PP-18

Grave Back Pain: A Case of Somatostatin Receptor Negative Metastatic Pancreatic Neuroendocrine Neoplasm

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INTRODUCTION
Somatostatin receptor (SSTR) status is an important prognostic marker in gastroenteropancreatic neuroendocrine neoplasms (NEN). Most NEN's are SSTR-positive with only approximately 7.6% being receptor negative. Among these SSTR-negative neoplasms, 75% are in the pancreas.

METHODOLOGY
We describe a 60-year-old Malay male who presented with progressive lower back pain for six months, associated with constitutional symptoms and a palpable left supraclavicular lymph node.

RESULTS
Chest radiography revealed a mediastinal mass. Computed tomography (CT) scan showed metastatic disease with generalised lymphadenopathies, a subcutaneous right anterior chest wall nodule and multiple lesions in the lung, liver and tail of the pancreas. Magnetic resonance imaging revealed extensive spinal metastases. CT-guided core biopsy of the mediastinal lymph node was suggestive of NEN with a low proliferative index, Ki-67 (<5%). An endoscopic ultrasound-guided biopsy of the pancreatic lesion confirmed primary tumour. Clinical and biochemical assessment concluded the NEN to be non-functional. We proceeded with a Gallium-68 DOTATOC PET-CT scan, which showed absence of SSTR avid disease. A multidisciplinary meeting conceded the disease to be unresectable and chemotherapy with spinal radiation was concurred as definitive management. Systemic therapy options were limited by the SSTR-negative status, rendering him unsuitable for SSTR-dependant therapies. He deteriorated with a rapidly enlarging anterior chest wall mass. Unfortunately, he was unfit for chemotherapy due to recurrent infections and succumbed within two months of diagnosis.

CONCLUSION
In our patient, the low Ki-67 index reported from the biopsied specimen didn't correlate with his rapid disease progression most probably due to intratumoural heterogeneity of Ki-67 expression in NEN. SSTR-negative status carries a poor prognosis. It is associated with high grade tumours with limited treatment options. More research is required to explore the best therapeutic strategy in this uncommon setting. The overall prognosis is poor in view of the negative SSTR status, bulky metastases and unresectable primary.

PP-19

Delay in Diagnosing Aldosterone-producing Adenoma: Can We Do Better?

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INTRODUCTION
Incidence of primary hyperaldosteronism is rising due to better detection and awareness among physicians. However, there is delay in patients receiving appropriate care. In these 2 case-series of aldosterone-producing adenoma, we summarized the key events until tumour removal.

METHODOLOGY
All case notes of new patients attending endocrine clinic Hospital Tengku Ampuan Afzan from 1st January 2017 till 1st May 2019 were screened. Cases fulfilling the diagnosis of primary hyperaldosteronism were included. Key events were reviewed and summarized.

RESULTS
Case 1 is a lady with hypertension for 7 years and hypokalaemia for 3 years. ARR done in December 2016 was positive. She was seen in endocrine clinic about 2 months later, saline suppression test (SST) was done after 2 weeks, but CT of the adrenals was done 3 months after SST. She was referred 3 months later to endocrine Hospital Putrajaya where adrenal vein sampling (AVS) was done. Results of AVS was made available after 1-month. She was referred to an endocrine surgeon after 6 weeks and operated 2 months later. Total time from ARR to table was 13 months. Case 2 is another lady with hypertension and hypokalaemia for 15 years. ARR was done in April 2017. She was only referred to endocrine clinic after 3 months. SST was done 6 weeks later, but the results were only reviewed after 3 months. CT of the adrenals was done after 3-month time. She was referred to endocrine Hospital Putrajaya for AVS which was done after 2 weeks. Finally, she was operated 6 months after AVS. Total time from ARR to table was 18 months.

CONCLUSION
There is delay from screening until definitive treatment. The process can be improved if there is proper diagnostic workflow for similar cases. Proper result-tracing and faster scans are desired. With improved diagnosis speed, hopefully there will be faster action to benefit the patients.