Computed tomography and magnetic resonance imaging features of a rare case of a primary epidermoid tumor of the jugular foramen

Parag Suresh Mahajan, Anuradha Parag Mahajan, Nawal M. Al Moosawi

Abstract

We present computed tomography (CT) and magnetic resonance imaging (MRI) features of a very rare case of a primary epidermoid tumor of the jugular foramen (JF). A 45-year-old male patient presented with gradually progressive vertigo and tinnitus. CT and MRI scans revealed a 3.5 cm right-sided JF tumor with characteristic bright signal (restricted diffusion) on diffusion-weighted MRI (DWI). DWI may be useful in accurately differentiating the lesion from other cystic neoplasms of the JF. We describe the imaging features of intracranial epidermoid and JF tumors and discuss its differential diagnosis.

Key words: Computed tomography, congenital cholesteatoma, jugular foramen, magnetic resonance, primary epidermoid

INTRODUCTION

Epidermoid tumors (ETs) are extra-axial lesions observed anywhere along the neuraxis and constitute up to 1.8% of all cerebral neoplasms.[1,2] Skull base is the most common location of ET, mainly involving the cerebellopontine angle and parasellar regions.[3] We describe the clinical imaging characteristics of a unique ET, primarily arising from and exclusively located in the jugular foramen (JF) and having the typical bright appearance on diffusion-weighted magnetic resonance imaging (DWI). Primary ET of the JF is also known as a congenital cholesteatoma. Although JF tumors may lead to lower cranial neuropathies, in some cases they present with few signs and symptoms due to compensation by the cranial nerves on the contralateral side.[4] JF tumors may demonstrate significant vascularity and involvement of crucial neurovascular structures. Diagnosis and management of the JF tumors can be difficult and challenging.[5]

CASE REPORT

A 45-year-old male patient was seen in the ear, nose and throat surgery outpatient department of our hospital with the complaints of gradually progressive tinnitus and vertigo since 6 months. His previous clinical history was noncontributory and had no signs or symptoms indicative of lower cranial neuropathy. Contrast-computed tomography (CT) scan of the temporal bones was performed, which revealed a 3.5 cm × 2.2 cm nonenhancing cystic density lesion involving the right JF resulting in expansion and thinning of JFs roof. No intrasional calcifications or erosions of the adjacent bone were noted [Figure 1a and b]. The differential diagnosis of primary arachnoid or epidermoid cyst of the JF was considered. Further evaluation was performed with magnetic resonance imaging (MRI), which indicated an oval lobulated nonenhancing lesion causing expansion of...
Case Reports

Paragangliomas constitute the commonest JF tumors. Differential diagnosis of JF tumors includes schwannomas, meningiomas, arachnoid cysts, chordomas, chondrosarcomas, dermoids, epidermoids, and metastatic lesions. Presenting symptoms of patients with JF tumors include pulsatile tinnitus, reduced hearing, aural region fullness, dysphagia, and hoarseness. In our case, the patient had progressive vertigo and dizziness, which can be better explained by compressive neuropathy of the vestibulocochlear nerve. Paragangliomas arise in paraganglia that are derivatives of the embryonic neural crest. Both glomus jugulare and glomus tympanicum paragangliomas originate in the temporal bone, the former originate from paraganglia associated with cranial nerves IX, X, and XI in the JF and the latter originate from paraganglia on the middle ear promontory. Schwannomas and meningiomas are the second and third commonest JF tumors, respectively. Schwannomas originate from the nerve sheath Schwann cells and meningiomas from the jugular bulb arachnoid cells. Interestingly metastatic lesions of the JF are rare.

Other lesions of the JF include pseudomass, which is a common misdiagnosis. Pseudomass includes normal vascular asymmetry, a high jugular bulb, a jugular diverticulum, or jugular bulb flow variants (wherein slow venous flow may be mistaken for contrast-enhancement). However, in these cases the bone-windowed CT images will show a normal JF bone structure, flow-sensitive MRI would confirm the vascular anatomic variant, and T2-weighted MRI will usually show a hyper-intense JF. ET also originates from ectodermal inclusions during the closure of the neural tube. This process likely occurs between the 3rd and the 5th weeks of embryonic life when the optic and the otic vesicles are developing. This explains the common occurrence of ETs in the parasellar and the cerebellopontine angle regions.

On histopathological examination, the ET demonstrate a wall made of stratified squamous epithelium supported by collagen from the outside. Repeated desquamation inside the lumen of the ET gives rise to lamellations. The internal contents are mostly soft, white and cheesy and are made up of keratinous debris with abundance of cholesterol crystals.

DISCUSSION

Paragangliomas constitute the commonest JF tumors. Differential diagnosis of JF tumors includes schwannomas, meningiomas, arachnoid cysts, chordomas, chondrosarcomas, dermoids, epidermoids, and metastatic lesions. Presenting symptoms of patients with JF tumors include pulsatile tinnitus, reduced hearing, aural region fullness, dysphagia, and hoarseness. In our case, the patient had progressive vertigo and dizziness, which can be better explained by compressive neuropathy of the vestibulocochlear nerve. Paragangliomas arise in paraganglia that are derivatives of the embryonic neural crest. Both glomus jugulare and glomus tympanicum paragangliomas originate in the temporal bone, the former originate from paraganglia associated with cranial nerves IX, X, and XI in the JF and the latter originate from paraganglia on the middle ear promontory. Schwannomas and meningiomas are the second and third commonest JF tumors, respectively. Schwannomas originate from the nerve sheath Schwann cells and meningiomas from the jugular bulb arachnoid cells. Interestingly metastatic lesions of the JF are rare.

Other lesions of the JF include pseudomass, which is a common misdiagnosis. Pseudomass includes normal vascular asymmetry, a high jugular bulb, a jugular diverticulum, or jugular bulb flow variants (wherein slow venous flow may be mistaken for contrast-enhancement). However, in these cases the bone-windowed CT images will show a normal JF bone structure, flow-sensitive MRI would confirm the vascular anatomic variant, and T2-weighted MRI will usually show a hyper-intense JF. ET also originates from ectodermal inclusions during the closure of the neural tube. This process likely occurs between the 3rd and the 5th weeks of embryonic life when the optic and the otic vesicles are developing. This explains the common occurrence of ETs in the parasellar and the cerebellopontine angle regions.

On histopathological examination, the ET demonstrate a wall made of stratified squamous epithelium supported by collagen from the outside. Repeated desquamation inside the lumen of the ET gives rise to lamellations. The internal contents are mostly soft, white and cheesy and are made up of keratinous debris with abundance of cholesterol crystals.

Figure 2: Axial T2-weighted images (a), axial fluid-attenuated inversion-recovery (FLAIR) image (b), coronal T2-weighted image (c), coronal precontrast T1-weighted image (d) and coronal postcontrast T1-weighted image (e) showing nonenhancing cystic intensity lesion in the right jugular foramen (arrows). Note heterogeneous signal intensity on FLAIR image (arrow).

Figure 3: Axial diffusion-weighted image shows hyperintense signal of the right jugular foramen lesion (arrow), suggesting restricted diffusion within the lesion.
In some cases, the internal contents may be thick, brown and viscid. Nevertheless, epidermoids are slow growing tumors.[3]

Characteristic CT features of ET consist of a cystic (like cerebrospinal fluid [CSF]) density rounded or a lobulated space occupying lesion, which may have characteristic serrated margins. Dense epidermoid is a rare variant of ET, which appears homogeneously hyper-dense on CT scans. This is thought to be due either to high protein content or saponification of debris within the cyst to soaps of calcium.[4] Calcification may be present in <10% of all intracranial epidermoids.[5] Calcification within the ETs may also be due to saponification of the desquamated cyst debris.[6]

Typical MRI features of ET include the hypointense appearance on T1-weighted images, hyperintense appearance on T2-weighted images and heterogeneous internal contents (in almost 65% of cases) in the FLAIR and proton-density (PD)-weighted images. Heterogeneous appearance on FLAIR and PD-weighted images helps in distinguishing ET from the closely mimicking arachnoid cysts. MRI may also show insinuation of the cystic lesion into the adjacent cisterns or fissures and serrated margins of the cyst, features that are not associated with arachnoid cysts. Typical ET are nonenhancing lesions. When contrast enhancement is present (35% of cases), it is minimal and peripheral.[7] In our case all characteristic imaging features of epidermoid were observed.

Diffusion-weighted magnetic resonance imaging is very useful in diagnosing an ET. ET appears significantly more hyperintense than CSF and cerebral white matter on T2-weighted images due to an amalgamation of T2 and diffusion effects. Apparent diffusion coefficient (ADC) of ET is more or less same as that of grey matter and less than that of CSF.[8,9] Arachnoid cysts and other cystic lesions in the cranium, on the other hand, do not show restricted diffusion, and have signal on DWI and ADC maps similar to that of CSF.[8,9] In this case, the ET showed a very bright signal on DWI, indicative of restricted diffusion. Sometimes, this bright signal may be caused by T2 shine-through. T2 shine-through is defined as significant DWI hyperintensity that is not because of restricted diffusion, but due to increased T2 signal which “shines-through” to the DWI image. Extended T2 decay time in few of the normal tissues leads to T2 shine-through. This is commonly seen in intracranial sub-acute infarcts having vasogenic edema but can also be seen in other pathologies such as epidermoid cysts. For confirmation of real restricted diffusion, comparison may be made between the DWI image and the ADC. In cases of real restricted diffusion, the area of high signal on DWI will appear as a low signal on ADC. ADC is a value that computes the effect of diffusion independent of the influence of T2 shine-through. ADC maps thus reveal restricted diffusion, such as in ischemic injury, as low signal lesions as compared to normal brain tissue. However, the T2 shine-through appears as normal or high signal on ADC. A previous study on ETs found very high DWI signal intensity and ADCs that are higher (and not lower) than that of the cerebral tissue, suggesting that the T2 properties (also called as T2 shine-through) dominated the contributions to the DWI signal intensity and even overwhelmed the effect of signal attenuation resulting from the increase in ADC.[10] They also mentioned that there were no examples of intracranial tumors with T2 shine-through phenomenon showing such intense contrast between the lesion and brain as the ETs.[10] This indicates that DWI has significant diagnostic value in imaging of intracranial ETs and correlation with ADC maps may not be useful. Our case demonstrated this characteristic DWI appearance.

To the best of our knowledge, this is the first report of primary ET of JF showing the characteristic DWI appearance. The other differential considerations of cystic ET of JF include arachnoid cyst and other cystic tumors, which have not been described to appear bright on DWI. When DWI is not available, the MRI features are not specific, and these JF lesions can be mistaken for arachnoid cysts or other cystic tumors.[10]

To conclude, we present CT and MRI features of a very rare case of a primary ET in an unusual location (JF), appearing characteristically bright on DWI, feature that helped differentiate it from other JF mass lesions.

REFERENCES

1. Johnston F, Crockard HA. Dermoid, epidermoid and neuroenteric cysts. In: Kaye A, Laws EJ, editors. Brain Tumors. 1st ed. New York, NY: Churchill Livingston; 1995. p. 895-905.
2. Aribandi M, Wilson NJ. CT and MR imaging features of intracerebral epidermoid – a rare lesion. Br J Radiol 2008;81:e97-9.
3. Robertson R, Caruso PA, Truwit CL, Barkovich AJ. Disorders of brain development. In: Atlas SW, editor. Magnetic Resonance Imaging of the Brain and Spine. 3rd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2002. p. 279-369.
4. Ramina R, Maniglia JJ, Fernandes YB, Paschoal JR, Pfeilsticker LN, Neto MC, et al. Jugular foramen tumors: Diagnosis and treatment. Neurosurg Focus 2004;17:E5.
5. Leonetti JP, Shirazi MA, Marzo S, Anderson D. Neuroendocrine carcinoma of the jugular foramen. Ear Nose Throat J 2008;87:88-91.
6. Timmer FA, Sluzewski M, Treskès M, van Rooij WJ, Teepen JL, Wijnaalda D. Chemical analysis of an epidermoid cyst with unusual CT and MR characteristics. AJNR Am J Neuroradiol 1998;19:1111-2.
7. Kallmes DF, Provenzale JM, Cloft HJ, McClendon RE. Typical and atypical MR imaging features of intracranial epidermoid tumors. AJR Am J Roentgenol 1997;169:883-7.
8. Tsuruda JS, Chew WM, Moseley ME, Norman D. Diffusion-weighted MR imaging of the brain: Value of differentiating between extraaxial cysts and epidermoid tumors. AJR Am J Roentgenol 1990;155:1059-65.
9. Schaefer PW, Grant PE, Gonzalez RG. Diffusion-weighted MR imaging of the brain. Radiology 2000;217:331-45.
10. Chen S, Ikawa F, Kurisu K, Arita K, Takaba J, Kanou Y. Quantitative MR evaluation of intracranial epidermoid tumors by fast fluid-attenuated inversion recovery imaging and echo-planar diffusion-weighted imaging. AJNR Am J Neuroradiol 2001;22:1089-96.
11. Watanabe K, Wakai S, Nagai M, Muramatsu H. Epidermoid tumor with unusual CT and MR findings – case report. Neurol Med Chir (Tokyo) 1990;30:977-9.

How to cite this article: Mahajan PS, Mahajan AP, Al Moosawi NM. Computed tomography and magnetic resonance imaging features of a rare case of a primary epidermoid tumor of the jugular foramen. J Nat Sc Biol Med 2015;6:236-9.

Source of Support: Nil. Conflict of Interest: None declared.