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ELSEVIER

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Papillary neoplasms of the endolymphatic sac and mucosal lining of the pneumatic spaces of the temporal bone: Role of imaging

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Primary adenomatous and adenocarcinomatous lesions of the temporal bone are rare tumors. Primary low-grade papillary adenocarcinomas of the endolymphatic sac origin were first reported by Heffner. These lesions, which are referred to as "Heffner tumors," are slow-growing, bone-destructive infiltrating tumors. Owing to the local bone destruction (usually centered at posterior petromastoid plate), the general consensus favors the endolymphatic sac as being the origin of these tumors. However, controversies regarding the cellular origins of adenomatous tumors of the temporal bone have been reported. Mafee and Shah reported a unique case in which histology was identical to papillary adenocarcinoma of endolymphatic sac in which the sac and duct showed normal findings on magnetic resonance images, which were confirmed on surgery. Computed tomography and magnetic resonance imaging are excellent imaging methods for evaluation of normal and anomaly of the vestibular aqueduct and endolymphatic duct and sac and for the diagnosis of these endolymphatic tumors. This article reviews computed tomography and magnetic resonance imaging characteristics of adenocarcinomatous tumors of temporal bone. This report also concerns a unique case of a papillary adenocarcinoma of presumed endolymphatic sac in a 12-year-old child, in which tumor replaced almost the entire petromastoid resulting in a markedly egg-shell appearance of the bone. To the best of our knowledge, this is the youngest child with papillary endolymphatic sac tumor reported.

Introduction

Primary adenoma and adenocarcinomatous lesions of the temporal bone are distinctly unusual rare tumors[1-5]. In 1989, Heffner[1] described 20 cases of a type of papillary-cystic adenocarcinoma in which the neoplasm destroyed a large portion of the posterior petromastoid portion of the temporal bone and included a prominent extension into the posterior cranial fossa.[1] Patients histories indicated slow growth rate of the lesions. There were resemblances of the normal distal endolymphatic sac tissue to some portions of many of the neoplasm.[1] Tissue submitted to the United States Armed Forces Institute of Pathology (AFIP) from a rare case of papillary adenomatous neoplasm of endolymphatic sac origin, reported by Hassard et al[6] had a strong similarity to the larger, destructive neoplasms reviewed by Heffner. In his final analysis, Heffner concluded that the neoplasms in his series grew slowly, but they manifested a destructive, infiltrating growth into the temporal bone. Heffner preferred to diagnose them as low-grade adenocarcinomas. Because the neoplasms were epithelial and manifested, a destructive growth along the posterior face of the temporal bone, Heffner inferred that the endolymphatic sac seems an ideal location to give rise to these tumors with their combined intrabony and posterior fossa components. These tumors are referred to endolymphatic sac tumors (Heffner's tumor).[2] Since Heffner's original report, many authors have reported

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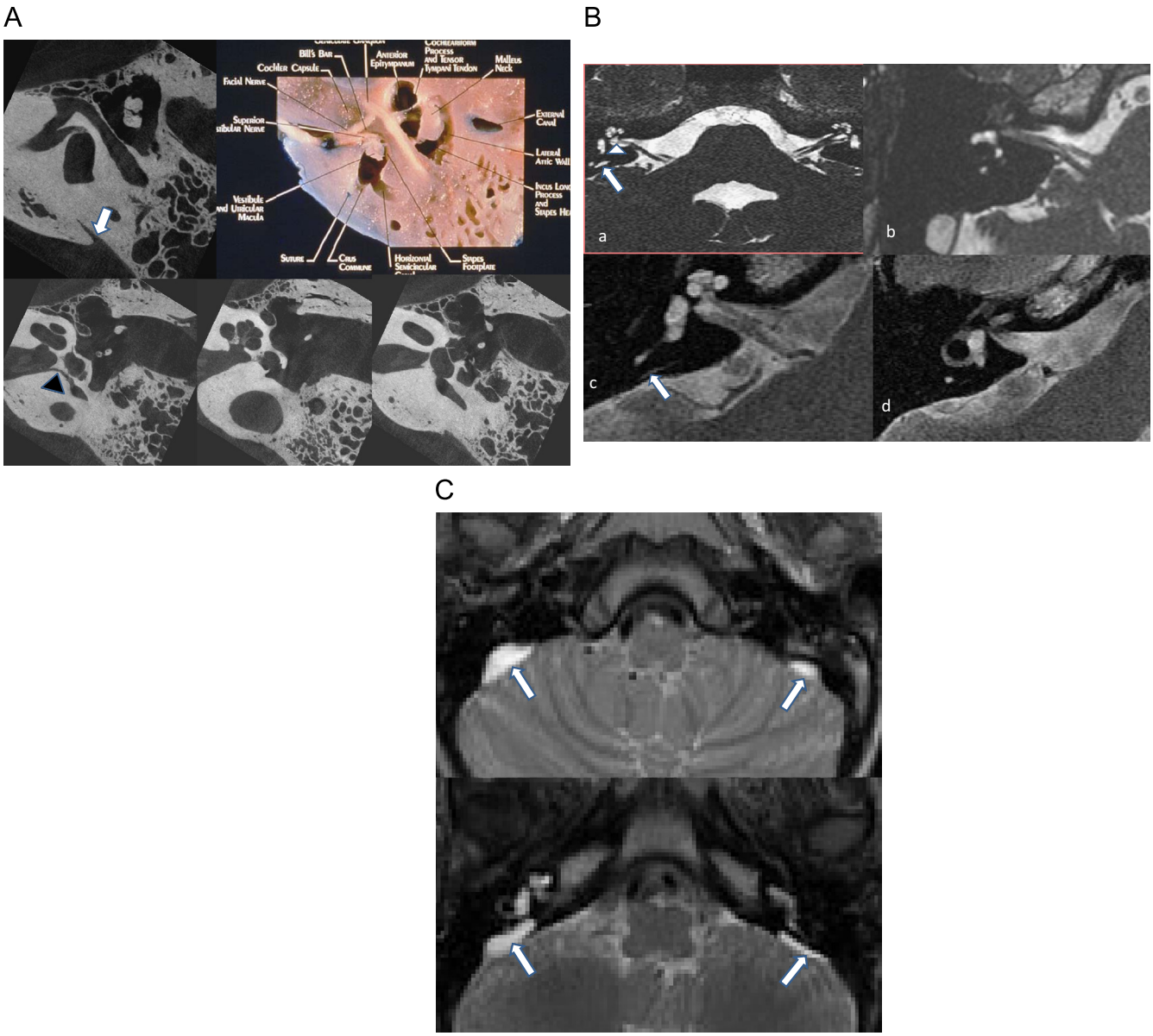

Figure 1 (A) Normal CT anatomy of a dehydrated temporal bone specimen, performed on a micro-CT unit at 18-μm in-plane resolution. Serial axial sections of a dried temporal bone, showing the labyrinthine segment, geniculate fossa, and tympanic segment of the fallopian canal. Note Bill's bar at the fundus of IAC, vestibular aqueduct (white arrow), and singular canal (arrowhead). Note the spiral laminae and tiny spiral foramina at the modiolus. Anatomical section, courtesy of Richard Buckingham, MD. (B) Normal MRI anatomy of the temporal bone. Axial T2W MR scans demonstrating cochlear and inferior vestibular nerves (a and c), facial and superior vestibular nerve (b and d), singular nerve (white arrowhead in a), and endolymphatic duct and sac (arrow in a and c). (C) Bilateral enlarged endolymphatic ducts and sacs. Axial T2W MR scans demonstrating markedly enlarged endolymphatic ducts and sacs (arrows). (Color version of figure is available online.)

these endolymphatic sac tumors (ELST).[4,5,7-11] These low-grade papillary adenocarcinomas characteristically cause destruction of the vestibular aqueduct (Figure 1) and at times marked erosion of the petromastoid plate along the posterior aspect of the petrous bone (Figure 2). There may be fine or coarse calcifications present within the tumor.[4,5,7-11] The neoplasm may be solid or a combination of solid and cystic components. There may be moderate to marked contrast enhancement on computed tomography (CT) and magnetic resonance imaging (MRI) (Figure 2). There may be extension of the tumor into the jugular fossa and middle ear cavity.[5,7-11] Some of the neoplasms with similar histopathologic features may arise from sublabyrinthine mastoid air cells submucosal glands with intact endolymphatic sac and vestibular aqueduct. Mafee and Shah[5] reported a papillary adenocarcinoma of the temporal bone, identical to what had been described as ELST, in which surgical evaluation revealed no pathogenic relation to the endolymphatic sac. The diagnosis of ELST is therefore a challenging one, with the cellular origins of these tumors of the temporal bone being endolymphatic sac or at times the mucosal lining of the pneumatic spaces of the temporal bone. Pollak et al[12] have