



Case Report

Neuroendocrine tumor of the inguinal node: A very rare presentation

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ABSTRACT

Neuroendocrine tumors are a broad family of tumors arising most commonly in the gastrointestinal tract and the bronchus pulmonary tree. The other common sounds are the parathyroid, pituitary and adrenal gland. Inguinal node as a primary presentation of a neuroendocrine tumor is an extremely rare presentation. We present the case of a 43-year-old-male who presented with the complaints of an inguinal node swelling without any other symptoms and on further evaluation was diagnosed to have a non-metastatic neuroendocrine tumor of the inguinal node. He was treated with a combination of chemotherapy and surgery and is presently awaiting completion chemotherapy.

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1. Introduction

Neuroendocrine tumors are a heterogeneous group of tumors, which can arise from the cells along the diffuse endocrine system. The majority of neuroendocrine tumors appear to be sporadic, but they may also arise in the context of inherited genetic syndromes, including multiple endocrine neoplasia types 1 and 2.¹ The common sites of these tumors are gastrointestinal tract and bronchopulmonary tree. Some rare sites include ovary and breast. However inguinal node being the primary presentation has been described very rarely in history.^{2–4} This article presents the case of a rare presentation of a neuroendocrine tumor presenting as an inguinal node swelling.

2. Case presentation

42-year-old male presented with history of swelling in the left inguinal region for a period of three months. The swelling was painless and progressive in nature which when started was the size of a marble but progressed to the present size. Local examination showed an inguinal swelling measuring 6 cm × 6 cm × 5 cm

swelling, which was hard, non tender and fixed. The overlying skin was stretched but non erythematous. He was completely asymptomatic without any clinical symptoms of the carcinoid syndrome such as flush, diarrhea and wheezing or other hormonal disturbances. There were no signs of localized infection within the pelvic or perianal region, abdomen and lower extremities.

A trucut biopsy from the inguinal node revealed obliterated sub capsular sinus, with lymph nodal architecture effaced and replaced by neoplastic cells. The neoplastic cells were small to moderate in size, round to oval with moderate granular cytoplasm, round to oval nuclei with stippled chromatin (Fig. 1). Occasional mitosis was seen with no necrosis noted. Immunohistochemistry was positive for CK, CD 56, synaptophysin and chromogranin. It was negative for CD 3, CD 20, LCA, SMA, EMA and desmin (Fig. 2). The final impression was of a poorly differentiated neuroendocrine carcinoma.

As a primary neuroendocrine carcinoma of the lymph nodes is extremely uncommon, this histological result was suspected to be a lymph node metastasis from a hitherto unrecognized primary tumor. Dermatological examinations showed no pathological findings, especially any evidence of Merkel cell carcinoma. There was also no evidence of lymphoma or melanoma. Laboratory investigations including 5-hydroxyindole acetic acid, chromogranin A, and neuron-specific enolase showed normal results.

He underwent a colonoscopy, which did not reveal any positive findings. He underwent Whole body Positron Emission Tomography with non contrast Computerized Tomography which revealed a 5.6 cm × 6 cm × 5 cm lymph nodal mass in the left inguinal region with increased metabolic activity and no other FDG active lesion

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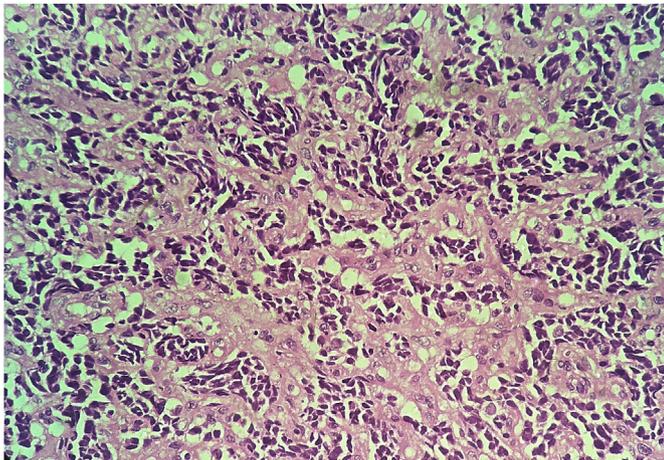


Fig. 1. Tru-cut biopsy showing effacement of lymph nodal architecture & replacement by small to medium neoplastic cells with round nuclei with stippled chromatin and occasional mitoses.

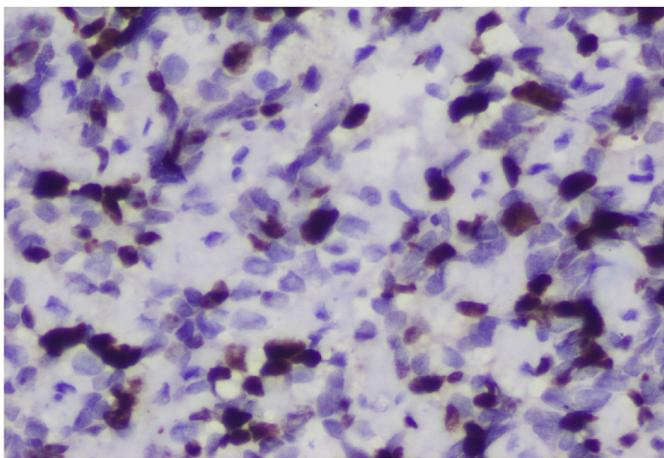


Fig. 2. Photomicrograph of immunohistochemistry slide showing a MIB-1 index of 40–45%.

(Fig. 3). He was further assessed with Gallium –68-DOTANOC whole body scan. Increased radiotracer uptake was noted in the left inguinal region with no somatostatin receptor avid occult primary detected (Fig. 4).

With the diagnosis of primary neuroendocrine tumor of the inguinal node, the patient was treated with 03 cycles of cisplatin and etoposide and taken up for clinical reassessment. The inguinal node decreased in size to around 4.5 cm × 4 cm × 5 cm and was mobile without fixation of overlying skin. He underwent wide local excision of the node.

The histopathology of the excised node showed intermediate sized cells with hyperchromatic nuclei, scant to moderate amount of eosinophilic cytoplasm and inconspicuous nucleoli. Mitosis is 5–7/High Power field. Immunohistochemistry showed cells positive for Cyto keratin, CD 56 AND NEGATIVE FOR CK 7, CK 20,S 100, synaptophysin and chromogranin. MIB labeling index is 25–27%. The final impression was of histomorphical and immunohistochemical features suggestive of poorly differentiated neuroendocrine tumor of inguinal node (post chemotherapy).

The patient was further treated with 3 more cycles of cisplatin and etoposide after surgery. He has remained on regular follow up since then and 06 months post completion of therapy, he remains disease free. DOTANOC and 18-FDG PET CT scan done at six months have shown no abnormal lesion in the body.

3. Discussion

Neuroendocrine tumors comprise of a family of tumors with preponderance in the gastrointestinal region. Rectum is the most common gastrointestinal site involved.^{5,6} According to a SEER database analysis, a primary tumor site could not be found in as many as (13%) neuroendocrine tumors.⁷ Histologically most of these tumors are aggressive and poorly differentiated mimicking small cell carcinoma.⁸ The initial evaluation of a patient with neuroendocrine tumors of unknown primary includes patient family history, clinical manifestations, laboratory studies, imaging studies, and immunohistochemical studies. Special investigations like OctreoScan and 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) scan may also be helpful in localizing certain neuroendocrine tumors.⁹

Inguinal node as the primary site in a non-metastatic condition is rarely described in literature. The presence of malignant cells in inguinal lymph nodes might be due to variable causes. One theory is the result of a metastatic spread from an occult or a regressed primary carcinoma. However even with an unknown primary the involvement of an inguinal node is not common.¹⁰ The patient was worked up to look for an unknown primary, but both imaging and endoscopy evaluation did not reveal anything.

Another explanation could be cutaneous manifestation of a malignant neuroendocrine tumor of the right leg with spread of



Fig. 3. Whole Body 18-FDG PET CT showing an isolated metabolically active lesion, 5.6 × 6 × 5 cm in size in the Left inguinal region.

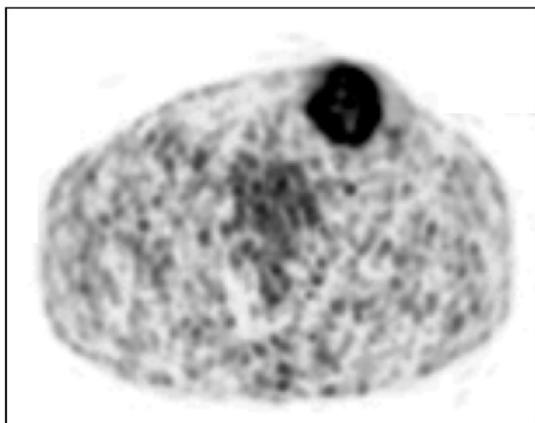


Fig. 4. Gallium-68-DOTANOC scan showing presence of somatostatin receptors over the lesion in the Left inguinal region.

metastatic neuroendocrine cells within a lymph node. Inguinal lymph nodes drain lymph from the lower extremity and the pelvic region. However, in spite of thorough investigations there was no evidence of a tumor within the anorectal region or lower abdomen. Neuroendocrine tumors can originate from neuroendocrine cells that reside within the basal layer of the epidermis (Merkel cell carcinoma).¹¹

Merkel cell carcinoma (MCC) is a rare but extremely aggressive skin cancer. Toker first described it¹² in 1972 as trabecular carcinoma of the skin. It is an aggressive tumor, which recurs locally and frequently disseminates to regional lymph nodes and distant organs, primarily to the liver and bones. These tumors typically arise from sun-exposed skin areas like the face and neck and in immunocompromised patients. However extensive dermatological examination did not reveal any positive findings.

Two possible explanations can be considered for what occurred in this patient. One is that the primary nodal disease is a distinct entity, which shows a less aggressive biological behavior as compared to a metastatic one. Another theory to explain the presence of the neuroendocrine carcinomas within lymph nodes is to postulate the malignant transformation of preexisting intranodal epithelial nests. The treatment of a poorly differentiated neuroendocrine resectable cancer remains a combination of surgery and small cell lung cancer chemotherapy regimens such as cisplatin, etoposide or carboplatin. For unresectable locoregional disease, radiotherapy in combination with chemotherapy (again, with a

small cell lung cancer regimen) is recommended. In a metastatic setting, chemotherapy alone (with a small cell lung cancer regimen) is recommended. Octreotide therapy can be considered for hormone-secreting tumors that are unresectable or metastatic.

In this case the clinical behavior of the tumor and response to treatment is yet to be assessed. Until then a close follow-up with local examination and imaging is planned. In addition to standard procedures, recent data indicate that positron emission tomography may present a valuable tool for the detection of neuroendocrine tumors or tumor recurrence.^{13,14}

In conclusion, this is a rare presentation of a neuroendocrine tumor. With the gamut of postulations explained above, a regressed primary or a malignant nodal transformation can account for this presentation. Further follow up and evaluation will give us a better picture of the behavior of an inguinal node neuroendocrine tumor.

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