Case Report:
Adenoid Cystic Carcinoma of External Auditory Canal
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URL: http://www.ojhas.org/issue41/2012-1-19.htm

Submitted: Jan 31, 2012; Accepted: Mar 18, 2012; Published: Apr 15, 2012

Abstract: Adenoid cystic carcinoma is extremely rare tumour that accounts for approximately 5% of primary malignancy of external auditory canal. These tumours are related with a high risk of recurrences and significant morbidities from surgical management and adjuvant radiotherapy. Despite the aggressive management for these tumours, many patients succumb to distant metastasis, making overall prognosis of these tumours poor. Although ACC of EAC has been reported in 5th decade, but its occurrence in young patient is very rare. We report a rare case of ACC in a young 22 years old female, who presented with ear canal mass and ear pain. Biopsy suggested mass to be ACC. Patient underwent wide local excision followed by adjuvant radiotherapy.

Key Words: Adenoid cystic carcinoma; External auditory canal; Malignant tumour.

Introduction:
Malignant tumours arising primarily from external auditory canal (EAC) are rare, with squamous cell carcinoma being the commonest. Glandular tumors accounts for 20% of EAC tumours, with adenoid cystic carcinoma (ACC) being exceptionally rare.[1-3] Earlier diagnosis of these tumours is of utmost importance, in view of the fact that delays in diagnosis may increases the risk of distant metastasis.[4] Appropriate imaging followed by biopsy should be considered early on in every patient presenting with otalgia, ear canal mass or chronic otorrhea.

Aggressive surgical resection with adjuvant radiotherapy is the standard treatment for these tumours.[4-6] The role of elective parotidectomy and its appropriate surgical extent remain a matter of controversy.[7,8] Despite these aggressive approaches, local recurrences and metastasis to the cervical lymph nodes, lungs, bones and liver can occur over many years.[9]

We report a rare case of ACC of EAC in a young 22 years old female, who presented with ear pain and EAC mass. Patient was managed by wide local excision and split thickness graft followed by adjuvant radiotherapy. Patient has been on regular follow-up for 2 months without any local recurrences and distal metastasis.

Case Report:
A 22 years old female referred to ENT OPD with one year history of a painful mass in her right EAC. She also had history of hearing loss, which was gradual in onset and progressive. She had one episode of ear discharge 6 months back, which stopped spontaneously. She had no history of tinnitus, bleeding from ear and vertigo. No previous history of surgery, trauma was noted. She underwent FNAC twice from private hospital, which was inconclusive. Physical examination showed approximately 1X1cm flat lesion on the posterior and inferior wall of the EAC, partially occluding the lumen of EAC (Figure. 1).

A contrast enhanced computed tomography (CECT) scan of temporal bone revealed isodense enhancing mass lesion measuring 2.1X1.8X1.5 cm arising from posterior and inferior canal wall, with no erosion of bone (Figure. 2). Biopsy of the swelling was performed. Histopathological examination revealed adenoid cystic carcinoma. The tumour was classified as a stage 1 (TNM0Mx) lesion based on university of Pittsburg TNM staging for EAC carcinoma. The patient underwent wide local excision, and reconstruction with split thickness graft, followed by adjuvant radiotherapy. Histopathology of the specimen revealed, tumour arranged in a cribriform and reticular pattern with cartilage destruction & perineural invasion, confirming the diagnosis of ACC.
local treatment. currences and distant metastasis occur even after successful avoidance distant metastasis. Long term follow-up is required, as re-emphasis upon the need for early diagnosis, as this may help torate imaging as FNAC could be inconclusive. We would like toAny patient presenting with otalgia, an ear canal mass or chronic ease at last follow up. A predilection for metastasis to the lungs10 patients[8] and 10 of 22 patients[4] with no evidence of disranged in a variety of patterns. The classical pattern consists of acribriform architecture admixed with areas of tubule formaerineural involvement. ACC may appear as a polypoidal mass, epithelial ulceration, associated granulation tissue, and poor de-marication from the surrounding tissue.[11]Microscopically these tumours composed of epithelial cells ar-anged in a variety of patterns. The classical pattern consists of acribriform architecture admixed with areas of tubule forma- tion or area of cellular growth. Perineural invasion is a charac-teristic and diagnostically helpful feature of adenoid cystic carcino-ma.[11]Aggressive surgical resection with adjuvant radiotherapy is thestandard treatment for local disease control.[4-6] Depending upon the clinical staging, surgical intervention varies from lat-eral temporal resection to total temporal resection. The role of elec-tive parotidectomy in improving the survival is controver-sial.[5] Many surgeons do prefer parotidectomy in dealing withEAC carcinoma.[12,13] while one report says that survival rate did not differ according to the performance of parotidectomy.[7] The involvement of parotid by tumour, positive surgical mor-bids, nerve, and bone involvement are important prognostic factors.[2,10]In spite of definitive treatment, patients with ACC of EAC of-ten experience recurrences. Metastasis to regional lymph nodes and distant sites are well-documented.[2-4] The larger studies to date showed 7 of 21 patients[14], 7 of 16 patients[10], 5 of 10 patients[8] and 10 of 22 patients[4] with no evidence of disease at last follow up. A predilection for metastasis to the lungs was noted in 2 of these series.[4,14]Discussion:Malignant tumours arising from the EAC are extremely rare with more than 80% being squamous cell carcinoma and ACC accounting for approximately 5%. [4] The mean age for ACC reported in the literature is fifth decade, and is two times more common in women than men.[4,8] But in our case the patient was much younger for mean age of ACC, than reported in the literature. Majority of the patients presenting with severe ear-pain of prolonged duration and mass in the ear canal.[10] The clinical presentation was similar in our case. The cause for ear-pain can be explained by tendency of these tumours for early perineural involvement. ACC may appear as a polypoidal mass, epithelial ulceration, associated granulation tissue, and poor del-marication from the surrounding tissue.[11]References: