Abstract: Merkel cell carcinoma is a rare and aggressive neuroendocrine carcinoma of the skin with tendency for rapid progression and nodal and distant metastases at presentation. The rarity of the tumour and need for multidisciplinary approach make the treatment decisions difficult. Here we report a case of Merkel cell carcinoma of the gluteal region, requiring multidisciplinary management.

Keyword: Merkel cell, multidisciplinary, gluteal region

INTRODUCTION

Merkel cell carcinoma is a rare and aggressive neuroendocrine carcinoma of the skin with tendency for rapid progression and nodal and distant metastases at presentation. The poor chances of survival with surgery alone and the systemic nature of the disease, have required multidisciplinary treatment for management of this tumour.

CASE SUMMARY

Merkel cell carcinoma is rare tumour with only a single case reported at our Institute over a period of 8 years. Here we report a 42 year old gentleman, who presented with left gluteal swelling of 5 months duration. He underwent excision elsewhere and final histopathology revealed a 6*6 cm tumour suggestive of well differentiated liposarcoma. He presented to us for further management. On examination, he had a 7cm vertical scar in left gluteal region with minimal induration. There were bilateral subcentimetric inguinal nodes. The paraffin blocks reviewed at our Institute were suggestive of a neoplasm with epithelial appearance. Immunohistochemistry revealed cells positive for keratin, desmin, S100-P, CD 57 and NSE. Chromogranin and synaptophysin were negative. The IHC was suggestive of Merkel cell carcinoma. MRI of the pelvis revealed T2 hyperintense lesion with surrounding hypointense rim measuring 4.3*3.6*5.0 cm in the skin and subcutaneous plane. Metastatic work up was negative. He underwent wide local excision of the left gluteal scar with superficial inguinal bloc dissection. The histopathology revealed foreign body granuloma with no evidence of malignancy. All 6 left inguinal nodes were free of tumour. After discussion in the multidisciplinary tumour board, he received adjuvant External Beam Radiation Therapy, total dose 62 Gy to local site and presently is under regular follow up.

DISCUSSION

Merkel cell carcinoma is a rare and highly aggressive skin cancer, known for its propensity to metastasize early. Merkel cell carcinoma affects predominantly the Caucasian population and is more frequent in men. MCC tends to affect the elderly, with a median age at presentation between 67 and 76 years. Few cases have been reported below 50 years and are usually related to immunosuppression. It usually occurs in sun-damaged skin of the head and neck region or extremities, but it also involves sun-protected sites, such as the buttocks. Pathogenetic factors such as UV irradiation may contribute to tumour development. A polyomavirus has been identified that is integrated into the genome of MCC and may play a role in the pathogenesis of these tumors.

Typically MCC has a tendency to progress rapidly, and in just a few months, the tumour may attain a large diameter with early and frequent metastasis to the regional lymph nodes. Therefore, early diagnosis and surgery are strongly advocated. Historical data have reported dismal 5-year survival rates ranging from 29% to 64%, 6. However, studies involving larger number of patients have suggested up to 74% survival rates at 5 years. 8. The most important prognostic determinant is tumor stage, which is most accurate if information of the pathologically confirmed status of the regional lymph nodes is included. Several studies established a close correlation between overall survival and tumour size. Size greater than 2 cm has known to affect survival.

HISTOLOGY

Three histopathologic patterns have been described, a trabecular type with connective tissue separating interconnecting cellular trabeculae, an intermediate cell type (most common) consisting of solid nests with trabeculae at the periphery, and a small cell type (least common) consisting of sheets of small cells with a diffusely infiltrative pattern. The cells in MCC have scant cytoplasm and round to oval nuclei that usually measure 14 to 20 μm in diameter and show finely dispersed chromatin.
Merkel cell carcinoma expresses low-molecular-weight cytokeratins (keratins 8,18,19 and 20), the simple epithelial type being the most marked. CK-20, a low-molecular-weight intermediate filament, is a highly sensitive marker, staining positively in a paranuclear, dot-like pattern. However it is not specific. In 5 – 25% of merkel cell carcinoma cases, CK-20 is negative 11. Absence of CK 7 and TTF-1 differentiate it from small cell lung carcinoma 10. Other markers with a high sensitivity for merkel cell carcinoma include neuron-specific enolase, chromogranin A, synaptophysin, BER-EF-4, and CAM 5.2. CD56, or neural cell adhesion molecule (NCAM), has recently been demonstrated to be a neuroendocrine marker of the pulmonary neuroendocrine cell system as well as merkel cell carcinoma 11.

REFERENCES

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University University Journal of Surgery and Surgical Specialties