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A rare case of left cerebellopontine angle endolymphatic sac tumour. Illustrative case

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Abstract

Endolymphatic sac tumours (ELST) are rare and slow-growing neoplasms that develop out of the endolymphatic sac of the temporal bone. Even though histologically low grade, they show locally aggressive behaviour, and may affect important neurovascular structures of the cerebellopontine angle. These tumours can present with auditory or vestibular dysfunction, and they are usually radiologically indistinguishable from other cerebellopontine angle lesions.

We present a rare case of a left cerebellopontine angle mass, which was a left endolymphatic sac tumour based on histopathological examination and immunohistochemical analysis. Under microscopy, the arrangements of the papillae of the cells demonstrated cuboidal to columnar eosinophilic to clear cytoplasmic cells, with no mitosis or necrosis. Immunohistochemistry showed focal positivity for CA IX, AE1/AE3, and CK7, and negativity for the rest (GFAP, SOX10, PAX8, TTF1, EMA, and CK20). The Ki-67 (MIB-1) labelling index indicated low proliferative activity at 1-2%.

This tumour should be recognised, as its clinical and radiological presentations may be similar to those of other lesions of the cerebellopontine angle, and it is extremely rare. The diagnosis and surgical care should be done early to avoid local invasion and neurological complications.

Keywords
Endolymphatic sac tumour,
Cerebellopontine angle,
Temporal bone tumour

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Introduction

ELSTs are infrequent epithelial neoplasms that start in the endolymphatic sac and duct of the posterior part of the petrous temporal bone. They are low-grade adenocarcinomas of local aggression, and these tumours were identified as a separate clinicopathological entity in the late twentieth century [1]. Though histologically low-grade, ELSTs may severely destroy the surrounding bone structures and may spread to the cerebellopontine angle and other nerves attached at the cranial end [2]. The cerebellopontine angle (CPA) is an anatomically complicated area found between the cerebellum and the pons. Numerous tumours may occur within this area, with the most frequent being vestibular schwannomas, meningiomas, and epidermoid cysts.

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Endolymphatic sac tumours constitute a very small percentage of lesions in this area, making them difficult to diagnose [3].

Patients with ELST commonly have hearing loss, tinnitus, vertigo, imbalance, or facial nerve dysfunction due to the proximity of the involved cranial nerves and vestibular apparatus. There are cases where ELST is related to Von Hippel-Lindau disease, a hereditary cancer syndrome that leads to the occurrence of multiple benign and malignant tumours in various organs [4]. Radiological examinations, such as CT and MRI, are helpful in determining the site and size of the lesion, but a definitive diagnosis is often made by histopathological and immunohistochemical tests [5]. Due to the infrequency of this tumour and its similarity to other CPA lesions, this can be a difficult diagnosis. This article describes an infrequent case of a left cerebellopontine angle endolymphatic sac tumour, presenting the clinical picture, histopathological diagnosis, immunohistochemical characteristics, and differential diagnosis.

Case Presentation

A patient presented with clinical signs indicative of a space-occupying lesion in the left cerebellopontine angle, describing a gradually increasing neurological dysfunction that prompted further clinical and radiological examination. Imaging examinations, including radiological examination of the brain, showed a clear mass lesion in the left cerebellopontine angle [6]. Radiological observation (MRI brain with contrast) raised suspicion of a neoplastic lesion arising from structures in the posterior cranial fossa (Figures 1 and 2). There was a large, heterogeneously enhancing mass on T1 contrast located at the cerebellopontine junction with upstream ventricular dilation and hydrocephalus. T1 contrast sequences showed heterogeneous enhancement with interspersed areas of non-enhancement, likely representing necrosis or cystic changes.

Based on the location and radiological appearance of the lesion, a surgical procedure was conducted first to place a left-sided ventriculoperitoneal shunt to relieve the CSF pressure, followed by definitive surgery of the tumour. The patient then underwent a retromastoid retrosigmoid craniotomy with gross total excision of the tumour with preservation of the facial nerve. The excised tissue was sent for histopathological assessment.

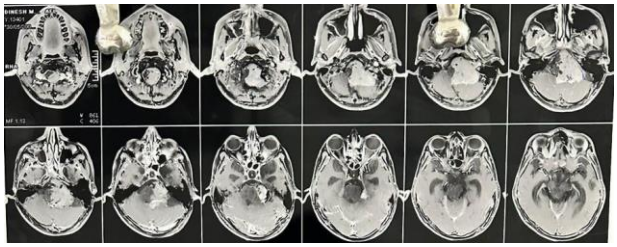

Figure 1. MRI brain, T1 contrast, axial images.

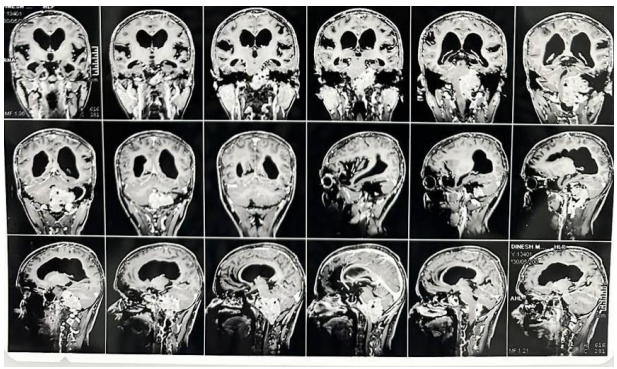

Figure 2. MRI brain, T1 contrast, sagittal and coronal images.

Pathology

Hematoxylin and eosin-stained sections from microscopic analysis showed that the tumour tissue was arranged predominantly in a papillary architecture. The papillae were covered with cuboidal to columnar epithelial cells having moderate eosinophilic to clear cytoplasm. The nuclei were fairly homogeneous, with little pleomorphism [8]. Notably, there was no evidence of mitotic figures or tumour necrosis, and as a result, low proliferative activity and relatively slow biological behaviour were found. Foamy macrophages were found in the tumour stroma, sometimes seen with papillary epithelial tumours. These histomorphological characteristics led to the consideration that an epithelial tumour could develop out of the endolymphatic sac.

Immunohistochemical Findings

The characterisation of the tumour, as well as its differentiation from other neoplasms that can occur in the cerebellopontine angle region, was performed using immunohistochemistry (IHC). Since there are several tumours in this anatomical location with

similar morphological features, a panel of immunohistochemical markers is necessary to make the proper diagnosis. The source of the tumour cells, and the exclusion of other possible entities based on a series of epithelial, glial, and neural crest markers, was determined in the present case [9]. The tumour cells were focally positive for CAIX, AE1/AE3, and CK7. The confirmation of an epithelial character of the tumour is achieved through positivity of AE1/AE3, which highlights cytokeratin, usually produced by epithelial tissues. CK7 is also a marker of epithelial differentiation and is common in tumours of glandular or ductal epithelium. Focal CAIX expression also supports epithelial origin, and has been reported in some papillary tumours, including tumours of the endolymphatic sac. All these findings suggest a tumour diagnosis based on epithelial structures of the temporal bone.

The tumour cells were negative for GFAP, SOX10, PAX8, TTF1, EMA, and CK20. The lack of GFAP staining was used to rule out glial tumours, which are expected to be strongly positive for GFAP. SOX10-negative staining was used as evidence against a schwannoma or other neural crest-derived tumour, which is usually prevalent in the cerebellopontine angle. Correspondingly, the absence of PAX8 expression helped eliminate metastatic renal cell carcinoma as a serious differential diagnosis given the similar histological features. TTF1-negative staining eliminated potential metastatic tumours of pulmonary and thyroid origin, and CK20-negative staining provided additional evidence of the lesion's non-gastrointestinal epithelial nature. Further immunohistochemical analysis showed that a few tumour cells were positive for p53, indicating little change in the p53 pathway [10]. In the most proliferative regions, the MIB-1 (Ki-67) labelling index was approximately 1-2%, indicating a low proliferation rate. This low Ki-67 index is consistent with the sluggish biological behaviour typically associated with endolymphatic sac tumours.

Pathological Impression

The combined histomorphology and immunohistochemical profile supported the conclusion of a left cerebellopontine angle space-occupying lesion, compatible with an endolymphatic sac tumour. Clinico-radiological correlation was suggested to support the pathological diagnosis and further clinical management.

Discussion

Endolymphatic sac tumour was first described by Hassard, and the term low-grade adenocarcinoma of endolymphatic sac origin was coined by Heffner in 1989. It presents in the 4th to 5th decade of life, with a female preponderance. It most commonly presents with hearing loss, vertigo, and tinnitus, all of which were seen in our patient. Other cranial nerve palsies and cerebellar involvement can also be present. It is mainly a benign lesion with no metastasis.

ELSTs are infrequent epithelial neoplasms that develop out of the endolymphatic sac epithelium situated in the posterior section of the petrous temporal bone [11]. These tumours are known to be locally aggressive despite being usually regarded as low histological grade. ELSTs can gradually erode the bone structures around them and can also spread to other areas, such as the cerebellopontine angle. Due to this invasive growth pattern, patients can experience neurological symptoms as a result of compression or involvement of the surrounding cranial nerves and vascular structures. The pathogenesis of ELST is not well understood. These tumours can be sporadic or associated with von Hippel-Lindau disease, a hereditary tumour syndrome resulting from mutations in the VHL tumour suppressor gene (autosomal dominant). Individuals with this condition are predisposed to benign and malignant tumours in various organs, including the central nervous system, kidney, pancreas, and adrenal glands. ELSTs in patients with this syndrome are usually more likely to develop at an earlier age and can be bilateral [12]. In the case of ELST, close clinical observation is commonly recommended to exclude related systemic abnormalities.

Histopathologically, ELSTs are often papillary, cystic, or glandular. Cuboidal-to-columnar eosinophilic or, at times, clear cytoplasmic epithelial cells usually line the papillae. The tumour cells tend to show mild nuclear atypia and a low level of mitosis, indicating their comparatively sluggish biological activity. Another typical feature is the presence of foamy macrophages in the tumour stroma [13]. These microscopic findings were consistent with those of the current case, in which the tumour showed a papillary appearance with cuboidal to columnar cells, with foamy macrophages present.

Immunohistochemistry is vital for distinguishing ELST from other tumours found in the cerebellopontine angle. ELSTs typically express epithelial markers, such as cytokeratin (e.g., CK7), and pan-cytokeratin markers, such as AE1/AE3. Conversely, neural crest markers such as SOX10 and S100 protein are commonly strongly positive in more common cerebellopontine angle tumours such as vestibular schwannomas [14]. Correspondingly, glial tumours of the central nervous system usually express glial fibrillary acidic protein (GFAP). In the current case, the tumour cells were positive for CK7 and AE1/AE3 but negative for GFAP and SOX10, thus ruling out glial and schwannian tumours.

The other significant differential diagnosis is metastatic clear cell renal cell carcinoma, which can present with similar histological characteristics, such as clear cytoplasm and papillary architecture [15]. Nonetheless, PAX8, a marker of renal epithelial differentiation, is typically highly expressed in metastatic renal cell carcinoma. In this case, the absence of PAX8 staining was used to rule this out. ELSTs are often radiologically manifested as destructive lesions of the petrous temporal bone and may show heterogeneous enhancement on magnetic resonance imaging (MRI). Nevertheless, imaging results are not always sufficient for a conclusive diagnosis, as many cerebellopontine angle tumours can appear similar on radiographs. As such, histopathological study and immunohistochemical staining remain crucial for accurate diagnosis of the tumour.

Excision via surgery is regarded as the first-line and most effective treatment for ELST. Total ablation of the tumour is essential for preventing local recurrence and further damage to adjacent structures [16]. Early diagnosis and proper surgical treatment are particularly important, as progressive tumour growth may result in severe neurological impairment due to compression of other cranial nerves, in particular the facial nerve (cranial nerve VII) and the nerve of the inner ear (cranial nerve VIII). Recurrence and optimal patient outcomes can thus be achieved through constant clinical follow-up.

Conclusion

Endolymphatic sac tumours (ELSTs) are infrequent tumours that develop out of the endolymphatic sac of the temporal bone and can manifest as masses in the cerebellopontine angle region. These tumours can readily be confused with more common lesions, such as vestibular schwannomas or meningiomas, because of their infrequent occurrence and similarity in clinical and radiological appearance. This makes precise diagnosis difficult and necessitates a multidisciplinary approach that incorporates clinical examination, radiological imaging, and meticulous pathological examination.

Histopathological analysis is very important for revealing the typical morphological characteristics of ELST, such as papillary architecture and eosinophilic or clear cytoplasmic epithelial cells. Immunohistochemical testing can also be useful in confirming the diagnosis by demonstrating epithelial biomarkers and the absence of markers associated with other neoplasms that can develop in the cerebellopontine angle. The integrated histomorphological and immunohistochemical profile in the current case was consistent with an endolymphatic sac tumour.

ELST should be recognised early, as even with a relatively low-grade histological appearance, these tumours may exhibit locally aggressive behaviour, destroying surrounding bone architecture and compressing surrounding cranial nerves. Surgical management and prompt diagnosis are therefore necessary to prevent disease progression and to preserve neurological function. Careful clinico-radiological correlation and extended follow-up are advisable to ensure accurate diagnosis, guide treatment, and detect potential recurrence.

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