ISSN: 2581 – 3633
PubMed - National Library of Medicine - ID: 101745081

International Journal of Medical Science and Advanced Clinical Research (IJMACR)

Available Online at:www.ijmacr.com

Volume – 7, Issue – 6, November - 2024, Page No. : 17 – 19

A Rare Case Report of Endolymphatic Sac Tumour

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How to citation this article: Dr. Sudindra SG, Dr.Madhusudhan HV, Dr.Praveen K.S, Dr.Vara Prasad BM, "A Rare Case report of Endolymphatic Sac Tumour", IJMACR- November - 2024, Volume – 7, Issue - 6, P. No. 17 – 19.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Here we present a case of tumour at the cerebellopontine angle in a 38-year-old man. He presented with reduced hearing in left ear with ataxia. On examination senoneural hearing loss and mild lower motor neural facial palsy was noted and was in favor of acoustic neuroma. MRI brain showed findings of left CP angle lesion extending into subtemporal area, with possibiity soft suckable vascular lesion was given. Surgical excision was performed and the diagnosis of the endolymphatic sac tumour was made. Endolymphatic tumour is a low grade adenocarcinoma that originates from the endolymphatic sac. The definitive diagnosis requires a combination of clinical features, radiological finding and pathological correlation.

Keywords: Endolymphatic sac tumors, Von Hippel-Lindau, Cerebellum, Adenocarcinoma

Introduction

Endolymphatic sac tumors (ELSTs) are very rare nonmetastasizing neuroectodermal tumors arising from the proximal, intrapetrous portion of the endolymphatic sac[1] and it has destructive behavior which occurs in the skull base and invades the posterior petrous bone, the mastoid and cerebellopontine angle (CPA). The first report concerning the ELST was in 1984[2],butitisin1989 that Heffner characterized ELST as a tumor deriving from the endolymphatic sac epithelium of the internal ear, referring to it as a "low-grade adenocarcinoma"[4]. It is synonymous with Heffner tumor, low grade adenocarcinoma of endolymphatic sac origin[2].It is rare in general population and has an association with Von Hippel-Lindau (VHL) disease [3].

Case Details

A 38 year-old man presented with chief complaints of reduced hearing in left ear with ataxia features, since 3

months and was in favor of acoustic neuroma. Otological examination revealed sensoneural hearing loss in left ear associated with left sided lower motor neuron facial paralysis. There was no history of trauma or surgeries in the past. The right ear was normal. Neither the symptoms nor a family history of VHL disease were found in the patient. On MRI, a 3.3×2x2 cm sized multi-lobulated extra-axial mass was detected in the left CPA, compressing the cerebellum. The radiological diagnosis at that time was vascular lesion. Microscopic examination of the excised material revealed on hematoxylin and eosin staining showed fragments of papillary tumour lined by pseudostratified columnar epithelium displaying round to oval nuclei, fine chromatin and moderate amount of eosinophilic cytoplasm. At places scattered glandular structures of varying calibre are observed. The tumor infiltrates bony trabeculae and adjacent skeletal muscle tissue. No evidemce of mitotic figures, atypia or necrosis seen. Immunohistochemical study was performed and the tumour showed positivity towards cytokeratin, CK 7, vimentin and MIB -1 labelling index was 1%. These findings support diagnosis of an endolymphatic sac papillary tumor.

Figure 1:

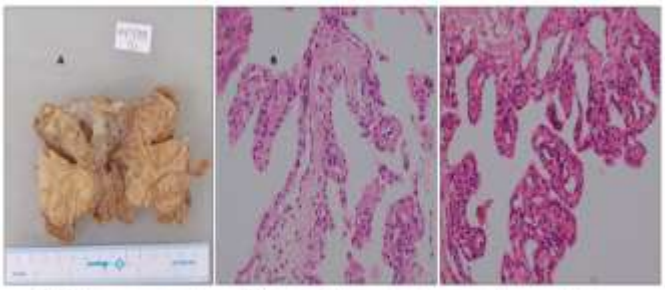

Fig 1 : A: showing gross picture of tumour. B(10x) & C(20x) H&E stain: showing fragments of papillary tumour lined by pseudostratified columnar epithelium displaying round to oval nuclei , fine chromatin and moderate amount of eosinophilic cytoplasm

Figure 2:

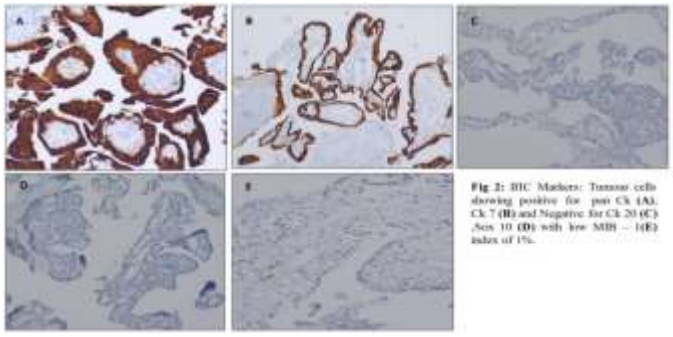

Fig 2: IHC Markers: Tumour cells showing positive for pan Ck (A), Ck 7 (B) and Negative for Ck 20 (C) ,Ses 10 (D) with low MIB – 1(E) index of 1%.

Discussion

The endolymphatic sac is an anatomic structure that lies along the posterior and medial petrous bone. The proximal portion is flared and lies in the vestibular aqueduct, covered by bone. The distal portion lies outside the vestibular aqueduct, between the dural layers. The endolymphatic sac is a network of interconnected sinuses and connecting ducts[1]. Endolymphatic sac tumor was first described byHassard[2]. Subsequently, the term low grade adenocarcinoma of endolymphatic sac origin was coined by Heffner in 1989 who found similarities in the anatomical location and histological features of the normal endolymphatic sac to this tumour [4]. ELST is reported to be more common in females with a mean presenting age of 45 years. Sensorineural hearing loss, tinnitus and vertigo are the typical presenting symptoms in patient with ELST. Cranial nerve paralysis including facial nerve palsy and cerebellar disorders develops as the tumor extends into the jugular foramen and cerebellopontine angle[5].This neoplasm behaves as a slow growing tumor which is locally invasive and exhibits bonedestruction[3].

The majority of ELST are sporadic while around 15% of patients have an autosomal dominant inherited disorder, von Hippel-Lindaudisease. Patients with VHL disease have a germ line mutation in the VHL tumor

suppressor gene that is responsible for their genetic susceptibility to various neoplasms. Apart from ELST, VHL disease is associated with haemangioblastoma of the central nervous system, choroid plexus papilloma, renal cell carcinoma, phaeochromocytoma and papillary cyst adenoma of the epididymis[3]. Our patient did not have any other associated abnormality on thorough examination. The differential diagnosis of ELST includes other destructive lesions of the temporal bone such as paraganglioma, meningioma, hemangiopericytoma, and metastases. Radiologically, hyper vascular mass near the temporal bone is strongly suggestive of a paraganglioma. Typical Zellballen pattern along with immuno positive for chromogranin and synaptophysin distinguishes it from ELST. Other differential image diagnoses for ELST include acoustic neuroma, which could lead to the destruction of the petrous apex of the temporal bone andcompressingthecerebellum[7].

The best treatment choice is the total removal of the lesion, which may sometimes necessitate sacrifice of cranial nerves, because total resection of the advanced tumors may be impossible due to the anatomic complexity[8],so postoperative radiotherapy was suggested as adjuvant therapy in most cases. Owing to their locally aggressive nature and difficult to extirpate surgically, local radiotherapy should be applied in time, depending on surgical excision status. In addition, radiotherapy may also be suitable as a salvage treatment in recurrent endolymphatic sac tumors[9].

Conclusion

ELST should be taken into consideration for differential diagnosis of CPA tumors. Detailed clinical and radiographic evaluation is required for appropriate management in every case. Radical excision is feasible using appropriate surgical approach. Early diagnosis, surgical excision and long-term regular follow-up may constitute an efficacious management.

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