Is Squamous Cell Carcinoma of the Middle Ear a Complication of Chronic Suppurative Otitis Media?

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Abstract

Primary neoplasia of the middle ear is a rare condition and in many patients, the precise site of origin remains doubtful. More than one third of the middle ear squamous cell carcinomas [SCC] are preceded by chronic suppurative otitis media [CSOM]. The co-existence of CSOM with or without cholesteatoma along with malignancy poses several diagnostic and therapeutic challenges for the clinician. These tumors retain a poor prognosis despite the development of increasingly radical surgical procedures and the advances in radiotherapy. Hence it is very important to detect these lesions very early. A high index of suspicion and a thorough investigation that includes multiple biopsies in suspicious cases may be required for the same. In view of the close association between CSOM and SCC of the middle ear and the evidence supporting the malignant transformation, the latter may be considered a rare complication of the former. It may be possible to prevent this dreaded complication by timely medical and surgical treatment of CSOM at an early stage. This paper discusses the controversial association between chronic suppurative otitis media and malignancy of the middle ear.

INTRODUCTION

Carcinoma of the middle ear is an uncommon malignancy of the head and neck. Carcinoma following chronic suppurative otitis media [CSOM] is reported to be 1 case per 4000 cases of CSOM. The association between the two remains unclear and is yet to be studied in detail. The rarity of the condition has made large scale studies difficult. Even though squamous cell carcinoma [SCC] of the middle ear and CSOM co-exist, no definitive correlation has been proven. How does a long standing CSOM predispose to a SCC? Do cholesteatoma and granulation tissue of CSOM facilitate bone destruction and rapid spread of tumor? Or does the tumor facilitate the complications of CSOM? The co-existence of cholesteatoma and malignancy has raised all these questions. In this paper we study the clinicopathological profiles of 3 such rare cases.

This paper has been presented to highlight the controversial relationship between CSOM and malignancy of the middle ear and the diagnostic and therapeutic challenges associated with their combination.

MATERIALS AND METHODS

This was a prospective descriptive study conducted in a tertiary referral medical college hospital catering to both urban and rural population with a relatively high incidence of CSOM and its complications. The study period was from July 2003 to December 2005. All cases of CSOM with or without cholesteatoma and associated with SCC of the middle ear were included in the study in this period. Cases with complications of CSOM and SCC were also included in the study. Cases from all ages and both sexes were included. Three cases fulfilled the criteria out of the 255 total cases studied in the above period. Two of them had cholesteatoma and all of them had an aural polyp in the ear. Longstanding history of ear discharge could be obtained in 2 cases. Diagnostic details of the individual cases are given in Table 1.
All of them underwent investigations like high resolution CT scan [HRCT] of the temporal bone, otoendoscopy, pure tone audiometry [PTA], biopsy of the ear mass [HPE] and microbiology of the ear swab. The otoendoscopic picture of the mass in the ear canal of case 1 is shown in figure 1. Bone erosion and destruction was evident only in case 1 and case 2 and both of them had advanced disease at the time of presentation [Fig 2,3,4].

Figure 1: Otoendoscopic picture of the mass in the ear canal of case 1
Figure 5
Figure 4: HRCT of the temporal bone [Coronal section] of case 3 showing bone erosion

Figure 7
Figure 6: Pictomicrograph of the section of the ear mass in case 2 showing keratin debris, granulation tissue, tumor and lymphocytic infiltration [H & E stain X100]

Figure 8
Figure 7: Pictomicrograph of the section of the ear mass in case 3 showing granulation tissue and tumor [H & E stain X100]

**OBSERVATIONS**

Pseudomonas aeruginosa was the organism encountered in the ear swab of all the 3 cases. The biopsy report of all the three suspected cases read moderately differentiated SCC. Case 1 and case 2 additionally showed cholesteatoma, granulation tissue and lymphocytic infiltration suggestive of chronic inflammation. The co-existence of CSOM, cholesteatoma and SCC is well demonstrated in the same tissue section [Fig 5,6,7].

Figure 6
Figure 5: Pictomicrograph of the section of the ear mass in case 1 showing keratin debris, granulation tissue, tumor and lymphocytic infiltration [H & E stain X40]

Intraoperatively, it was found that the tumor could not be delineated from the disease of otitis media and was inseparable from the cholesteatoma and granulation tissue. This indicates the common region of involvement of the 2 disease conditions. The tumor along with the granulation tissue was found to enter and involve the cochlea through the round and oval windows. This must have lead to the severe sensorineural hearing loss in case 1 and case 2. The latter even showed a fistula in the lateral semicircular canal. Case 1 and case 3 underwent a radical mastoidectomy and case 2 a lateral temporal bone resection. All of them were treated with postoperative radiotherapy and chemotherapy. Case 1
and case 2 had a remission period of 2 years after which there were signs of tumor recurrence. Case 3 has been free of symptoms up to this date.

DISCUSSION

SCC accounts for 60 to 80% of the carcinomas of the temporal bone. Carcinomas of the middle ear histologically are most often of the squamous type and often associated with a history of chronic or recurrent ear infection with discharge for a long period of time. The coexistence of CSOM with or without cholesteatoma along with malignancy poses several diagnostic and therapeutic challenges. Middle ear cancer masquerades CSOM for at least 6 months before a diagnosis of malignancy is made. A pre-existing CSOM can further delay the diagnosis. After the diagnosis is made, the exact site of primary tumor is difficult to ascertain as the tumor is considerably advanced at the time of presentation. Further, it becomes difficult to demarcate the tumor from the normal tissue as there is long standing inflammation of CSOM all around the region. The extent of tumor is difficult to ascertain even on radiography since both cholesteatoma and malignancy are liable to cause destruction of the tissue and bone even though the latter may be more aggressive. Some of the cases would have previously undergone mastoid surgery for CSOM and that could complicate the picture further. Both CSOM and malignancy can present with an aural polyp as was seen in our cases and could be mistaken for the polyp of the former that is much commoner, unless a biopsy is taken. Fortunately, there are other warning symptoms of a development of malignancy in an ear with CSOM like severe earache, bloody discharge and facial palsy.

It is widely accepted that one of the most commonly identified predisposing factors in tumor development is chronic suppuration and in most series, the duration of ear discharge has been long, in the region of 20 years. Lodge et al considered that malignancy developed from cellular metaplasia subsequent to infection. Michaels and Wells doubt this theory, referring to the high frequency of CSOM contrary to the low frequency of middle ear carcinomas. Besides malignancy is not a common complication of CSOM and hence doesn't even find a mention at most places in the long list of complications of otitis media. Nevertheless, several studies have reported the much higher incidence of the malignancy of the middle ear in the presence of longstanding CSOM than without it. Conley raised the possibility that the biologic features that predispose to chronic recurring infections may be more responsible for the development of cancer than the infection per se. Kenyon et al confirmed that a history of chronic suppuration with or without cholesteatoma predisposed the patient to tumor development and that, in the presence of continuing otorrhea, patients were not protected by mastoid surgery.

There are no clear-cut theories in literature on the facilitation of the genesis of malignancy by CSOM or cholesteatoma. Lodge et al suggested that chronic otitis might promote the development of a carcinoma in the middle ear in a manner analogous to the development of carcinoma of the skin adjacent to draining sinuses resulting from chronic osteomyelitis [Marjolin's ulcer].

In 2 of our cases, cholesteatoma, granulation tissue and carcinoma co-existed in the same histopathological tissue section. This supports the hypothesis that the carcinoma had probably originated from the cholesteatoma epithelium itself in these cases.

Surgery is the primary modality of treatment and lateral temporal bone resection is the commonly performed surgery. In any definitive surgical procedure for the resection of the mass, it may be extremely difficult or impossible not to violate a tumor margin, especially if the tumor size is underestimated by the imaging studies. The situation may only get worsened when malignancy is superimposed on pre-existing chronic otitis media. Despite the advances in surgery and radiotherapy, the prognosis of most cases remains bad because of late presentation and rapid intracranial spread. The poor prognostic factors include facial nerve paralysis, positive tumor margins, dural involvement, regional lymph node involvement, multiple cranial involvement and severe pain. The 5 year survival rate remains to be 25 to 35%.

CONCLUSION

In an ear with longstanding or recurrent CSOM, a sudden onset of sinister symptoms like severe earache, bleeding or facial palsy should alert the development of a malignancy. Early diagnosis of malignancy in such a case rests on a high index of suspicion and a thorough investigation that should always include multiple biopsies of the suspicious areas. Radiography is useful to indicate the extent and type of bone erosion. In view of the close association between CSOM and SCC of the middle ear and the intraoperative and histopathological evidence supporting the genesis of the malignancy from the same epithelium, the latter may be considered a rare complication of the former. It may be
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possible to prevent this dreaded complication by timely medical and surgical treatment of CSOM at an early stage.

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