ANNALI ITALIANI DI CHIRURGIA

Digital Edition
e-publish on-line
ISSN 2239-253X
Direttore Nicola Picardi

Multiple enlarged lymph nodes in axilla

A case report of Merkel Cell Carcinoma

Ann Ital Chir, Digital Edition 2017, 6
pii: S2239253X17026378 - Epub, Jan. 16
free reading: www.annitalchir.com

Muatasim Noorelahi*, Rana Mahmood**

**Intern, College of Medicine, Taibah University, Madimah, Saudi Arabia*
***Radiation Oncologist Consultant, Oncolology Centre, King Faisal Specialist Hospital & Research Center, Riyadh, Saudi Arabia*

Multiple enlarged lymph nodes in axilla: A case report of Merkel Cell Carcinoma

We present the case of a 59-year-old man with a growing mass in his left axillary area. A biopsy and immunostainings demonstrated neuroendocrine carcinoma, which is Merkel cell carcinoma (MCC). The disease is characterised by neurosecretory granules in tumor cells. MCC is a rare entity. The disease is predominantly seen in the inguinal region or the axilla and typically found incidentally. It presents clinically as multiple lymph nodes enlargement. Yet controversy exists regarding treatment modality of MCC. We report the first case of MCC in (Left) Axilla where there were two cases reported previously in (Right) Axilla.

KEY WORDS: Lymph nodes enlargement, Merkel cell carcinoma, Radiotherapy

Introduction

Merkel cell carcinoma (MCC) is a rare aggressive tumor; arising from mechanoreceptors of the skin, which is cutaneous neuroendocrine carcinoma [1]. In majority, MCC inclines to appear on sun-exposed area: head, neck, and extremities of elderly [2] MCC is highly associated with propensity to local relapse, loco-regional nodal involvement and distance metastasis, which has the potential to be fatal and poor prognosis [3].

Controversy exists regarding therapeutic approach and optimal management of MCC, with mainstay of management with surgery and radiation, and no evidence-based effective chemotherapy treatment has been made available to date [4]. We hereby describe a patient with MCC of the left axilla.

Pervenuto in Redazione Settembre 2016. Accettato per la pubblicazione Novembre 2016
Correspondence to: Muatasim Noorelahi, MBBS, Al-Hijra str, Medina, Saudi Arabia (e-mail: motasimelahi@aol.com)

Case Report

A 59-year-old male ex-smoker patient, a known case of hypertension, dyslipidemia, ischemic heart disease, diabetes and post cardiac catheterization, noticed left axillary (LA) mass increasing in size for 1 year. No history of weight loss or other symptoms. Systemic review was unremarkable. The patient had mild penicillin allergy but no reactions were documented. Written consent for publication was obtained from the patient. According to physical examination, he was in generally good condition and vitally stable. There was a very large, fixed, non-tender mass in the LA. There was no obvious primary malignancy anywhere on examination. The Patient had a LA lymph node (LN) biopsy and examined tissue revealed presence of sheets of small blue cells exhibiting hyperchromatic nucleus with nuclear molding and no nucleoli. Immunostainings were performed and tumor cells were positive for Chromogranin, Synaptophysin and Pancytokeratin and negative for TTF1. These results were consistent with neuroendocrine carcinoma. Workup including Gallium-68 Positron Emission Tomography scan (Ga68 PET/CT) revealed that there were multiple enlarged LNs in the left subpectoral and axillary regions

with corresponding increased tracer activity (SUV max 8.2 in the subpectoral region and 9.4 at the axillary region). No other significant tracer-avid lymphadenopathy noted elsewhere in the study (Fig. 1).

The plan was initially to do an axillary dissection and an intraoperative radiation (IORT). The patient was taken to the operative room and the LA dissection removed very large tumor. However at the apex of the axilla the tumor was infiltrating into the brachial plexus. In fact, the axillary vein (AV) was a double vein, fortunately only one part of the vein was completely invaded. We removed that part of AV with the tumour, however we had to leave a very small remnant of the tumor that was attached to the neurovascular bundle rather than risking neurovascular damage. In consultation with radiation oncologist in the operating room, a decision was made as not to deliver IORT and consider external radiation for the whole tumour bed and residual gross disease in post-operative setting. After surgery, patient had gallium scan which confirmed that this tumor takes up gallium isotope. The tumor was confirmed as a MCC with immuno-positive CK, CAM, synaptophysin and chromogranin, which was patchy. The Ki-67 proliferative index was around 80-90%. Pathology specimen revealed multiple nodes, largest 9 cm with extra nodal disease. Unsurprisingly, surgical margin was positive while gross disease was left during surgery.

Case was discussed in tumour board and in view of gallium uptake by the residual tumour; decision was made to start with radionuclide receptor therapy. He was given the 1st dose of 147 mCi of 177 lutetium-DOTATOC treatment and this was confirmed with the scan postoperatively with good therapy uptake (Fig. 2). Then, he was given another dose of 140 mCi of lutetium 177 DOTATOC IV and again showed good uptake in the LA and no other uptake anywhere else. Ga68 PET/CT post Lu177 therapy showed progressive disease (Fig. 3). After further multidisciplinary discussion, we offered the patient a radical course of Chemoradiotherapy. Patient was started on 1st cycle of Intravenous Etoposide 100 mg/m^2 and Cisplatin 80 mg/m^2 and radiotherapy was planned as concurrent treatment with 2nd and 3rd cycle.

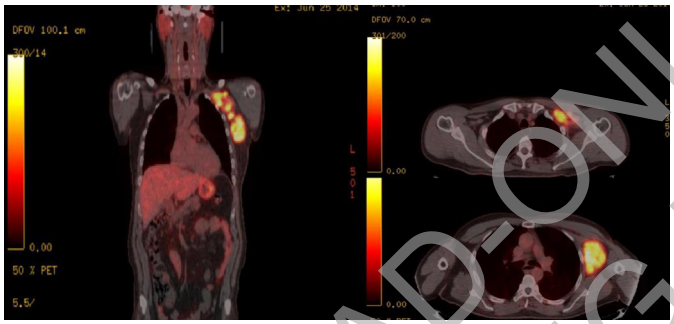

Fig. 1: Ga68 PET/CT scan – Pre-operative Baseline scan.

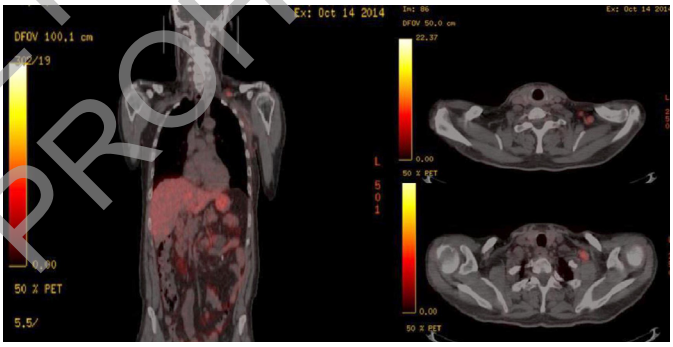

Fig. 3: Ga68 PET/CT scan – Post Lu177 progressive disease.

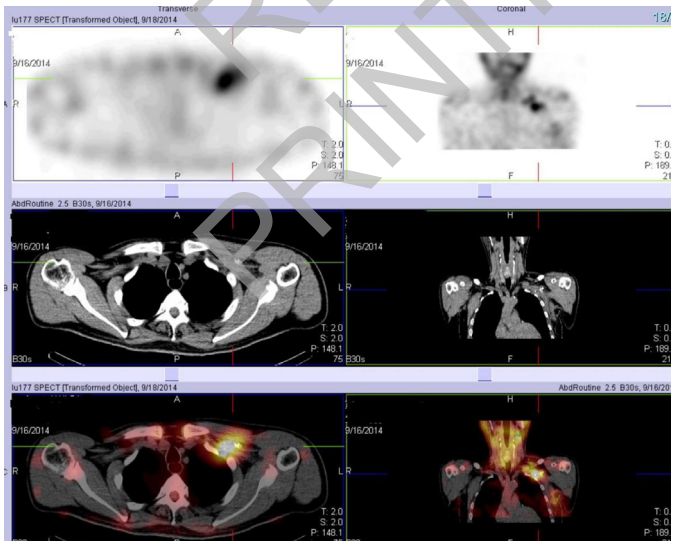

Fig. 2: Lu177 SPECT therapy scan showing uptake in residual disease.

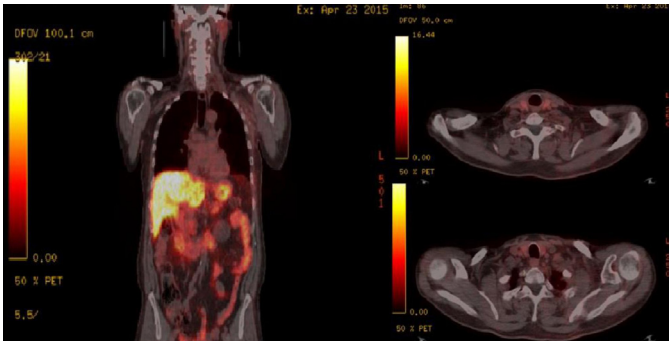

Fig. 4: Ga68 PET/CT scan – Post Chemoradiotherapy showing complete response.

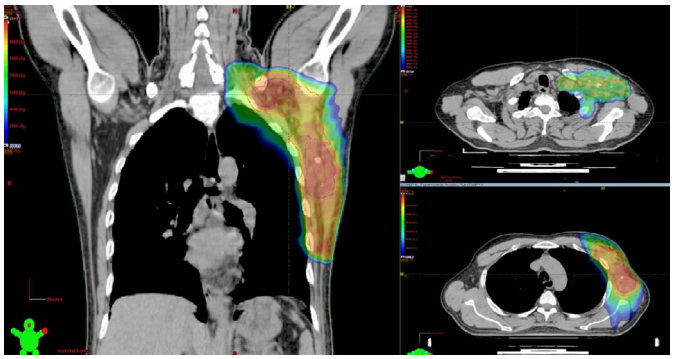

Fig. 5: Planning CT scan showing dose colour wash of IGRT plan with Simultaneous Boost: (Red) 66Gy for gross disease and (Yellow) 60Gy tumour bed.

Total dose of 60Gy to whole tumour bed and 66Gy in 30 fractions was delivered using Volumetric Arc Therapy (VMAT). Dose colour wash displaying dose coverage of tumour bed and escalated dose at residual disease is shown in Fig. 5.
After 6 months follow up, Ga68 Whole Body PET/CT was done and revealed that previously seen LNs in the LA/subpectoral and left supraclavicular region are no longer visualized (Fig. 4).

Discussions

In 1972, MCC was discovered by Toker and described as Trabecular carcinoma [5]. The aggregation of diagnostic imaging and pathology demonstrated electron dense neurosecretory granules in tumor cells and it was classified as tumor of neuroendocrine origin [6].
Incidence of MCC is rising due to increase exposure to the risk factors: Ultraviolet (UA) exposure and increasing in number of people living with immunosuppressive disease [7]. However, incidental finding of a LN enlargement is the most common clinical presentation of MCC, usually at the inguinal region or the axilla and it is usually discovered after excisional biopsy and pathology examination. Recently the role of [8] F-FDG PET or PET/CT in patients with MCC has been successfully reported in depicting the sites of LN disease due to its high reported specificity (98%) and sensitivity (90)[8].

Conclusion

Up to today the main treatment modality of MCC is undefined, however options include regional LN surgical dissection, radiotherapy and/or Chemotherapy with etoposide and cisplatin or carboplatin. Although, the rarity of MCC tumors makes almost impossible to extract good quality of evidence in the form of randomized control trials addressing this issue [9].

Riassunto

Si presenta il caso di un uomo di 59 anni portatore di una massa in accrescimento nella ascella sinistra. Lo studio bioptico ha dimostrato trattarsi di un carcinoma neuroendocrino, e precisamente un carcinoma a cellule di Merkel (MCC).
Questa patologia è caratterizzata da granuli neurosecretori nelle cellule tumorali, e si tratta di una entità rara e localizzata prevalentemente nelle ragioni inguinali e ascellari ed in genere di scoperta casuale. Si presenta come ingrandimento di linfonodi multipli.
Esistono controversie tutt'ora circa le modalità del trattamento d intraprebdere.
Questo è il primo caso di una localizzazione di MCC nell'ascella sinistra, mentre vi sono in letteratura due casi localizzati nell'ascella destra.

References

1. Goodwin CR, Mehta AI, Adogwa O, Sarabia-Estrada R, Sciubba DM: *Merkel Cell Spinal Metastasis: Management in the Setting of a Poor Prognosis.* Glob Spine J, 2015; 5(4):e39-43. Doi: 10.1055/s-0034-1398488.

2. Kritikos N, Priftakis D, Stavrinides S, Kleanthous S, Sarafianou E: *Nuclear medicine techniques in Merkel cell carcinoma: A case report and review of the literature*. Oncol Lett, 2015; 10(3):1610-6. Doi: 10.3892/ol.2015.3377.

3. Gorayski P, Tripcony L, Poulsen M: *Chemotherapy compliance in high-risk Merkel cell carcinoma treated with chemoradiotherapy*. Australas J Dermatol, 2015; Doi: 10.1111/ajd.12419.

4. Prieto I, de la Fuente TP, Medina S, Castelo B, Sobrino B, Fortes JR, et a.: *Merkel cell carcinoma: An algorithm for multidisciplinary management and decision-making.* Crit Rev Oncol Hematol, 2015; Doi: 10.1016/j.critrevonc.2015.10.008.

5. Toker C: *Trabecular carcinoma of the skin.* Arch Dermatol 1972; 105(1):107-10.

6. 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:E68-72.

7. Holdsworth R, Liew S: *Electrochemotherapy for treatment of Merkel cell carcinoma: A case report.* JPRAS Open, 2015; 6:49-52. Doi: 10.1016/j.jpra.2015.06.001.

8. Treglia G, Kakhki VRD, Giovanella L, Sadeghi R: *Diagnostic performance of fluorine-18-fluorodeoxyglucose positron emission tomography in patients with Merkel cell carcinoma: a systematic review and meta-analysis.* Am J Clin Dermatol, 2013; 14(6):437-47. Doi: 10.1007/s40257-013-0040-x.

9. Kontis E, Vezakis A, Pantiora E, Stasinopoulou S, Polydorou A, Voros D, et al.: *Merkel cell carcinoma of unknown primary site; case presentation and review of the literature.* Ann Med Surg, 2015; 4(4):434-37. Doi: 10.1016/j.amsu.2015.10.013.