Neuroendocrine Tumors (NET’s) Unusual Clinical Presentations

R Parikh, P Desai, S Arneja
Breach Candy Hospital, Mumbai, India

Introduction

Neuroendocrine tumors are a heterogenous group of malignant tumors. Earlier thought to be rare, forming 1% of all neoplasms, the current data shows that NET form 10% of all neoplasms. They vary in localization, clinical presentation, tumor biology and prognosis. The tumors can occur at any site in the neuro-endocrine system. Seventy five percent of NET’s arise from gastro-intestinal tract i.e. the small intestine, appendix, stomach and rarely the colo-rectum. The lung and bronchus are the second most common sites forming 20% of all NET’s. The pancreato-biliary tree, thymus and ovary are the other known sites. Some tumors present with clinical syndromes related to peptides and amine production, or the carcinoid syndromes related to serotonin and tachykinin production from small intestinal NET’s (carcinoids).

Case Study: Case 1

A 62 year old male presented with a swelling and pain in the right scapular region. MRI and PET-CT scan showed a localized lesion of the scapular spine without any dissemination. The entire lesion was widely resected. Final histopathology revealed a primary NET. Patient is followed up over 4 years and has remained disease free clinically and on imaging

Case Study: Case 2

A 22 year old female was diagnosed as a small cell lung carcinoma and treated with chemotherapy and radiation at another institution. Three years later, she presented with a large abdomino pelvic mass. MRI and PET-CT scan showed a large lobulated mass arising from the pelvis. The lungs were normal on imaging. A core biopsy reported a NET. Chromogranin A levels were elevated. She had no symptoms to suggest a carcinoid syndrome. Exploratory laprotomy revealed huge bilateral ovarian masses. A total hysterectomy with bilateral salpingo-oophrectomy was performed. The final histopathology showed a high grade NET consistent with metastasis from a small cell neuroendocrine carcinoma of the lung. Post-operative chemotherapy (Carboptatin, Etoposide and Capcetabine) were administered. She remains disease free 3 years later

Conclusions

1. Primary skeletal NET is extremely rare, literature has reported sacrum and pre-sacral NET’s; however, primary scapular spine NET remains unreported.
2. Metastasis from a primary NET’s are known to occur in the liver, lung and bones; however, metastasis to the ovary occur in only 2%. Metastasis to the ovary usually occur from primary carcinoids in the ileum, pancreas, appendix; or jejunum. Metastasis to the ovary from a primary lung NET is extremely rare as in this case. Small cell lung carcinoma (SCLC) is considered similar to NET of the lung due to common morphology, immuno-histochemical and molecular features. SCLC is, in fact, a large cell neuroendocrine carcinoma as reported in this patient.