

Case Report

Extracranial Clear Cell Meningioma in Neck: Report of a Rare Case with Diagnostic Challenge to the Cytopathologist

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Keywords

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- Clear cell variant
- Neck mass
- Extracranial extension

Abstract

Extracranial clear cell meningioma is a rare, distinctive and intrinsically aggressive subset of meningioma affecting younger females. The early recognition of this variant is important as it is associated with a high recurrence rate & mortality. We present a case of a 27 year old female who presented with a swelling in the right infra-auricular region. Fine Needle Aspiration Cytology (FNAC) showed pattern less sheets and focal acinar clusters of polygonal cells with vacuolated cytoplasm. A wide differential diagnosis was considered (metastatic renal cell carcinoma, nodular hidradenoma, acinic cell carcinoma, oncocytoma, parathyroid adenoma & sweat gland carcinoma). A clinico-radiological & histopathological correlation was advised. Biopsy showed sheets & whorls of clear round to ovoid cells which were positive for CK, EMA, Vimentin and NSE. Computed tomography scans were reviewed and showed an intracranial extension of the lesion. Based on the above features, a diagnosis of clear cell meningioma was made. This case highlights the need for incorporating clear cell meningioma as a differential diagnosis whenever a clear cell epithelial neoplasm is encountered on FNAC, considering each differential has a distinctive behaviour, treatment protocol and prognosis.

ABBREVIATIONS

CCM: Clear Cell Meningioma; FNAC: Fine Needle Aspiration Cytology; IHC: Immunohistochemistry; ICC: Immunocytochemistry; CK: Cytokeratin; EMA: Epithelial Membrane Antigen; NSE: Neuron Specific Enolase

INTRODUCTION

Meningiomas are frequently encountered intracranial tumors occurring throughout the craniospinal axis. These tumors are dura-based and usually found along the external surface of brain and rarely within the ventricular system. Ectopic or extracranial presentation of meningiomas is very rare and located entirely outside the craniospinal confines, usually encountered in the head & neck region, skin, lung, retroperitoneum, mediastinum, foot and external auditory canal [1,2]. The clinicians and the cytopathologists may encounter a diagnostic difficulty when these ectopic meningiomas present at uncommon sites

with an uncommon histology. We report a case of Clear Cell Meningioma (CCM) in the posterior triangle of neck, missed on Fine Needle Aspiration Cytology (FNAC) but subsequently diagnosed on histopathology & Immunohistochemistry (IHC). A wide differential diagnosis on FNA smears and the use of Immunocytochemistry (ICC) can help us arrive at a specific diagnosis when one encounters morphological mimics.

CASE PRESENTATION

A 22 year old female presented to the ENT clinic with a gradually progressive swelling in the posterior aspect of neck for the past 7 months. The swelling was associated with pain, muffling of voice and difficulty in swallowing. No history of blurring of vision or neurological deficit was present. On examination, a 6 X 7cm swelling was seen in the right posterior triangle of neck, with a bulge in the oral cavity and pushing the tonsil to the left side (Figure 1A, 1B). FNAC was performed using a

22 gauge needle and yielded a blood mixed aspirate. The smears were stained with May Grunwald Giemsa and showed moderate to high cellularity comprising of round to polygonal cells with moderate amount of vacuolated cytoplasm & minimal nuclear pleomorphism. The cells were arranged in diffuse sheets and focal attempted acinar pattern (Figure 2A, 2B). No necrosis or mitotic figure was seen. A wide differential diagnosis was considered (metastatic renal cell carcinoma, nodular hidradenoma, acinic cell carcinoma, oncocytoma, parathyroid adenoma & sweat gland carcinoma). A clinico-radiological and an excision of the lesion for histopathological correlation were advised. CECT scan revealed a 7 X 6 cm homogenously enhancing soft tissue mass in the right posterior cervical space, displacing the parapharyngeal space anteriorly and superiorly causing destruction of the right temporal bone with intracranial extension (Figure 2C, 2D). Possibility of a malignant neoplastic mass was suggested. A biopsy was performed and it was received in multiple bits together measuring 1 X 0.6 X 0.5 cms. Hematoxylin & eosin stained sections showed round to polygonal cells in a prominent whorling pattern. The nuclei of these cells were round to oval, vesicular with mild to moderate pleomorphism. No mitosis was seen (Figure 3A). In many areas, these cells showed cytoplasmic clearing (Figure 3B). A provisional diagnosis of meningioma was considered and immunohistochemistry for Cytokeratin (CK), Vimentin, Epithelial Membrane Antigen (EMA), Synaptophysin and Neuron Specific Enolase (NSE) was performed. The cells were strongly positive for EMA (Figure 3C), NSE, Vimentin (Figure 3D), focally expressed CK and negative for Synaptophysin. In view of the radiological, histopathological and IHC findings, a definitive diagnosis of clear cell meningioma was given. Surgical excision

was planned with the neurosurgical team; however the patient refused to permit the procedure.

DISCUSSION

Meningiomas account for 24% of primary neoplasms of the central nervous system [3]. Although, most meningiomas are limited to the intracranial space and attached to the duramater, few may extend extracranially, lying completely outside the craniospinal confines (ectopic meningioma). Some intracranial meningiomas may behave poorly and permeate the neighbouring skull to present as skull masses, however maintaining their intracranial connections (secondary meningiomas) [4]. Various mechanisms have been described for the formation of extracranial meningiomas: direct extension of an intracranial mass, origin from arachnoid cells within the sheaths of cranial nerves, origin from embryonic rests of arachnoid cells and metastasising meningioma [5]. Meningiomas usually present in middle or late adult life and show moderate female preponderance (female to male ratio - 3:2) [4].

Although most meningiomas are benign, they have a surprisingly broad spectrum of clinical characteristics and histologically distinct subsets with variable risk of recurrence & prognosis. Classical meningiomas display a wide variety of cytohistological patterns, of which whorl formation is a characteristic feature and evident at least focally. WHO has defined 16 various histologic types of meningiomas based on their risk of recurrence and aggressive behaviour [6]. There are three types of meningiomas according to their grade of malignancy: benign (WHO grade I), atypical (WHO grade II) and anaplastic (WHO grade III) [7].

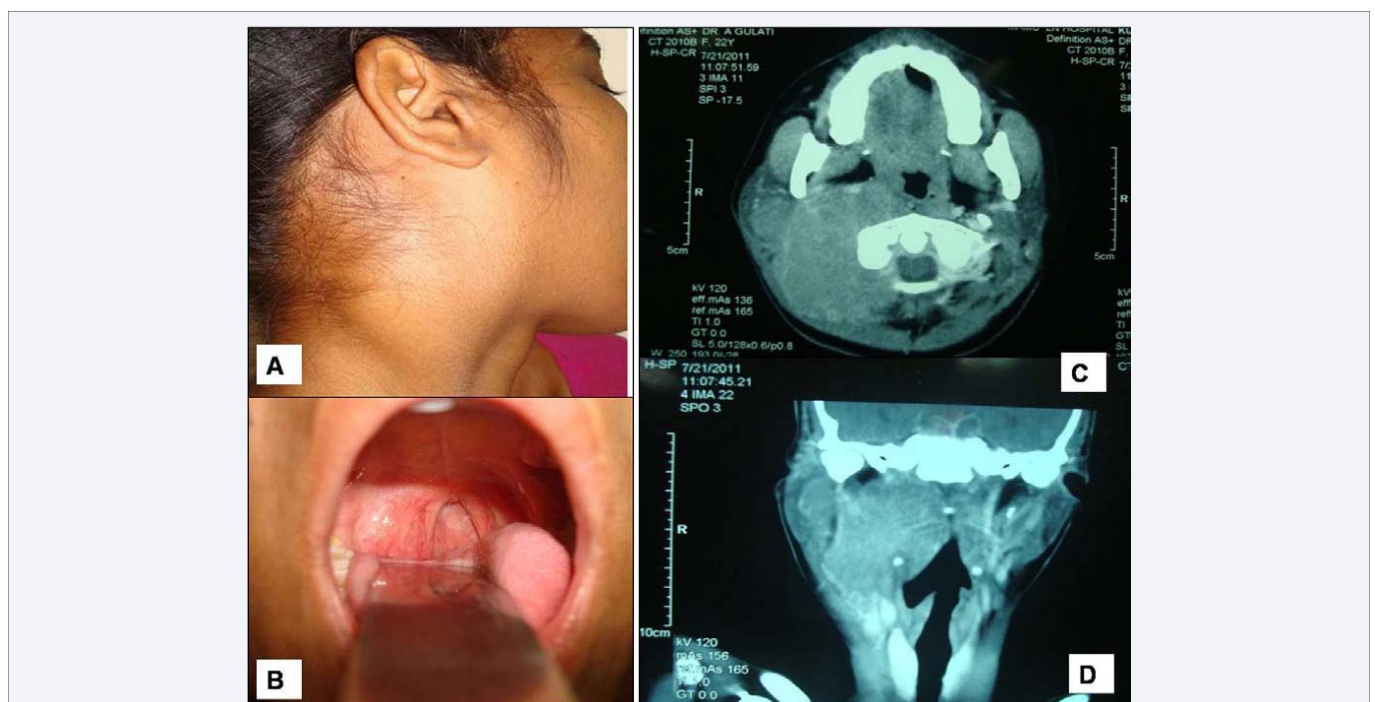


Figure 1 1A- Clinical photograph showing 6 X 5cm swelling in the right infra-auricular region (left)
1B-producing a bulge in the right side of the oral cavity
1C- CECT scan showing temporoparietal space occupying lesion
1D- 7 X 6 cm homogeneously enhancing mass in the right posterior cervical space, displacing the parapharyngeal space

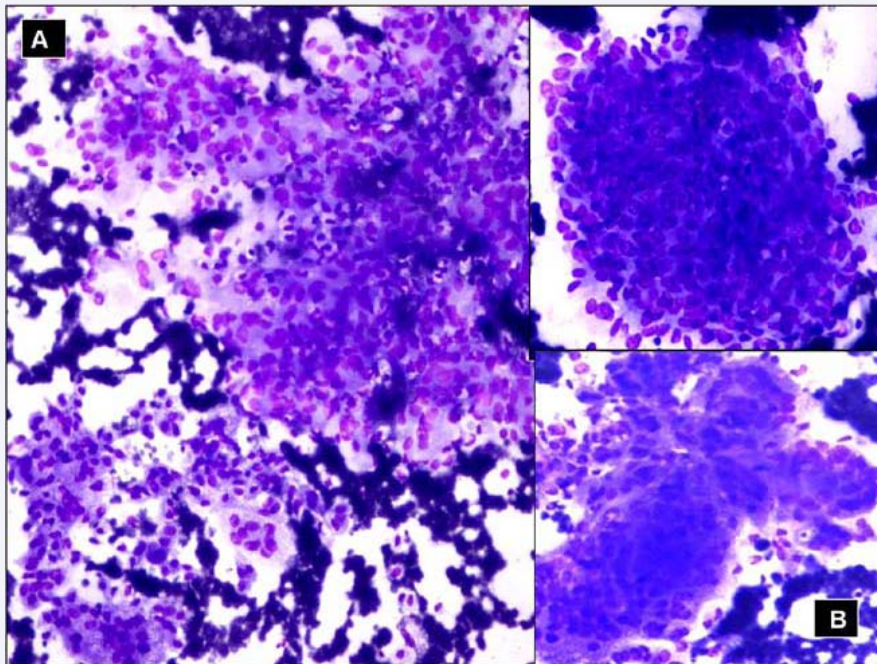


Figure 2 2A - Photomicrograph showing moderately cellular smears comprising of patternless sheets of vacuolated cells with focal attempted whorling pattern (2B). (May Grunwald Giemsa stain, X 400).

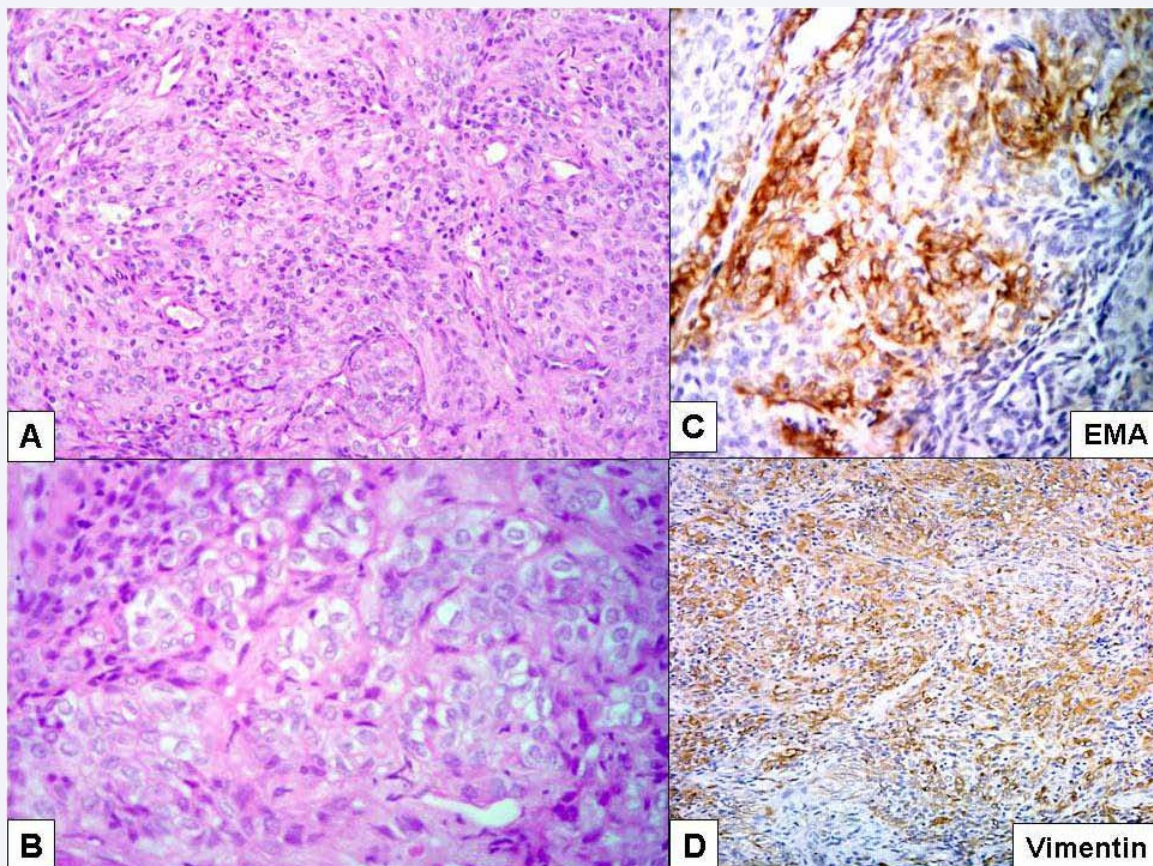


Figure 3 3A, 3B- Photomicrograph showing cells arranged in fascicles and concentric whorls, with prominent clear cell change. (Hematoxylin & eosin stain, X100)

Figure 3C, 3D - Photomicrograph showing tumor cells expressing EMA & Vimentin respectively (Immunohistochemistry, X100)

Clear cell meningioma is a rare subset of meningotheial neoplasms and despite its bland histological appearance shows a potential aggressive behaviour. Zorludemir et al first described this neoplasm in 1995 and it was subsequently listed as a distinct entity in the WHO classification of brain tumors in 2000 [7]. It is associated with a greater likelihood of recurrence and is classified as one of the WHO Grade II tumors. Most CCMs present in the first three decades of life, usually extra axial and often found in the spinal canal, cerebellopontine angle and foramen magnum. In our case, the site of the lesion and the unusual morphology misled the diagnosis. The differential diagnosis of clear cell lesions in the neck includes metastatic renal cell carcinoma, nodular hidradenoma, acinic cell carcinoma, oncocytoma, parathyroid adenoma and myoepithelial variant of sweat gland carcinoma.

Metastatic renal cell carcinoma presents with hematuria, flank pain and a renal mass. Cytology shows large round to polyhedral cells arranged in tubular & acinar pattern, with large vesicular nucleus and prominent nucleoli [4]. Nodular hidradenoma presents as subcutaneous cystic swellings containing clear cells admixed with squamoid cells [4]. Sweat gland carcinomas are usually adenocarcinomas, few of which show signet ring cells. Parathyroid adenomas show a varied morphology comprising of cohesive clusters, disorganised sheets, microfollicles and pseudopapillary fragments. The cells are small, round to oval with pale blue finely vacuolated cytoplasm and a coarse nuclear chromatin [8]. Acinic cell carcinoma shows round to polygonal cells in a glandular or lobular pattern with abundant fine granular cytoplasm. Periodic acid schiff with diastase (PAS- D) highlights the secretory granular cytoplasm of these cells [9]. Oncocytoma is characterised by cohesive clusters and sheets of oncocytes-large cells with abundant granular, eosinophilic cytoplasm with central too eccentric nucleus and prominent nucleolus [10].

The treatment for clear cell meningioma is surgical resection with radiotherapy being reserved for recurrent cases.

To conclude, this report documents an uncommon variant of meningioma presenting as a neck mass and posing a diagnostic difficulty for both the clinician and cytopathologist. It also emphasises the need for incorporating clear cell meningioma as a differential diagnosis whenever a clear cell epithelial neoplasm is encountered on FNAC. Although this tumor is

unusual, the characteristic cytological features and appropriate immunocytochemistry if applied can help establish an early definitive diagnosis.

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