Endolymphatic Sac Tumor: Rare Tumor of Internal Ear.  
A Case Report and Literature Review

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ABSTRACT

Papillary tumors of the temporal bone are rare and aggressive neoplasms. Recently described, these tumors had initially a presumed middle-ear origin. Only recently, convincing anatomic, morphological and immunohistochemical arguments exist for an endolymphatic sac origin (inner-ear origin). We report one case of endolymphatic sac tumor. These tumors can be encountered sporadically or in Von Hippel-Lindau disease. They classically grow very slowly, resulting in late clinical manifestations with expansive mass invading temporal bone and extending in posterior fossa. Radiologically, these endolymphatic sac tumors can mimic metastatic carcinoma, paraganglioma, or cerebellar haemangioblastoma specially in von Hippel-Lindau disease. Histology shows a papillary epithelial tumor with hypervascular stroma, without atypia. The treatment for these tumors is surgical and curative when early diagnosed. In apparently sporadic cases, genetic analysis for Von Hippel-Lindau disease should be considered.

Keywords: Endolymphatic, sac, tumor, internal ear

1. INTRODUCTION

Endolymphatic sac is an established source of low-grade neoplasms, posing a difficult problem in local tumor control. These tumors may occur sporadically or in relation to von Hippel-Lindau disease [1]. Endolymphatic sac tumor is grows slowly. In addition, because the natural course and clinical behavior of this neoplasm are not yet established, the sporadic reports of such cases continue to provide basis for better understanding. The papillary tumors of the middle ear behave more aggressively with extension to the petrous apex and cranial cavity and frequently involve the facial nerve and the otic capsule [2].

In 1989, Heffner proposed that papillary tumors of the middle ear and temporal bone arise from the endolymphatic sac [3]. These tumors share a common origin from the mucosa of the endolymphatic sac and have clinical, radiologic, and histopathologic features that distinguish them from other tumors occurring in the petrous temporal bone and cerebellopontine angle. These tumors warrant distinction as a distinct clinicopathologic entity.

2. CASE REPORT

This is a 61-year-old patient without antecedent who presented otorrhea with hyperacusis and vertigo. The clinical examination found a patient in good general condition with left purulent otorrhea, decreased left visual acuity and left auditory acuity, without any neurological deficit. A CT scan of the rocks revealed a left otitis with bone lysis of the mastoid, the rock, the occipital bone with filling of the mastoid cells, the external auditory canal and the left lateral sinus (Figure 1).

A transmastoid biopsy was performed in favor of a papillary adenocarcinoma of the endolymphatic sac of the inner ear and the rock (AE1 / AE3 and Vimentin positive, PS 100 negative).

The magnetic resonance imaging of the facial mass showed a heterogeneous lesional process enhanced after injection of gadolinium 70x32x48 mm and extends from the posterior cerebral fossa lysing the clivus with infiltration of the tent of the cerebellum without extension of the tentorium with bilateral jugulo-carotid lymph nodes (Figure 2; Figure 3).

Figure 1: CT scan of the rocks: axial section

Figure 2: Magnetic resonance imaging coronal view
The excision of the tumor was impossible considering the extension of the tumor. The patient received 3D conformational radiotherapy at a dose of 60 Gy in 30 sessions at a rate of 2 Gy per fraction on the tumor and 50 Gy in 25 sessions at a rate of 2 Gy per fraction on the cervical lymph nodes II and III and supra clavicular. Tolerance of radiotherapy was marked by photophobia and lacrimation. The radiological evaluation at 3, 6 and 9 months with magnetic resonance imaging showed a stabilization of the process.

3. DISCUSSION

Several recent reports have suggested that locally aggressive adenomatous tumors with papillary histological architecture occurring in the posteromedial petrous temporal bone may be distinguished from other adenomas and adenocarcinomas of the middle ear and temporal bone. The pathological findings confirm that most of these tumors originate from the pars rugosa of the sac [4,5]. The tumor extension may show distinct patterns of growth. Laterally, they spread to the middle and external ear via a transmastoid route. Medially, the tumor extends to the cerebellopontine angle and the jugular foramen after Dural transgression. In large tumors, both patterns may coexist [5].

The clinical manifestations depend on tumor extension. The otological complaints in the form of unilateral, sudden onset hearing loss remains the commonest otological presentation [5]. In cases of posterior fossa extension, cerebellopontine angle syndrome, lower cranial nerve palsies, and obstructive hydrocephalus have been reported [2]. However, our patient presented with a history of ear discharge and decreased hearing, misdiagnosed as chronic suppurative otitis media.

Imaging features in endolymphatic tumors may be nonspecific [6] or highly characteristic [7]. These tumors appear to be centered between the sigmoid sinus and the internal auditory canal along the posterior petrosal plate, in the region of the vestibular aqueduct [8].

These tumors should be included in the differential diagnosis of destructive or nondestructive tumors about the posterior-medial face of the petrous bone as well as extra-axial cerebellopontine tumors. The CT scan may show prominent intratumoral calcific speculation or rim calcification [7]. The mastoid air cells remain pneumatized, distinguishing the lesion from neoplasms arising from the middle ear [8]. The MRI scan shows heterogenous foci of low and high signal intensity on both T1- and T2-weighted images, probably because of repeated intraparenchymal hemorrhages, leading to the deposition of hemosiderin, methemoglobin, and cholesterol crystals. The other tumors at this location, with similar radiological features, may be glomus tympanicum, atypical meningioma, metastasis, primary bone tumor, or chondrosarcoma [3]. Angiography identifies blood
supply of tumors especially for tumors more than 3 cm, where external carotid, internal carotid, and posterior circulation vessels are involved [7]. The clinicopathologic evidence of a large destructive papillary tumor of the posteromedial petrous temporal bone and a concomitant contralateral papillary tumor confined to the endolymphatic sac further supports the hypothesis that these tumors have an endolymphatic sac origin.

Macrosopically, the tumor is described as a friable, highly vascular, reddish polypoidal mass. However, the texture of the tumor may be much denser, fibrous, firm, and lobulated. The papillary architecture, with cuboidal and low columnar cells forming the epithelial lining, and subepithelial vascularity are typical of papillary cystic adenocarcinomas. In spite of benign histological features (with no mitotic activity and slight cellular polymorphism), these tumors are classified as adenocarcinomas because of their clinical course [5]. Immunohistochemical expression of neuron-related antigens, such as glial fibrillary acidic proteins, is explained by the neuroectodermal origin of the endolymphatic sac. This pattern may not always exist.

Many investigators [9,10,11,12] have reported the association between aggressive papillary tumors of the petrous temporal bone of endolymphatic sac origin and von Hippel Lindau disease, an autosomal dominant multisystem neoplastic disorder with angiomatosis of the retina and cerebellum and similar lesions present in a variety of organs. These patients may demand early radiologic assessment for endolymphatic sac tumor when symptoms referable to inner ear disease manifest.

Early gross total microsurgical resection is the treatment of choice, as definitive radiotherapy has yielded disappointing results. In a large series, 90% cure rate has been documented without radiation therapy. Hence, complete tumor extirpation seems to be unachievable in cases of tumors more than 3 cm [13,14]. The preservation of hearing can be obtained in early stages and must be tried in the context of a VHL disease where tumors are more frequently bilateral [15]. Total surgical excision is usually done by a transtemporal approach with postoperative radiation therapy as an adjunct when only subtotal excision is possible [16,17]. The selection of radiotherapy should be standard external irradiation or gamma knife radiosurgery [13]. Both pre- and post-operative external irradiation (50–60 Gy) was given to several patients in some series [18,19,20]. Heffner reported 3 of 4 patients developing tumor recurrence after subtotal resection with adjuvant irradiation. The analysis of the published series showed that 50% of the patients who received postoperative radiotherapy following subtotal resection of ELST had further growth of the residual tumor on follow up, and 20% of those who received radiation following a complete removal of the disease had recurrence within one year [21]. Consequently, the role of postoperative radiotherapy (fractionated or stereotactic) in the treatment of ELST is still controversial [21].

4. CONCLUSION

These tumors are a specific clinical entity and should be included in the differential diagnosis of petrous temporal bone and cerebellopontine angle tumors and in patients with symptoms of auditory and vestibular pathology. The locally aggressive nature of these “benign” neoplasms belies their bland histopathologic appearance, and the pathogenesis of their destructive capacity is deserving of further research.

COMPETING INTERESTS

The authors have declared that no competing interest exists.

FUNDING

This Case Report was funded by the Department of Radiotherapy, National Institute of Oncology of Rabat.

ETHICAL APPROVAL

Ethics Committee of the National institute of Oncology, Mohammed V University, Rabat, Morocco

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

ACKNOWLEDGEMENTS

We thank Radiotherapy department and our radiotherapist’s colleagues at National institute of Oncology of Rabat who provided care and support for this patient.

REFERENCES