

# A Rare Association of Deaf Mutism, Syndactyly and Squamous cell carcinoma of the Temporal bone

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## Abstract

Malignant tumours of the temporal bone are rare, the majority being squamous cell carcinoma. We here report a rare syndromic complex of squamous cell carcinoma of the temporal bone, deaf-mutism & syndactyle in a 12 year old male child, presented in Department of Otorhinolaryngology, Hamidia Hospital, Bhopal, with one month history of mass in left external auditory canal & lower motor neuron facial nerve palsy on a background of left chronic suppurative otitis media since childhood.

## INTRODUCTION

Malignant tumours of mastoid and middle ear are rare, accounting for the 5-26% of all ear neoplasms<sup>1,2</sup>. Of these neoplasms, squamous cell carcinoma is most common with an incidence of one case in one million and peak age of 60 years<sup>1,2</sup>. We here reported a rare presentation of a squamous cell carcinoma of the temporal bone in a 12-year-old child in association with syndactyly and deafness mutism.

## CASE REPORT

A 12-year-old deaf & dumb child presented in department of ENT with mass in left ear and left facial nerve palsy since one month. He had a history of intermittent recurrent left ear discharge since childhood.

Examination showed a fleshy granular mass filling the left external auditory canal with swelling in lower part of pre and postauricular region (fig 1). Tympanic membrane was not seen. Patient also had a complete infranuclear facial palsy. Bony syndactyly of right hand was also present. (fig 2).

## Figure 1

Figure 1: Showing fleshy granular mass in left external auditory canal



**Figure 2**

Figure 2: Syndactyly of right hand with facial palsy left side

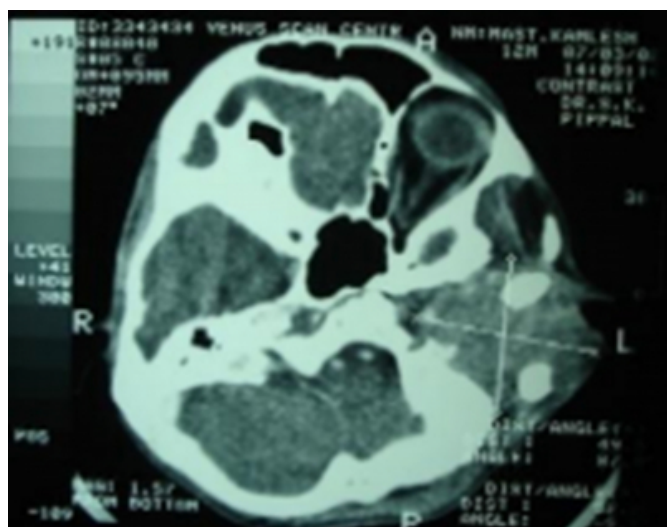


Histology of mass was suggestive of moderately differentiated squamous cell carcinoma grade 2.

CT Scan revealed large inhomogenous density heterogeneously enhancing mass in the region of the left external auditory canal with complete destruction of the bony part of the auditory canal and infra-temporal extension up to the base of the skull with destruction of the left temporomandibular joint (fig 3). Left nasopharyngeal space was compressed by the mass. Medial extension was seen till the middle ear cavity with destruction of the ossicular chain. Left mastoid was also eroded by the mass. Inner ear structures were normal.

**Figure 3**

Figure 3: Showing mass in the region of left external auditory canal with complete destruction of the bony part of auditory canal



## DISCUSSION

Carcinoma of temporal bone is a rare tumour, though the peak age incidence is 60 year. However, cases have been reported in children as young as 8 years<sup>3</sup>. It is usually a unilateral disease however bilateral cases has been reported<sup>4,5</sup>. Chronic otitis media has been implicated as the main etiological factor in this tumour. Hence, the need for a detailed and regular evaluation of such patients. Radiation is another factor implicated.

More recently Jin et al. have shown that there is high prevalence of human papilloma virus (HPV) 16 and 18 at both the tissue level and molecular level of patients with middle ear squamous cell carcinoma.

Middle ear squamous cell carcinoma should be considered when refractory granulation, long-standing otorrhoea, otalgia and facial nerve palsy are observed. Absence of otorrhoea in the history indicates that the origin of tumour is in the external auditory canal.

Standard treatment for early tumour is extended total petrosectomy with postoperative radiotherapy, however the value of such major surgery for palliation with significant mortality and morbidity is rarely profitable in terms of cure. In younger patient especially with more or limited disease, a lateral or subtotal temporal resection may be considered. These procedures are not always curative but may provide the relief of the severe pain, which is so often, the most distressing aspect of the problem. Sole radiotherapy is mainly used for the palliation where surgery is made impossible because of poor general condition or very tiny tumour. Most authors advocate full course post-operative radiation to stage T3 or T4 tumors as defined by University of Pittsburgh staging system.

In this case there was a history of long standing chronic otitis media in a deaf and mute child. Examination showed a mass in external auditory canal, infranuclear facial nerve palsy and syndactyly. Histological confirmation was done by punch biopsy from mass in the external auditory canal & accurate assessment of the extent of tumor is by high resolution CT scan. As tumour was extensive so surgery was not done. Chemotherapy followed by radiotherapy was given and tumor reduced in size but did not completely dissolve. More research needs to be carried out with regards to the association of the clinical features in this case.

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