Case Report

Adenoid Cystic Carcinoma of Cervix: Treatment Strategy

ABSTRACT

Adenoid Cystic Carcinoma (ACC) of cervix behaves aggressively and it has a propensity for local infiltration, invasion in lymphatic vessels as well as perineural spaces, with subsequent haematogenous spread. In advanced disease (Stage III and IV), outcome is invariably poor. Moreover, these cases have a high tendency to recur locally and also to metastasize to distant sites. There are no standard treatment protocols because of rarity of disease. The guidelines for its management are established similarly as for staged squamous cell carcinoma of cervix. Therefore, recommended treatment in advanced disease is aggressive multimodality treatment, which includes chemotherapy and radiotherapy. We are presenting a case of adenoid cystic carcinoma of cervix in 55 years old patient, staged as III B. The patient was treated with concomitant radiotherapy and chemotherapy. The patient responded well to treatment and is disease free, three years after treatment.

BACKGROUND

ACCs arising from female genital tract are very rare and they constitute less than 0.1 % of all malignancies of the cervix [1]. Local infiltrations, vascular, lymphatic and neural invasion, with subsequent haematogenous spread, are its characteristics features [2]. These cases usually follow the guidelines which are established for similarly staged patients with squamous cell carcinoma of cervix. Surgery and radiation therapy, either alone, or in a combined setting, have been used in the treatment of these cases. Considering high local failures and distant relapses, an aggressive local and systemic treatment is recommended in advanced stage and inoperable disease.

CASE REPORT

A 55-year-old postmenopausal woman presented with complaints of watery discharge per vagina, burning micturition of 4 months duration. Discharge was purulent in nature, sometimes it was blood tinged. General physical examination was normal. Per speculum and per vaginal examination showed an ulceroproliferative growth arising from cervix, involving all fornices, and extending up to upper half of vagina. The disease extended up to lateral pelvic wall on either side. Per abdomen examination was normal. Blood investigations, renal function test and liver function test were normal. Chest X-ray was normal. Ultrasound examination of abdomen and pelvis revealed a mass over the cervical region. Histopathology examination of growth cervix showed adenoid cystic carcinoma [Table/Fig-1]. The patient was finally diagnosed as having adenoid cystic carcinoma cervix, stage III B, according to classification of International Federation of Gynecology and Obstetrics (FIGO).

The patient had an inoperable disease, so she was planned for radical radiotherapy with dose 50 Gray in 25 fractions, one fraction per day, five days a week over 5 weeks. Per fraction dose was 200 centiGray. Radiation was given with chemotherapy. Chemotherapeutic agent was Injection Carboplatin which was given concomitantly (when both modalities i.e Radiation and Chemotherapy were given simultaneously). Concomitant Carboplatin 150 mg was given on day of start of radiotherapy, on 7th, 14 th, 21th and 28th days of radiation; a total of five courses were administered. One week after completion of this treatment, the patient was given high dose intracavitary brachytherapy. Dose of brachytherapy was 7 Gray per fraction, once a week, for three weeks. Total 21 Gray was prescribed to point A by high dose rate brachytherapy. The patient is asymptomatic, with no evidence of disease at primary as well as distant sites, after three years of completion of treatment.

DISCUSSION

In 1856, Billroth first described Adenoid Cystic Carcinoma (ACC) [3]. Paalman and Conseller first reported that adenoid cystic carcinoma appeared in cervix in 1949 as Cylindroma [4]. Adenoid Cystic Carcinoma of cervix is characterized by highly aggressive behaviour. Therefore, outcome in advanced disease (stage II and IV) is invariably poor. Management of cervical ACCs comprises surgical intervention as the initial strategy for early stages; radiation therapy is usually recommended as adjuvant treatment. The role of chemotherapy

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[Table/Fig-1]: Photomicrograph showing cribriform pattern – Adenoid Cystic Carcinoma. (H& E 4 x)
is undefined as adjuvant or primary treatment, although in some series, chemotherapy has been used for recurrent or metastatic disease [1,2]. ACC of cervix is considered as radiosensitive tumour and better results have been seen in reported cases as compared to those seen in cases where surgery has been done alone, in early stages [5,6].

Distant metastases are the main determinant of the survival. Therefore, majority of patients require aggressive local and systemic therapy, considering high local and distant relapses.

Based on these recommendations, the present case was treated with concurrent chemoradiation. Chemotherapeutic agents which have been investigated are bleomycin, Adriamycin and 5 FU. However, cisplatin is most commonly used agent.

Ethassani et al., reported 13 case of Staged III B ACC cervix in 2008 and they claimed that the first case was successfully managed with concurrent chemoradiation [7]. Nishida et al., treated a case of stage III B with radiotherapy alone; the patient showed no evidence of recurrent tumour at 5 years after radiotherapy [8]. Dixit et al., reported eight patients of Stage III who had taken radiotherapy treatment. Only one patient remained disease free at 11 months and none remained disease free for more than two years, with NED rate of 12.5% (l/8) [9].

CONCLUSION

ACC of cervix is a rare entity and it is considered as a radiosensitive tumour, but at same time, highly aggressive biological behaviour of the tumour leads to early recurrences and metastases. In absence of optimal treatment guidelines, the patient was treated as per treatment protocol for stage III B Squamous cell carcinoma of cervix. The patient had an inoperable disease, so she was treated with concomitant chemoradiation.

Single agent chemotherapy with carboplatin 150 mg weekly was given to the patient. The patient is asymptomatic with no evidence of disease at primary as well as distant sites, after three years of completion of treatment. The appropriate management which includes CRT, may have a major impact on the survival of advanced cases of ACC of cervix.

REFERENCES