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Case Report

# Endolymphatic Sac Tumor: Case Report and Review

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ABSTRACT: OBJECTIVE: To report a case of endolymphatic sac tumor which has local aggressive pattern. Methods Design: A Case Report. Setting: Tertiary Referral Centre. Patients: One. Result: The patient presented with a left otorrhoea and left facial nerve palsy with worsening past one year in 2008. Physical examination showed left facial nerve palsy grade V and reddish middle ear mass seen. Computed tomography (CT) scan and Magnetic Resonance Imaging (MRI) showed a cerebellopontine angle tumor. Tumor excision via intracranial approach performed and histopathology report consistent with endolymphatic sac tumor (ELST). During surveillance follow-up a year later, this patient developed tumor recurrence and translabyrinthine excision was performed. He has been completed radiotherapy 30 fraction with 60 Gry. Post surgery he is well and still under yearly follow up.
Key Words: Endolymphatic sac tumour, vestibular aqueduct, pathology.

INTRODUCTION: Endolymphatic sac tumor is a locally aggressive tumor which results in late clinical manifestations with expansive mass invading the temporal bone and posterior fossa. Surgery and post-operative radiotheraphy are the main treatments modality. Follow up of patients have to focus on both local recurrence and metastases

CASE REPORT: A 40 year old man presented with left facial nerve palsy with left otorrhoea for one year and gradually worsening in symptoms. He presented with reduced hearing over left ear with presence of tinnitus for few months with no history of vertigo. Physical examination showed left facial nerve palsy grade V and left reddish middle ear mass seen. Nasoendoscopy showed normal findings of nasopharynx. Pure tone audiometry showed left moderate to severe mixed hearing loss with right normal hearing. The post contrasted computed tomography scan (CT scan ) showed a mass at left Cerebello-Pontine (CP) angle tumor with heterogeneous enhancement of the solid component and enhancement of the wall of the cystic component. (Figure 1) There were calcifications within the solid component. The petrous temporal bone was irregular with lytic lesion. Magnetic Resonance Imaging (MRI) showed left CP angle tumor measuring 3x2cm and extends into the left middle cranial fossa 2x2x1cm, enhancement of adjacent durra. (Figure 2) Excision of tumor done via craniotomy was performed successfully. Post operative period was uneventful and he was discharged well. Histopathological examination showed endolymphatic sac papillary tumor. During follow-up surveillance at one year later, MRI showed distorted left temporal bone signal density with presence of residual tumor at the CP angle component measures 2x0.9x1.5cm and extends into the left middle cranial fossa measures 1.4x1.8x1.6cm. He underwent tumor excision via translabyrinthine approach and intraoperative findings showed a mass at the left CP angle eroding the posterior wall of the middle ear and sigmoid sinus. Histopathology report showed remnant of endolymphatic sac tumor. He completed radiotherapy 30 fraction with 60 Gry with no complications.

DISCUSSION: Initially ELST described as a low grade papillary adenocarcinoma, the histological appearance and apparent lack of metastatic potential of these tumours has convinced some to reclassify them as benign papillary adenomatous tumours. However to date, it is classify as a locally aggressive tumour with malignant features and possibility of drop metastases. The high overall survival following surgical resection, despite locally aggressive behaviour, is likely due to the underlying benign histology of the tumour. The first report of drop metastasis to the spine is by Bambakidis in 2005[1]. The second report of such occurrence reported by Bambakidis is by Tay KY which shows the need for close follow up and assessment in patients diagnosed with ELST[2]. Endolymphatic sac tumor (ELST) is a locally aggressive tumor which shows destruction to adjacent area. The immunochemical analysis of these tumors usually reveals cytokeratin, vimentin, epithelial membrane antigen and less frequently S-100 protein and neuron specific enolase. In sporadic cases genetic analysis for Von Hippel-Lindau disease should be considered. Local excision is curative for end lymphatic sac papillary tumors. The currently favored method of treatment consists of excision and long term follow up. The treatment of choice is a radical resection, although complete resection is impossible in most of the cases[3]. The role of adjuvant radiotherapy as treatment is controversial[4]. Endolymphatic sac tumors could be controlled by surgery, radiotherapy or conservative management and strongly influenced by tumor stage and co-morbidity[5]. It is important to make a distinction between ELST and the more benign middle-ear adenomas, since this leads to a different treatment and prognosis. ELST frequently invades the surrounding structures and extends intracranially. The long history of chronic otorrhoea in this patient with the presence of reduced hearing and tinnitus warrants an earlier investigations, namely CT scan or MRI for further evaluation to role out other causes of chronic discharging ear namely chronic suppurative otitis media with cholesteatoma. The nature of local aggresiveness of this

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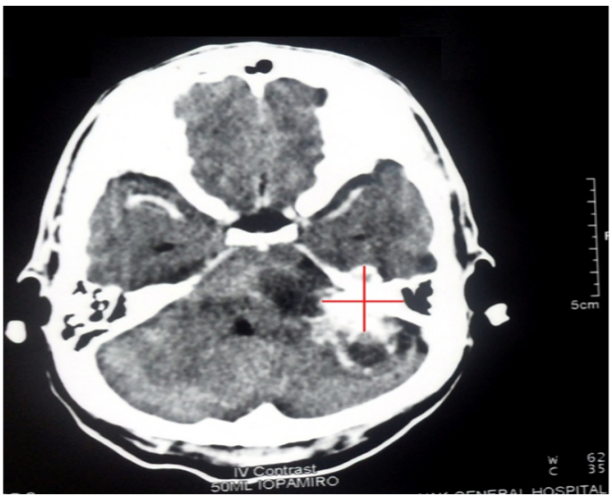


Figure 1: Axial cut of post contrasted computed tomography scan (CT scan ) shows a mass at left Cerebello-Pontine (CP) angle with heterogenous enhancement of the solid component and enhancement of the wall of the cystic component.

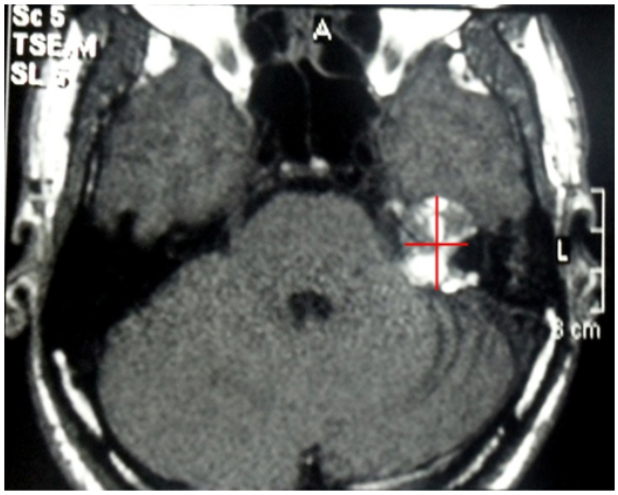


Figure 2: Magnetic Resonance Imaging (MRI) of left CP angle shows tumor measuring 3x2cm and extends into the left middle cranial fossa 2x2x1cm, enhancement of adjacent durr.

tumour was presented in this patient whereby it eroded the posterior wall of the external ear canal. The diagnosis dilemma was at the time of presentation there is presence of reddish mass in the middle ear which could also be glomus jugulare having in mind of the CP angle tumour could be acoustic neuroma which has different entity and management.With regards to this patient the remnant of tumour was excised via translabyrinth approach compromising the hearing as to ensure complete tumour resection. He was given radiotherapy post operation in order to prevent metastases. Despite given post operative radiotherapy patient still has to be on surveillance follow up and spinal imaging should be considered in patients complaints of back pain, cauda equina and radicular symptoms[2]. In patients undergoing gamma knife surgery for recurrence tumour still shows neurologically stable although not disease free still warrants a close surveillance of symptoms[6]. To date role of radiotherapy is still controversial, 50% efficacy has been reported for the treatment of ELST with only radiotherapy, rate of cure of 90% has been reported for complete ELST excision without radiation[7]. With the possibility of residual tumour it is important to look at the origin of this tumour for complete resection to avoid recurrence and metastases. The precise location of the origin of tumour is at the vestibular aqueduct of the endolymphatic sac. In a reported case series by Russell RL et al in 2009 it correlates the HPE and the CT scan and MRI findings for and made a conclusion that the tumour is confined in the intraosseous portion (vestibular aqueduct) of endolymphatic sac and the radiological investigations showed the epicentre is at the same area[8]. Special attention should be consider in excision of tumour in this area as it will prevent the recurrence of disease. In patients with excellent preoperative hearing and a small ELST, such a hearing conservation approach may be warranted. However, the completeness of tumor resection should not be compromised for the sake of hearing conservation. In some tumours, total resection cannot be achieved without risk of catastrophic loss of function or death, and in these patients subtotal resection may be warranted. Overall survival characteristics for all reported cases of ELSTs are: 74% of patients have no evidence of disease, 20% patients alive with disease and 4% died of disease. Stereotactic radiotherapy has shown no increased benefit above standard fractionated radiotherapy in survival or recurrence rates, and subtotal resection followed by stereotactic radiotherapy has uniformly resulted in tumor regrowth[9,10].

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