranial space occupying lesion is extremely uncommon.

Clear cell carcinoma NOS, is a malignant epithelial neoplasm composed of a monomorphous population of cells that have optically clear cytoplasm with standard hematoxylin and eosin stains [2]. This low-grade malignancy has a predilection to arise in the oral cavity, especially in the base of the tongue, of adult women and is also known as hyalinizing clear cell carcinoma (because of their occasional prominent fibro hyaline stroma) [3, 4]. The palate, buccal mucosa, lip, and retro molar and tonsillar areas can also be affected [2]. In our case the parotid gland was involved, which is a rare location for this tumor.

The peak occurrence of this tumor is in patients in the 40–70 year age range [2]. Our patient was a 41-year old woman.

The microscopic appearance of the present tumor was consistent with clear cell carcinoma NOS type of parotid gland. However other clear cell neoplasms that occur either from the native tissue or of metastatic nature such as epithelial myoepithelial carcinoma, and Paraganglioma needed to be differentiated by IHC. The other possible primary sites for clear cell carcinomas were ruled out. As intracranial extension of the tumor was radiologically evident, IHC was done on both parts of the tumor, although they were histologically identical.

IHC was positive for both Pancytokeratin and EMA; negative for GFAP, Chromogranin, S100 and SMA (Figs. 2, 3).

The final diagnosis thus given was clear cell carcinoma NOS type with intracranial extension through JF.

Conclusion

We present this unique case of intracranial extension of a clear cell carcinoma of the parotid gland through the JF. Tumors extending intracranially through this foramen are usually glomus tumors, neural tumors and very rarely salivary gland tumors. We highlight this case to illustrate the point that a clear cell carcinoma of the parotid gland must be considered in the differential diagnosis in a patient presenting with a tumor in the neck extending intracranially through the JF.

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