Middle Ear Carcinoid Tumor: A Report of Three Cases

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ABSTRACT

OBJECTIVES

Carcinoid tumors of the middle ear are an exceedingly rare yet indolent neoplasm. There are approximately 50 reported cases to date. The objective of this study is to describe our experience with three cases of middle ear carcinoid tumors.

METHODS

Three cases of adults with histopathological diagnosis of middle ear carcinoid tumors at a tertiary care academic center were reviewed.

RESULTS

First, was a case of middle ear carcinoid tumor in a 33-year-old female. She underwent a canal wall up tympanomastoidectomy for removal of a mass involving the middle ear and mastoid air cells. Following resection, the patient had minimal residual disease and no evidence of recurrence at last follow up visit. The second was a case of carcinoid tumor presenting in the middle ear of a 36-year-old male who had evidence of systemic tumor secretion and visible visceral metastases. The patient underwent a radical tympanomastoidectomy for removal of her primary tumor. The third was a case of previously partially removed carcinoid tumor in the middle ear of a 28-year-old man who presented with a local recurrence and underwent additional surgical resection with a canal wall up tympanomastoidectomy.

CONCLUSION

We found three of these rare tumors with a high index of suspicion and careful clinical, radiological and pathological evaluation. Surgical resection is the mainstay of definitive treatment and in the setting of delayed metastatic potential and possible local and regional recurrence, long-term follow up is recommended.

REFERENCES:


IMAGING

CASE 1

**CLINICAL PRESENTATION**

- A 33-year-old female referred to our department in April 2014 presented with complaints of right ear fullness and hearing loss for four months.
- The patient underwent a right middle ear exploratory tympanotomy in November 2014 for diagnostic and therapeutic purposes.
- Intra-operative frozen section analysis was concerning for a malignant process, but a final diagnosis could not be determined at that time.
- Final pathology was consistent with a neuroendocrine adenoma/carcinoid tumor.
- Metastatic work-up:
  - Patient underwent a whole-body nuclear medicine iodine-123 meta-iodobenzylguanidine (MIBG) scintigraphy scan to identify and localize possible metastatic disease.
  - Brain MRI was obtained to rule out other mass lesions.
- Both were negative for additional pathology.

**TREATMENT**

- The patient underwent a right canal wall down tympanomastoidectomy in March 2015.

**RESULTS**

- The patient had evidence of a near maximal conductive hearing loss on the right. The incus and stapes superstructure were removed, so this was an unexpected finding. At that time, the patient had no further evidence of recurrent or residual disease.

**DISCUSSION**

- Middle ear carcinoid tumors are rare but represent a distinct otologic disease.
- They commonly present as a nonspecific middle ear mass with an associated conductive hearing loss.
- Radiological evaluation is imperative to further characterize the primary tumor and to guide surgical planning.
- Metastatic work-up is variable, obviating the necessity of a standardized protocol with regards to imaging modality type and the optimal timing of obtaining them.
- Serological and urinary work-up should be initiated to identify bioactivity of the tumor, even in the absence of clinical symptoms of carcinoid syndrome.
- In the setting of tumor secretion, peri-operative management to prevent carcinoid crisis is essential, and the administration of somatostatin analogues such as octreotide is recommended.

**FOLLOW UP**

- Two months post-operatively, the patient had evidence of a near maximal conductive hearing loss on the right. The incus and stapes superstructure were removed, so this was an unexpected finding. At that time, the patient had no further evidence of recurrent or residual disease.

**CONCLUSION**

We found three of these rare tumors with a high index of suspicion and careful clinical, radiological and pathological evaluation. Surgical resection is the mainstay of definitive treatment and in the setting of delayed metastatic potential and possible local and regional recurrence, long-term follow up is recommended.