

Cystic Acoustic Schwannoma: A Case Report and Review of Literature

Agrawal A¹, Vagh S.J², Gosavi S.N³, Pachpande B⁴, Ubeja G⁵

Abstract

A cystic acoustic neurinoma almost entirely consists of a cystic component and is a well recognized subtype of acoustic schwannoma. We report a case of a 40-year-old male patient who presented with clinical features of a left cerebello-pontine angle mass lesion and CT scan findings mimicking the diagnosis of an epidermoid cyst. However intraoperative and histopathological findings were suggestive of a large cystic acoustic schwannoma. We have also analyzed the characteristics, clinical symptoms and radiological findings of these tumors. In the presence of a large cystic cerebellopontine angle tumor without a significant hearing loss and no destruction of the internal auditory canal, the possibility of the cystic acoustic neurinoma should be considered.

Keywords: vestibular schwannoma, acoustic neurinoma, cerebellopontine angle tumor, cystic schwannoma

¹. Associate Professor in Neurosurgery, Department of Surgery, Datta Meghe Institute of Medical Sciences, Sawangi (Meghe), Wardha (India)

². Professor and Head, Department of Pathology, Datta Meghe Institute of Medical Sciences, Sawangi (Meghe), Wardha (India)

³. Assistant Professor in Anesthesiology, Department of Anesthesiology, Datta Meghe Institute of Medical Sciences, Sawangi (Meghe), Wardha (India)

⁴. Resident in Anesthesiology, Department of Anesthesiology, Datta Meghe Institute of Medical Sciences, Sawangi (Meghe), Wardha (India)

⁵. Resident in Surgery, Department of Surgery, Datta Meghe Institute of Medical Sciences, Sawangi (Meghe), Wardha (India)

Corresponding author:

Amit Agrawal

Phone: +91-9102-240129, 243073

Fax: +91-9102-2231902

Email: dramitagrawal@gmail.com

IJCP 2008; 4: 187-190

Introduction

A cystic acoustic neurinoma almost entirely consists of a cystic component and is a well recognized subtype of acoustic schwannoma [1-11]. These lesions can be misdiagnosed preoperatively because their computed tomography appearances can resemble many other cystic lesions in the cerebellopontine angle. [2] Based on the available literature and our case, we have analyzed various clinical symptoms and CT scan findings of these tumors.

Case Report

A 40-year-old male presented with headache, dizziness, difficulty in walking, weakness of left

upper and lower limbs, deviation of angle of mouth to the right side and dysarthria from three months ago. The patient did complain of a hearing loss, tinnitus, or vertigo. Higher mental functions were normal. On cranial nerve examination he showed gaze evoked nystagmus and mild left facial nerve paresis. An audiogram showed a 70-dB sensorineural hearing disturbance at 8000 Hz on the left side. Plain CT scan of brain showed a well defined, low-density cystic lesion, mildly hyperdense to cerebrospinal fluid, along with displacement of the fourth ventricle and distortion of the brain stem. There were no bony changes of the internal auditory canal or enhancement after contrast administration

Table 1: Comparative clinical and CT scan characteristics of typical acoustic schwannoma and large cystic schwannoma

Features	Typical acoustic neuroma	Cystic acoustic neuroma
Clinical features	Evidence of 5th and 7th cranial nerve and pontine-cerebellar disturbances	Similar
	A history of progressive, unilateral sensorineural hearing loss, usually encompassing many months or years ^{10,17}	A short clinical history, an atypical initial symptom, facial nerve involvement ¹
CT scan ¹⁻¹¹	Usually almost isodense with the adjacent cerebellar tissue After infusion of a contrast agent, almost all acoustic neurinomas exhibit enhancement Two thirds of these tumors enhance homogeneously and have well defined margins, whereas one third exhibit ring enhancement	Usually appear as a well demarcated, low-density area on precontrast CT scan Exhibit either no contrast enhancement or a thin, discontinuous, faint ring of enhancement in the cyst wall Occasionally a tiny solid portion can be identified ⁵ Occasionally horizontal fluid levels on plain CT scan ¹¹
	Widening of the cistern around the tumor An acute angle between the lateral tumor border and the petrous bone ^{10,18}	These features were lacking or subtle ^{1-2,10,19}
	Enlargement and/or erosion of the IAC (One of the most important radiological diagnostic features of acoustic neurinomas) ^{10,17}	There may be no abnormality of the IAC and/or bone erosion ¹⁻²

(Figure 1 A-D). The patient underwent a left retromastoid suboccipital craniectomy. There was a thin walled cystic lesion with very fragile capsule containing yellowish serous fluid in the left CPA. The cystic portion was easily dissected from the cerebellum and excised; however, fragility of the capsule made it extremely difficult to dissect it free from the surrounding structures particularly from the 7th and 8th cranial nerves and brain stem and part of the capsule was left behind (Figure 2). The postoperative course was uneventful, with preserved 7th and 8th cranial nerve functions. Histological examination of the surgical specimen revealed that the cyst walls were composed of typical Schwann cells with a predominant Antoni type-A pattern and confirmed the diagnosis of acoustic schwannoma (Figure 3). The patient is doing well at one year follow up.

Discussion

The pathogenesis of the unusual cystic alteration of cystic acoustic neurinomas may be similar to that of the small cysts that often accompany solid tumors (necrosis, degeneration, and hemorrhage within the tumor). [12, 13] The only difference may be in the degree of such changes. [2] Computed tomography (CT) and magnetic resonance imaging (MRI) have considerably simplified the diagnosis of acoustic

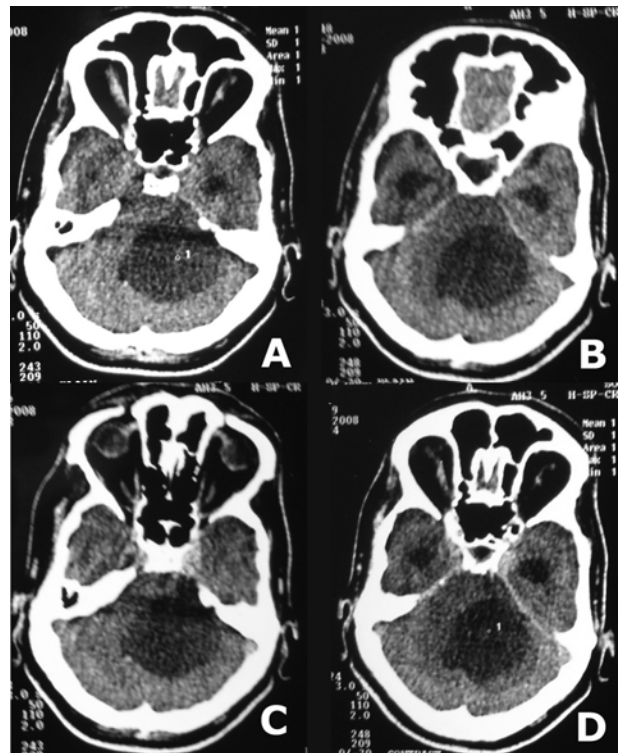


Figure 1. Plain CT scan (A & B) showing a large, well defined, low-density cystic mass with high density fluid levels in the left CPA region, Post contrast CT scan (C & D) did not show any enhancement in the lesion

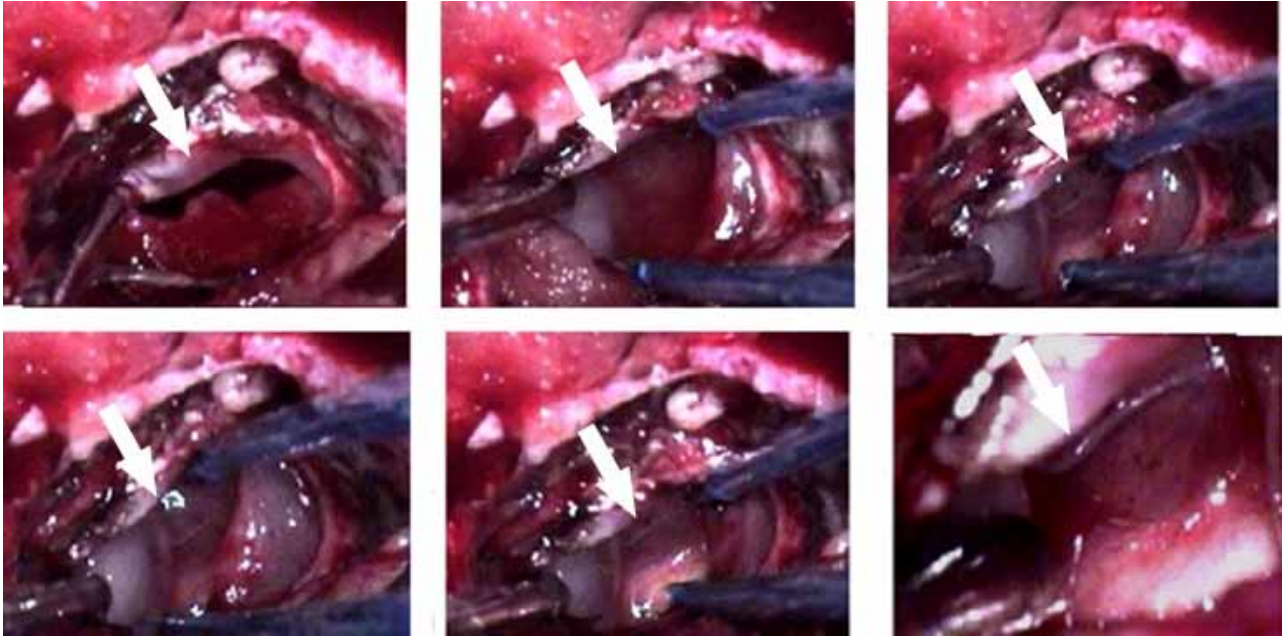


Figure 2. Intra-operative photograph showing the thin wall of the cyst contained clear and was densely adherent to the surrounding structures

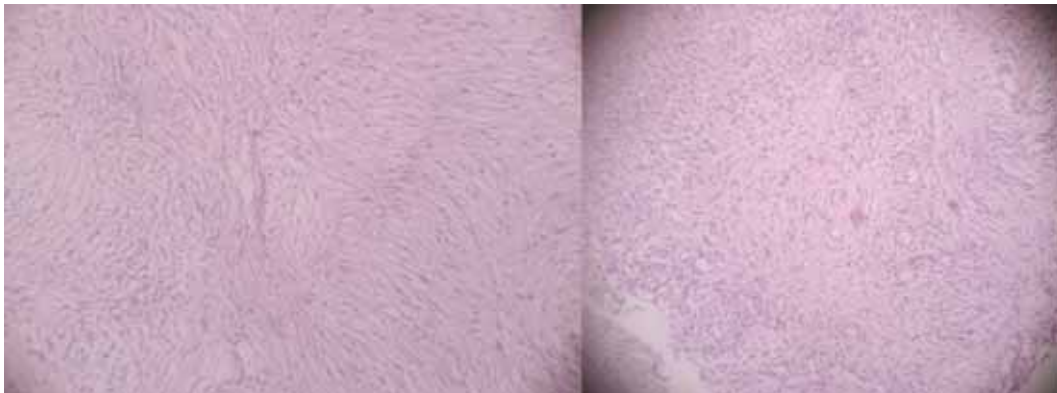


Figure 3. Photomicrograph showing that the cyst wall is composed of typical Schwann cells with a predominant Antoni type-A pattern (Haematoxylin and Eosin $\times 40$)

neuromas. [14] The differential diagnosis of cystic acoustic neurinomas can include epidermoid cyst, arachnoid cyst, neurenteric cyst, dermoid cyst and lipoma. [15] On computed tomographic scans, cystic acoustic neurinomas may mimic these cystic lesions; however, based on the clinical features and imaging findings, a diagnosis of cystic acoustic schwannoma can be suspected (Table 1). A gadolinium-enhanced MRI scan is more sensitive and superior to CT scan to diagnose and differentiate a cystic acoustic neurinoma from other cystic lesions of the CP angle [16, 17]. The majority of the cystic acoustic neurinomas are histologically composed of typical Schwann cells without any special features.

[18, 19] Occasionally, the cyst wall may contain abnormal sinusoid and telangiectasia-like vessels, endothelial proliferations with myxoid degeneration and necrosis. [20] It is largely unknown why large cystic acoustic neurinomas often produce atypical clinical symptoms and do not cause widening of the IAC. It has been postulated that these tumors, unlike solid tumors, tend to originate not within the IAC, but directly from the junction between the neurilemmal sheath and the neuroglial fibers located in the cerebellopontine cistern. Another possibility is that these tumors, being cystic, may be too soft to destroy bone or disrupt nerve fibers. [21] Cystic tumors differ from solid acoustic neuromas by having a rapid

growth pattern, short clinical history and more frequent facial nerve involvement. [٦] The role of matrix metalloproteinase (MMP) expression in the pathogenesis of tumor associated cyst formation in brain tumors as well as in acoustic schwannoma has been investigated and it has been found that MMPs may be partly involved in the pathogenesis of tumor associated cysts and peritumoral edema. [٨,٩] It is believed that matrix metalloproteinase ٢ (MMP-٢) may aggravate adhesion to the facial nerve, either by promoting the enlargement of the tumor or engendering the degradation of the tumor-nerve barrier proteolytically, making the dissection of the tumor and facial nerve preservation difficult. [٨] In the presence of a large cystic cerebellopontine angle tumor especially if there is no significant hearing loss and no destruction of the internal auditory canal, the possibility of the cystic acoustic neurinoma should be considered. [١٠-١٢] Because of these characteristics, it has been found that the cystic component in vestibular schwannoma is associated with a less favorable surgical outcome, probably due to the rapid tumor growth and associated compression of the posterior fossa structures. [٦, ٩, ١٠, ١١] It has also been found that a cystic acoustic schwannoma is less responsive to gamma knife surgery than a solid counterpart. [١٢]

References

- Miyagi A, Maeda K, Sugawara T. [Large cystic neurinoma: a case report][Article in Japanese] *No Shinkei Geka*. ١٩٩٧;٢٥(٧):٦٤٧-٥٤.
- Lin CL, Hwang SL, Howng SL. Unusual large cystic acoustic neurinomas: report of two cases. *Kaohsiung J Med Sci*. ١٩٩٧;١٣(٣):١٩٠-٤.
- Kameyama S, Tanaka R, Kawaguchi T, et al. Cystic acoustic neurinomas: studies of ١٤ cases. *Acta Neurochir*. ١٩٩٦;١٣٨(٦):٦٩٥-٩.
- Lunardi P, Missori P, Mastronardi L. Cystic acoustic schwannomas. *Acta Neurochir*. ١٩٩١;١١٠(٣-٤):١٢٠-٣.
- Yamada S, Aiba T. Cystic acoustic neurinomas--report of two cases. *Neurol Med Chir*. ١٩٨٩;٢٩(٩):٨٣٠-٣.
- Sinha S, Sharma BS. Cystic acoustic neuromas: surgical outcome in a series of ٥٨ patients. *J Clin Neurosci*. ٢٠٠٨;١٥(٥):٥١١-٥.
- delpolyi AR, Yang I, Buckley A, et al. Fluctuating response of a cystic vestibular schwannoma to radiosurgery: case report. *Neurosurgery*. ٢٠٠٨;٦٢(٥): ١١٦٤-٥.
- Moon KS, Jung S, Seo SK, et al. Cystic vestibular schwannomas: a possible role of matrix metalloproteinase-٢ in cyst development and unfavorable surgical outcome. *J Neurosurg*. ٢٠٠٧;١٠٦(٥):٨٦٦-٧١.
- Wandong S, Meng L, Xingang L, et al. Cystic acoustic neuroma. *J Clin Neurosci*. ٢٠٠٥;١٢(٣):٢٥٣-٥.
- Fundová P, Charabi S, Tos M, et al. Cystic vestibular schwannoma: surgical outcome. *J Laryngol Otol*. ٢٠٠٠;١١٤(١٢):٩٣٥-٩.
- Su WD, Li XG, Liu R, et al. Diagnosis and treatment of cystic acoustic neuroma: Article in Chinese. *Zhonghua Wai Ke Za Zhi*. ٢٠٠٣;٤١(٣):٢٠٥-٧.
- Dubois P. Tumors of the cerebellopontine angle. In: *Radiology*, in Wilkins RH, Rengachary SS (eds): *Neurosurgery*, vol ١. New York. McGraw-Hill. ١٩٨٥: ٧٠٤-٧١٩.
- Krassanakis K, Sourtsis E, Karvounis P. Unusual appearance of an acoustic neuroma on computed tomography. *Neuroradiol*. ١٩٨١;٣١: ٥١-٥٣.
- Curtin HD, Hirsch WL. Imaging of acoustic neuromas. *Neurosurg Clin N Am*. ٢٠٠٨;١٩(٢): ١٧٥-٢٠٥.
- Harner SG, Laws ER. Clinical findings in patients with acoustic neurinoma. *Mayo Clin Proc*. ١٩٨٣;٥٨: ٧٢١-٨.
- Valvassori GE. Radiologic evaluation of eighth nerve tumors. *Am J Otolaryngol*. ١٩٨٤;٥: ٢٧٠-٨٠.
- Wu EH, Tang YS, Zhang YT, et al. CT in diagnosis of neurinomas. *AJNR*. ١٩٨٦;٧: ٦٤٥-٦٥٠.
- Pinto RS, Kricheff II. Neuroradiology of intracranial neuromas. *Semin Roentgenol*. ١٩٨٤;١٩: ٤٤-٥٢.
- Hart RG, Gardner DP, Howieson tumors. Atypical features and recent diagnostic tests. *Neurology (NY)*. ١٩٨٣;٣٣: ٢١١-٢٢١.
- Bonneville F, Savatovsky J, Chiras J. Imaging of cerebellopontine angle lesions: an update. Part ٢: intra-axial lesions, skull base lesions that may invade the CPA region, and non-enhancing extra-axial lesions. *Eur Radiol*. ٢٠٠٧;١٧(١١):٢٩٠٨-٢٠.
- Jung S, Moon KS, Kim ST, et al. Increased expression of intracystic matrix metalloproteinases in brain tumors: relationship to the pathogenesis of brain tumor-associated cysts and peritumoral edema. *J Clin Neurosci*. ٢٠٠٧;١٤(١٢):١١٩٢-٨.