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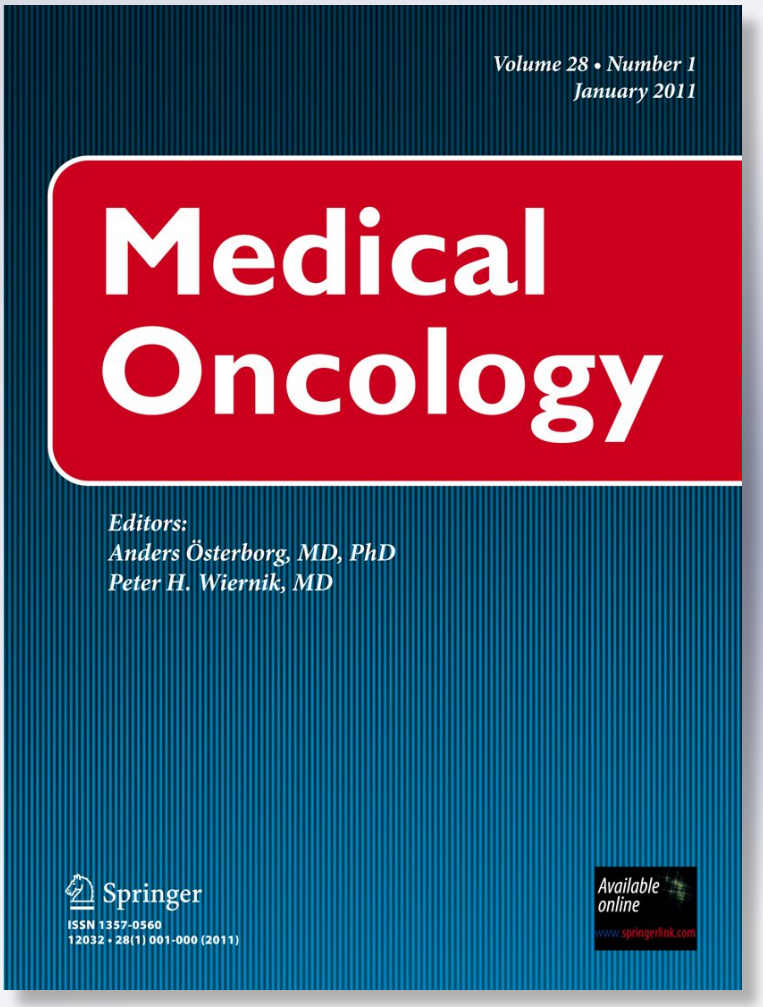

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REVIEW ARTICLE

Primary cutaneous small-cell carcinoma: a case report

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Abstract Most commonly arising in the gastrointestinal and genitourinary tracts, extrapulmonary small-cell carcinoma is quite rare. Although its prognosis is very poor, complete remission or even cure can be achieved by combination of different treatment modalities. We report here a 32-year-old woman with a cutaneous gluteal mass diagnosed as small-cell carcinoma of the skin. Combination chemotherapy containing cisplatin and etoposide was started. Radiotherapy was administered after two courses of chemotherapy. Following radiotherapy, additional 4 courses of chemotherapy were given. She has been in remission for three years with no evidence of tumor recurrence.

Keywords Cutaneous · Small-cell carcinoma · Chemotherapy · Radiotherapy

Introduction

Small-cell carcinoma commonly originates from lungs. However, it can also arise from the extrapulmonary sites. Constituting 2.5% of all small-cell cancers, extrapulmonary small-cell carcinoma (EPSCC) is very rare with an incidence of 0.1–0.4%. It was initially described by Duguid and Kennedy in 1930 [1]. The most common sites of involvement are gastrointestinal and genitourinary tracts. There is limited knowledge about its clinical course and prognosis. Although its prognosis is poor, complete remission or even cure can be achieved by combination of different treatment modalities [2].

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Case report

A 32-year-old woman was admitted to another hospital because of a cutaneous gluteal mass. After total excision of the mass, she was referred to our hospital for further management. Histopathological examination was consistent with small-cell carcinoma of the skin. Positron emission tomography–computed tomography (PET-CT) was performed, and recurrence and metastatic focus were not found. Combination chemotherapy containing cisplatin and etoposide was started. Radiotherapy was administered after two courses of chemotherapy. Following radiotherapy, additional 4 courses of chemotherapy were given. After completion of therapy, she has been in complete remission for three years with no evidence of tumor recurrence.

Discussion

Although the natural course of EPSCC resembles small-cell carcinoma of the lung, it is a completely distinct entity. The most important factor affecting the prognosis is the stage of the disease [2]. Different primary localizations of EPSCC have been reported as case reports. These are esophagus [3] and colon [4] in gastrointestinal tract, bladder [5] and cervix [6] in genitourinary tract, and larynx [7] in head and neck region. EPSCC with unknown primary has also been reported [8]. The frequency of these localizations is shown in Table 1. Cutaneous EPSCC must be differentiated from primary metastatic tumors of skin [9]. Reported only as case reports or a part of retrospective

Table 1 The frequency of the sites of involvement

Frequency of EPSCC per site of origin	
Localization	Percentage of SCC/total per site of origin (%)
Pulmonary	15–20
Esophagus	0.8–2.4
Larynx	0.5–1
Bladder	0.3–1
Cervix	1
Prostate	2
Unknown primary	7–30

small series, our knowledge about cutaneous EPSCC is extremely limited, even compared to other locations. Furthermore, the prognosis of this rare entity is so poor that long-term survival is usually not expected [10]. In a study by Taxy et al., radiotherapy was administered for the second time and systemic chemotherapy was given thereafter due to recurrence of lesions 3 months after first radiotherapy [10]. Despite the combination of chemotherapy and radiotherapy, the patient died at the 13th month of follow-up.

Treatment options are based on the scarce information coming from case reports and small case series. Randomized clinical trials are absent because of the limited number of the patients. Locoregional disease can be managed as in tumors of similar regions. However, despite an aggressive locoregional treatment composing of surgical excision and/or radiotherapy, recurrence is common and adjuvant chemotherapy is recommended. First-line chemotherapy regimens include drug combinations such as cisplatin and etoposide (EP), irinotecan and cisplatin (IP), carboplatin and etoposide (CE), cyclophosphamide, adriamycine, vincristine (CAV), and etoposide (ACE), ifosphamide, carboplatin, and etoposide (ICE). Despite adjuvant treatment, prognosis remains poor and many patients develop systemic metastatic disease [9]. In EPSCC originating from salivary glands and paranasal sinuses in the head and neck region, prognosis is better and local recurrence is more common than distant metastasis. In the present case, we administered chemotherapy and radiotherapy similar to the strategy in limited-stage small-cell lung cancer. Platinum-based chemotherapy protocols are recommended in these cases. Response to treatment is usually partial and of short duration in systemic EPSCC. The efficacy of maintenance therapy has not been shown. Topotecan, taxane, or gemcitabine have been used as single agents in patients with recurrence and good performance status. Due to low incidence of symptomatic brain metastases, prophylactic brain irradiation is not recommended except for EPSCC cases originating from head and neck region [9].

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