*The Korean Journal of Pathology 2010; 44: 543-6*
*DOI: 10.4132/KoreanJPathol.2010.44.5.543*

# Jugulotympanic Paraganglioma, Mimicking a Vascular Tumor
## – A Brief Case Report –

**Ji-Youn Sung · Chang Il Cha[1] Yong-Koo Park**

Departments of Pathology and [1]Otorhinolaryngology, Kyung Hee University Medical Center, Kyung Hee University College of Medicine, Seoul, Korea

**Received** : July 10, 2009
**Accepted** : September 30, 2009

**Corresponding Author**
Yong-Koo Park, M.D.
Department of Pathology, Kyung Hee University Medical Center, 1 Hoegi-dong, Dongdaemun-gu, Seoul 130-701, Korea
Tel: +82-2-958-8742
Fax: +82-2-958-8740
E-mail: ykpark@khmc.or.kr

Jugulotympanic paragangliomas (JTPs) known as glomus tumors, are neoplasms of variable invasiveness that arise from the paraganglia situated around the jugular bulb or middle ear. We now report a rare case of JTP in an 18-year-old male. Preoperative diagnoses through external auditory canal biopsy and radiologic examination both failed. Even using a frozen section, an informative finding was not obtained because mostly granulation tissue was present along with associated squeezing artifacts. On permanent histologic examination, small cell nests between many ectatic small vessels and fibrotic stroma were seen, and those cells were positive for CD56, synaptophysin and chromogranin. Because JTPs are rare and have rather different histologic findings – higher vascularity, smaller and less uniform tumor cells than other paragangliomas – they are easy to misdiagnose. However, remembering those differences may help the physician avoid missing JTPs.

**Key Words :** Paraganglioma, extra-adrenal; Glomus tympanicum; Glomus jugulare

Head and neck paragangliomas are relatively uncommon tumors, representing 0.6% of all neoplasms of the head and neck.[1] Among them, jugulotympanic paragangliomas (JTPs) arise from anatomically dispersed paraganglia near the base of the skull and middle ear. Actually, jugular paragangliomas are those arising from paraganglia situated in the vicinity of the jugular bulb, and tympanic paragangliomas are usually located along the course of Jacobson's nerve in the middle ear cavity. However, it may not always be possible to neatly distinguish between these two entities and the term JTPs is favored. Under microscopic examination, JTPs tend to be more vascular, and cell nests are less uniform and frequently smaller compared with other paragangliomas. Dense sclerotic matrix is another characteristic of JTPs. Vascular tumor, middle ear adenoma and even a reactive condition such as granulation tissue in chronic otomastoiditis are diagnostic pitfalls of this tumor. Therefore, when we interpret a tumor presenting as a middle ear or temporal bone mass, this pitfall should be kept in mind. Herein we report a case of jugulotympanic paraganglioma in an 18-year-old male.

## CASE REPORT

An 18-year-old male was admitted for evaluation of a mass in the right external auditory canal. He had complained of right sided otorrhea and a hearing disturbance for the last two months. On physical examination, the right external auditory canal was narrowed and was clogged by soft tissue with an invisible tympanic membrane. On temporal computed tomography, the 2.7 cm soft tissue mass was seen, and it was located in the right external auditory canal, middle ear cavity and mastoid process (Fig. 1A). The mass showed an irregular high signal intensity in a T2 weighted image and was isodense in a T1 weighted image from temporal magnetic resonance imaging. It was located in the medial portion of the right mastoid air cell and petrous apex of the temporal bone and was associated with bone destruction (Fig. 1B). The impression of the radiologist was that there was a malignant mass.

Two tissue biopsies through the external auditory canal were done. However, they failed to provide diagnostic information except that the tissue showed inflammation and necrosis. So, an

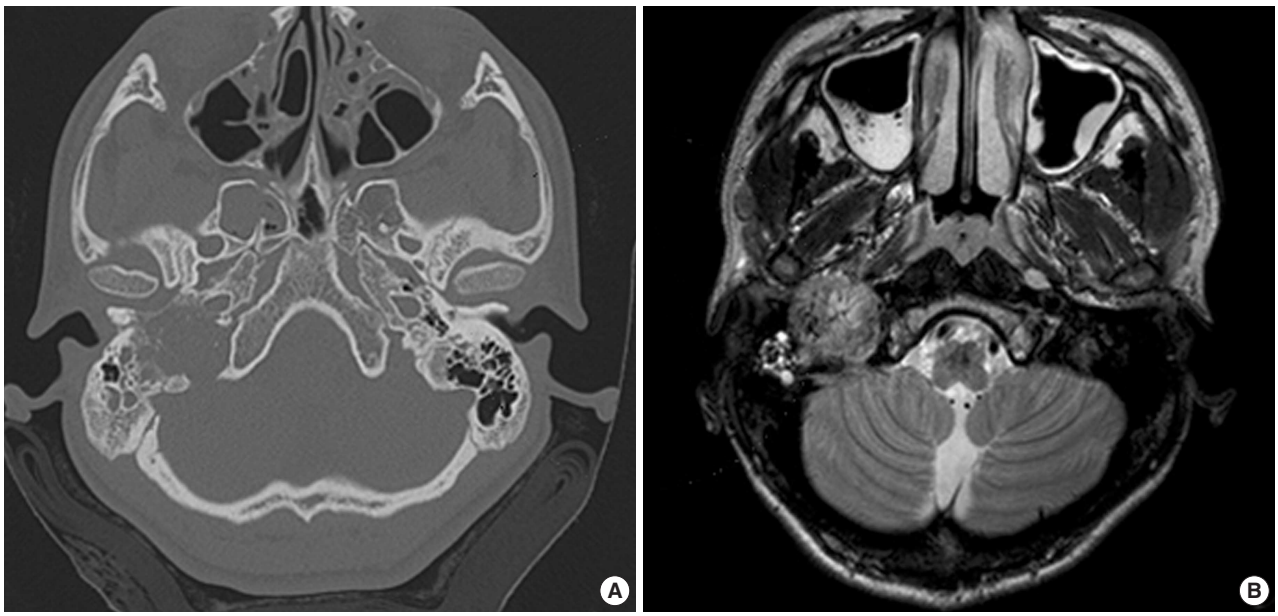


Fig. 1. (A) Computed tomography shows a destructive lesion of the right temporal mastoid process and the middle ear cavity having communication with the right external auditory canal. (B) T2-weighted magnetic resonance image shows a mildly high signal intensity middle ear mass that extends to the right mastoid air-cell and petrous apex area with encasement of the right internal carotid artery.

intraoperative biopsy was done under general anesthesia. After a retroauricular incision and open cavity tympanomastoidectomy, a polypoid mass with a bleeding tendency was found, one that filled the external auditory canal, middle ear cavity and mastoid process. Whereas the tympanic membrane was not found, and auditory ossicles were disrupted, the sigmoid sinus, dura matter and facial canal were intact. Most of the mass was removed, but some remained because of massive bleeding.

On histologic examination, highly vascular tissue looking like granulation tissue was seen in a low power microscope field. However, in a high power field, we observed cell nests composed of less uniform small cells between the increased ectatic small vessels (Fig. 2A, B). The small cells had relatively hyperchromatic nuclei and showed immunoreactivity for CD56 (Fig. 2C), synaptophysin, chromogranin and neuron specific enolase. Spindle shaped sustentacular cells surrounding cell nests were positive for S100 (Fig. 2D). These cells were negative for cytokeratin and CD34. Moderate stromal sclerosis was also noted.

Although considerable squeezing artifacts were present, this case was diagnosed as a jugulotympanic paraganglioma on the basis of the histologic and immunohistochemical findings.

## DISCUSSION

JTPs can be confused with glomus tumors, which are tumors of perivascular cells. The reason is that many clinicians call JTPs "glomus" tumors although the term is not a pathologic diagnosis. These two entities–glomus tumors and JTPs that are called "glomus" tumors, are definitely different diseases. JTPs are tumors arising from paraganglia whereas glomus tumors arise from a modified smooth muscle cell located in the walls of specialized arteriovenous anastomoses involved in temperature regulation. Hence, it is better to use the term JTPs rather than glomus tumors to avoid confusion.

The major differential diagnosis of JTPs includes ruling out a vascular tumor such as an epithelioid hemangioendothelioma (EHE) because of the high vascularity of JTPs. EHE can be differentiated from JTPs by the findings of tumor cells having a moderate amount of pale eosinophilic cytoplasm and immunoreactivity for vascular markers such as CD31, CD34 and factor VIII.[2] We also thought this case was a vascular tumor, because there were numerous small-sized vessels and the small cells between the vessels were missed the first time. However, through careful microscopic examination and immunohistochemical staining, the mass was shown to be a neuroendocrine tumor. Another important alternative in the differential diagnosis is middle ear adenoma (MEA), which is thought to arise from pluripotent cells in the middle ear mucosa and may have mixed patterns of differentiation, ranging from glandular to neuroendocrine.[3] Because these tumors may have a neuroendocrine component, MEAs can be confused with paragangliomas. However,

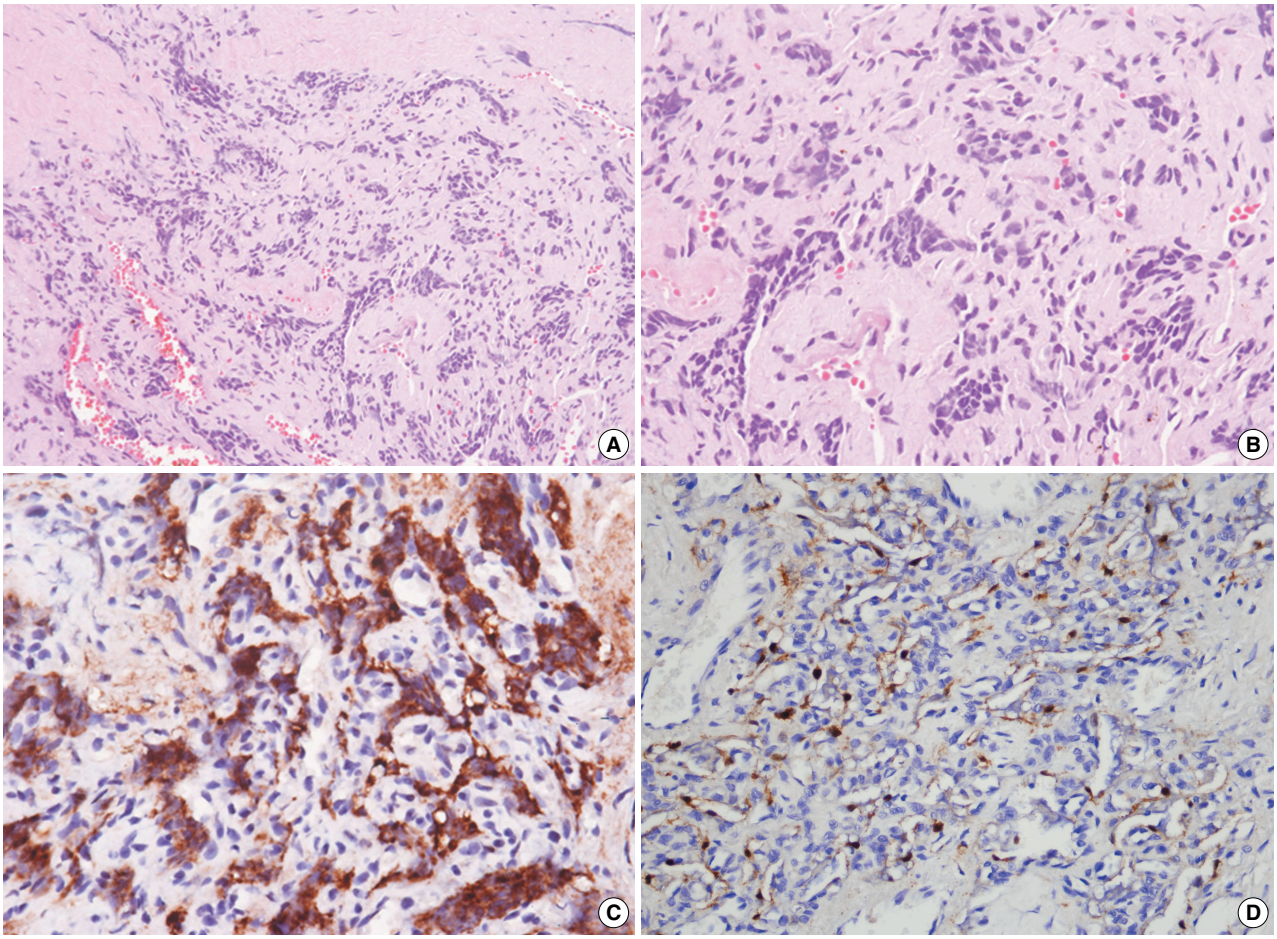


Fig. 2. (A, B) Tumor tissue with squeezing artifacts consists of small and less uniform hyperchromatic cells with increased vascularity and dense sclerotic stroma. These tumor cells show strong immunoreactivity for CD56 (C). And sustentacular cells surrounding tumor cell nests are positive for S100 (D).

Table 1. Clinical presentation of 9 Korean cases of jugular paraganglioma[7] and our case

| Case No. | Sex | Age (yr) | Side | Size (cm) | Initial symptom |
|---|---|---|---|---|---|
| 1 | F | 32 | Left | 1.5 × 1 | Pulsatile tinnitus, headache |
| 2 | M | 35 | Right | 3 × 3 | Facial palsy |
| 3 | M | 26 | Left | 3 × 2 | Pulsitile tinnitus, otalgia |
| 4 | M | 48 | Left | 2.2 × 2 | Hoarseness |
| 5 | M | 45 | Left | 4.5 × 3.5 | Hoarseness, chronic cough |
| 6 | F | 50 | Right | 2.2 × 2.5 | Sensorineural hearing disturbance |
| 7 | F | 60 | Right | 4.5 × 2.2 | Hoarseness |
| 8 | F | 29 | Right | 4.5 × 9 | Hoarseness, facial palsy |
| 9 | F | 42 | Right | 2.5 × 1.5 | Sensorineural hearing loss, pulasitile tinnitus |
| Our case | M | 18 | Right | 2.7 × 2.5 | Otorrhea, hearing disturbance |

F, female; M, male.

less vascularity and mixed patterns of differentiation of MEAs may be helpful to differentiate it from paragangliomas.

Head and neck paragangliomas can be functional and show hyperadrenergic syndrome despite being less frequent than pheochromocytoma or abdominal/pelvic paraganglioma.[4] So, in the patient showing hyperadrenergic manifestations without an adrenal, abdominal or pelvic mass, head and neck paraganglioma must be considered.

Only 15 reports in the English language literature are about malignant head and neck paragangliomas, including JTPs (Pub-

Med search). Kliewer *et al.*[5] previously investigated the aggressiveness of JTPs, and the amount of S100 or glial fibrillary acid protein expressed in sustentacular cells. The intensity of tumor cells for neuroendocrine markers was inversely related to tumor grade. Based on their results, our case, which showed well-preserved sustentacular cells and strong immunoreactivity for CD-56, chromogranin and synaptophysin was thought to be a low grade paraganglioma.

JTPs usually develop in adults in the 5th to 6th decade of life, but our patient was an 18-year-old boy. Although there have been no reports about the difference between familial and sporadic cases of JTPs, Kliewer *et al*.[5] reported that familial paragangliomas were found in significantly younger patients (mean age, 25.8 years) than in the rest of paraganglioma patients (mean age, 42.5 years). And, paragangliomas are occasionally associated with a variety of genetic multisystemic disorders such as von-Hippel-Lindau disease, multiple endocrine neoplasia type 2 and neurofibromatosis type 1.[6] So, we recommend evaluations regarding familial tendencies of paragangliomas and close follow up of such patients.

Recently, Chung *et al.*[7] published a clinical review of about 9 Korean cases of jugular paraganglioma (Table 1). The cases included 4 men and 5 women who underwent surgery between 1986 and 2005. Mean age at the time of diagnosis was 40.8 years (range, 26 to 60 years). They concluded that angiography with embolization is crucial for successful tumor removal without massive bleeding. Unfortunately, the authors didn't review the microscopic findings associated with their cases.

In conclusion, JTP is a rare tumor and can be missed or misinterpreted due to its different histology from pheochromocytoma or abdominal and pelvic paraganglioma. Hence, if we remember this entity and its characteristics, it is helpful when we interpret middle ear or temporal bone masses.

## REFERENCES

1. Borba LA, Al-Mefty O. Intravagal paragangliomas: report of four cases. Neurosurgery 1996; 38: 569-75.
2. Panda NK, Rao SS, Karuppiah S, Vaiphei K, Singh P. Epithelioid hemangioendothelioma of middle ear masquerading as glomus tumor. Am J Otolaryngol 2007; 28: 69-71.
3. Berns S, Pearl G. Middle ear adenoma. Arch Pathol Lab Med 2006; 130: 1067-9.
4. Al-Harthy M, Al-Harthy S, Al-Otieschan A, Velagapudi S, Alzahrani AS. Comparison of pheochromocytomas and abdominal and pelvic paragangliomas with head and neck paragangliomas. Endocr Pract 2009; 15: 194-202.
5. Kliewer KE, Wen DR, Cancilla PA, Cochran AJ. Paragangliomas: assessment of prognosis by histologic, immunohistochemical, and ultrastructural techniques. Hum Pathol 1989; 20: 29-39.
6. Suárez C, Sevilla MA, Llorente JL. Temporal paragangliomas. Eur Arch Otorhinolaryngol 2007; 264: 719-31.
7. Chung SM, Kim HS, Jung J, Lee HK, Lee WS. Clinical presentation and management of jugular foramen paraganglioma. Clin Exp Otorhinolaryngol 2009; 2: 28-32.