G Chir Vol. 40 - n. 4 - pp. 364-367
July-August 2019

clinical practice

Jejunal metastasis of Merkel cell carcinoma: case report

S. MANCINI[1], L. SOLINAS[1], L. LEONE[1], B. BATTAGLIA[1], M.G. NOTARANGELO[2], R. HASSAN[2], C. DI COSIMO[1], R. ANGELONI[1], A. BELARDI[1], L. COSENTINO[1], A. BAKACS[3], A. SAGNOTTA[1,4]

SUMMARY: **Jejunal metastasis of Merkel cell carcinoma: case report.**

S. MANCINI, L. SOLINAS, L. LEONE, B. BATTAGLIA, M.G. NOTARANGELO, R. HASSAN, C. DI COSIMO, R. ANGELONI, A. BELARDI, L. COSENTINO, A. BAKACS, A. SAGNOTTA

Merkel cell carcinoma (MCC) of the skin is a rare but aggressive cutaneous neuroendocrine-derived malignancy that predominantly affects elderly white males. The presence of distant nodal metastases significantly impacts survival. Typical metastatic sites of MCC are liver, bone, brain and skin. Gastrointestinal metastases are uncommon and small bowel is the most common site followed by stomach. We report a case of symptomatic MCC jejunal metastasis.

KEY WORDS: Merkel cell carcinoma - Jejunal MCC metastasis - Small bowel obstruction.

Introduction

Merkel cell carcinoma (MCC) of the skin is a rare but aggressive cutaneous neuroendocrine-derived malignancy that predominantly affects elderly white males (1). MCC has a propensity for local recurrence and regional lymph node metastases. The aetiology is likely multifactorial: immunosuppression, UV-induced skin damage and viral factors, such as Merkel Cell Polyomavirus (MCPyV) contribute to its development (2, 3). MCC typically presents as a rapidly enlarging, painless red to purple nodule located on sun-exposed areas such as head, neck and arms (4). Although rare, cases of MCC on non sun-exposed skin have been documented and typically have worse prognosis (5). This malignancy affects almost exclusively Caucasian population, with 98% of all cases, suggesting possible protection by darker skin pigmentation (6). Prognosis of MCC depends on the original MCC localization, sex, age and other comorbidities. The presence of distant nodal metastases significantly impacts survival. Typical metastatic sites of MCC are liver, bone, brain and skin (7). It is very uncommon for MCC to metastasize to the gastrointestinal (GI) tract and small intestine is the most common site followed by stomach (8). In those cases, MCC is easily confused with primary tumors of visceral organs (9-14). Histopathologic diagnosis frequently requires support by immunohistochemistry. We hereby describe a patient with symptomatic MCC jejunal metastasis.

Case report

A 72-year old caucasian woman was admitted to our Department with mesogastric pain, nausea and vomiting during the last 3 months. In 2015 the patient underwent excision of a Merkel cell carcinoma on right gluteus followed by adjuvant radiotherapy.

[1] Department of General Surgery and Surgical Oncology, San Filippo Neri Hospital, Rome, Italy
[2] Department of Surgical Sciences, Faculty of Medicine and Dentistry, "Sapienza" University of Rome, Rome, Italy
[3] Department of Histopathology, San Filippo Neri Hospital, Rome, Italy
[4] Surgical and Medical Department of Translational Medicine, Sant'Andrea Hospital, Faculty of Medicine and Psychology, "Sapienza" University of Rome, Rome, Italy

Corresponding author: Andrea Sagnotta, e-mail: andrea.sagnotta@aslroma1.it

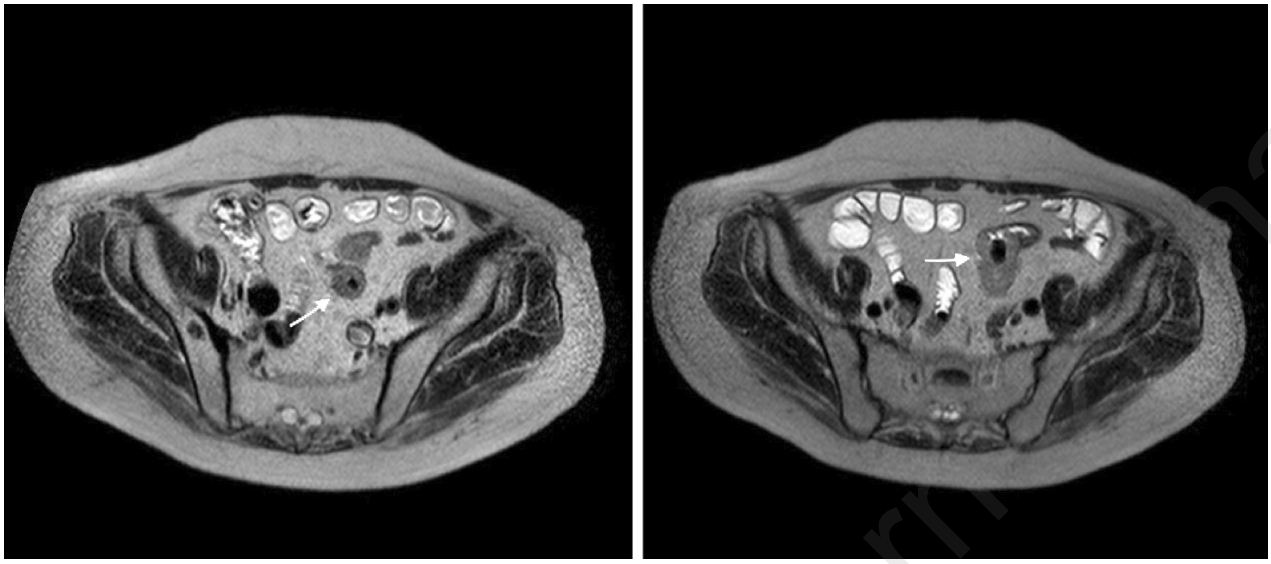

Figure 1 - Entero-MRI findings showing a long stenosis (arrows) of proximal jejunum.

EGDS and CT scan did not show any evidence of pathological findings. An entero-MRI showed a 6 cm long stenosis of proximal jejunum with contrastografic enhancement and locoregional lymphoadenopathies (Figure 1). Patient underwent laparotomy. A moderate duodenojejunal dilatation due to fourth jejunal loop stenosing lesion was found. Small bowel resection with an isoperistaltic JJ laterolateral anastomosis was then performed. Postoperative course was uneventful and patient was discharged eight days after surgery. Histology demonstrated a small bowel wall full-thickness involvement with mucosal ulceration, moderate lymphocytic infiltrate and necrosis areas. Microscopically, hematoxylin and eosin staining showed widespread proliferation of monomorphic tumor cells, nuclei with finely granular chromatin, small nucleoli, numerous mitotic figures and necrosis areas (Figure 2 A, B). Immunohistochemistry was positive for Neuron Specific Enolase (NSE), CD56, Chromogranin, Synaptophysin and CK20 (Figure 2 C). Based on these findings, a Merkel cell carcinoma jejunal metastasis was finally diagnosed. Locoregional lymphnodes and surgical margins were disease free. After a 1 year follow-up the patient is without evidence of disease.

Discussion

MCC is a rare but highly aggressive cutaneous reddish cancer affecting elderly white males (1). It was first described by Toker in 1972 as trabecular carcinoma (15). Rarely a Merkel cell carcinoma is suspected before surgery due to its aspecific and benign clinical appearance (11). If MCC is suspected based on initial hematoxylin and eosin staining (H&E) examination, further confirmation should be performed by immunohistochemistry. MCC is tipically positive for CK20 and most low-molecolar-weight cytocheratin markers (16). In patients with asymptomatic primary MCC, sentinel lymph node biopsy (SLNB) is the most sensitive method to diagnose nodal metastasis considering that about 30% of clinical negative nodes are involved (17). Positron emission tomography- computerized tomography (PET-CT) scan is preferred when distant metastasis is suspected but CT scan and MRI may be used too (16, 18). Surgery with a 1-2 cm free margin is the main treatment for this neoplasia followed by radiotherapy (18, 19). According to the National Comprehensive Cancer Network (NCCN) the efficacy of chemotherapy or molecular-target-therapy is questionable (18).

The most common sites of metastases from MCC are distant lymph nodes (27-60%), distant skin (9-30%), lung (10-23%), central nervous system (18.4%) and bone (15.2%) (6). Gastrointestinal metastases are uncommon and small bowel is the most common location followed by stomach (8). In our case a jejunal MCC metastasis was diagnosed af-

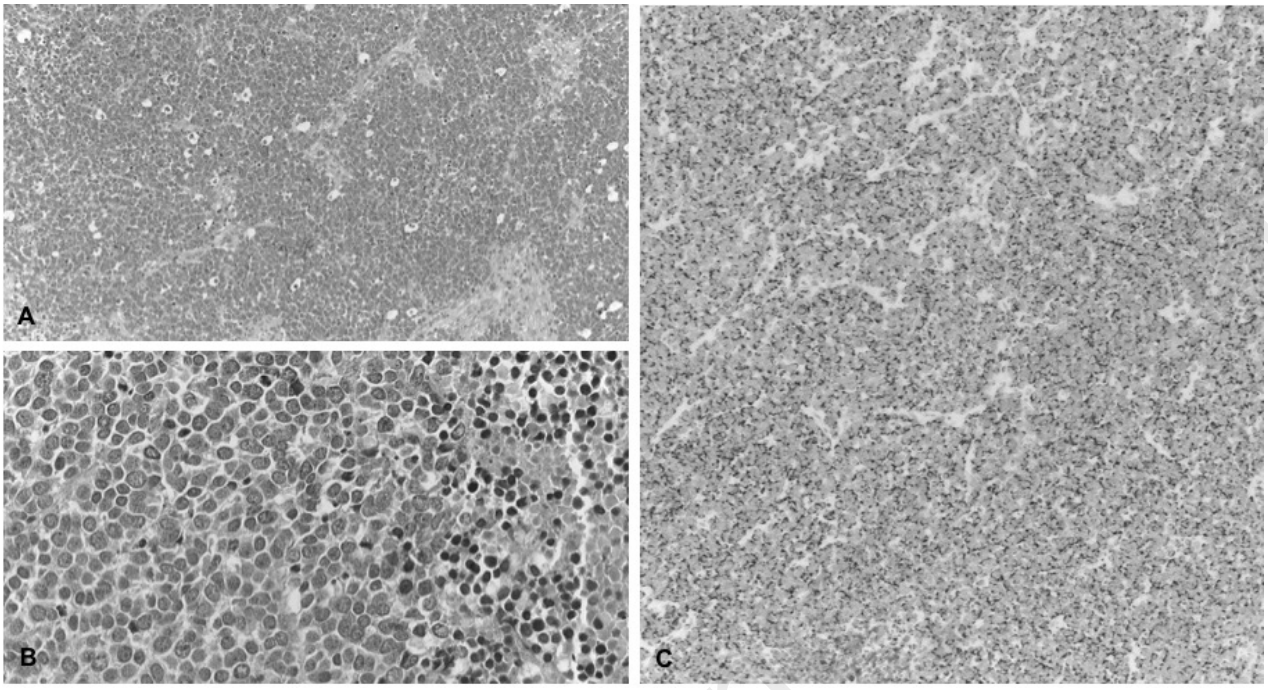

Figure 2 - Histological images. A) 20x Hematoxylin and eosin (H&E) stained image showing widespread proliferation of medium-size monomorphic tumor cells. B) 40x Hematoxylin and eosin (H&E) stained image of nuclei with finely granular chromatin, small nucleoli, numerous mitotic figures and necrosis areas. C) CK-20 positive tumor cells showing a characteristic perinuclear dot-like staining pattern. These findings were consistent with Merkel cell carcinoma metastasis.

ter histological report. Clinical presentation and preoperatory imaging were suggestive for a primary small bowel neoplasia. Thus, a radical small bowel resection was performed. Follow-up is very important due to high recurrence rate: the incidence of local recurrence is 25-30%, regional disease is 52-59% and distant metastatic disease is 34-36% of all cases of MCC (20).

In conclusion, MCC represents a very aggressive skin neoplasm with a high rate of nodes dissemination and distant metastases. Thus, radical primary surgical excision, sentinel node examination and adjuvant radiotherapy are needed. Small bowel metastases are rarely reported and related to dismal prognosis. This rare entity should be kept in mind when gastrointestinal symptoms or findings occurred in patients with MCC history. Given the rarity of this tumor, no consensus exists about optimal management and a long-term follow-up is then suggested.

References

1. Becker JC. Merkel cell carcinoma. Ann Oncol. 2010;21 Suppl 7:vii81-5.
2. Bachmann J, Kleeff J, Bergmann F, Shrikhande SV, Hartschuh W, Buchler MW, et al. Pancreatic metastasis of Merkel cell carcinoma and concomitant insulinoma: case report and literature review. World J Surg Oncol. 2005;3:58.
3. Shuda M, Kwun HJ, Feng H, Chang Y, Moore PS. Human Merkel cell polyomavirus small T antigen is an oncoprotein targeting the 4E-BP1 translation regulator. J Clin Invest. 2011;121(9):3623-34.
4. Saini AT, Miles BA. Merkel cell carcinoma of the head and neck: pathogenesis, current and emerging treatment options. Onco Targets Ther. 2015;8:2157-67.
5. Marcoval J, Ferreres JR, Penin RM, Perez D, Vinals JM. Merkel cell carcinoma: differences between sun-exposed and non-sun-exposed variants-a clinical analysis of 36 cases. Dermatology. 2014;229(3):205-9.
6. Medina-Franco H, Urist MM, Fiveash J, Heslin MJ, Bland KI, Beenken SW. Multimodality treatment of Merkel cell carcinoma: case series and literature review of 1024 cases. Ann Surg Oncol. 2001;8(3):204-8.
7. Heath M, Jaimes N, Lemos B, Mostaghimi A, Wang LC, Penas PF, et al. Clinical characteristics of Merkel cell carcinoma at diagnosis in 195 patients: the AEIOU features. J Am Acad Dermatol. 2008;58(3):375-81.
8. Syal NG, Dang S, Rose J, Chen C, Aduli F. Gastric metastasis of merkel cell cancer - uncommon complication of a rare neoplasm. J Ark Med Soc. 2012;109(7):134-6.
9. Idowu MO, Contos M, Gill S, Powers C. Merkel cell carcinoma: a report of gastrointestinal metastasis and review of the

literature. Arch Pathol Lab Med. 2003;127(3):367-9.
10. Cheung M, Lee H, Purkayastha S, Goldin R, Ziprin P. Ileocaecal recurrence of Merkel cell carcinoma of the skin: a case report. J Med Case Rep. 2010;4:43.
11. Matkowskyj KA, Hosseini A, Linn JG, Yang GY, Kuzel TM, Wayne JD. Merkel cell carcinoma metastatic to the small bowel mesentery. Rare Tumors. 2011;3(1):e2.
12. Li M, Liu C. Cytokeratin 20 confirms merkel cell metastasis to stomach. Appl Immunohistochem Mol Morphol. 2004;12 (4):346-9.
13. Parikh MP, Samo S, Ganipisetti V, Krishnan S, Dhandha M, Yungbluth M, et al. Gastric metastasis of Merkel cell carcinoma, a rare cause of gastrointestinal bleeding: case report and review of the literature. J Gastrointest Oncol. 2014;5(4):E68-72.
14. Shalhub S, Clarke L, Morgan MB. Metastatic Merkel cell carcinoma masquerading as colon cancer. Gastrointest Endosc. 2004;60(5):856-8.
15. Toker C. Trabecular carcinoma of the skin. Arch Dermatol. 1972;105(1):107-10.
16. Stein JM, Hrabovsky S, Schuller DE, Siegle RJ. Mohs micrographic surgery and the otolaryngologist. Am J Otolaryngol. 2004;25(6):385-93.
17. Gupta SG, Wang LC, Penas PF, Gellenthin M, Lee SJ, Nghiem P. Sentinel lymph node biopsy for evaluation and treatment of patients with Merkel cell carcinoma: The Dana-Farber experience and meta-analysis of the literature. Arch Dermatol. 2006;142(6):685-90.
18. Bichakjian CK, Olencki T, Aasi SZ, Alam M, Andersen JS, Blitzblau R, et al. Merkel Cell Carcinoma, Version 1.2018, NCCN Clinical Practice Guidelines in Oncology. J Natl Compr Canc Netw. 2018;16(6):742-74.
19. Poulsen M, Harvey J. Is there a diminishing role for surgery for Merkel cell carcinoma of the skin? a review of current management. ANZ J Surg. 2002;72(2):142-6.
20. Allen PJ, Bowne WB, Jaques DP, Brennan MF, Busam K, Coit DG. Merkel cell carcinoma: prognosis and treatment of patients from a single institution. J Clin Oncol. 2005;23 (10):2300-9.