The Clinicopathologic Characteristics of Primary Presacral Neuroendocrine Tumor: A Single Center Experience

Guoqing Yang¹; Run Yu²; Nissen Nicholas²; Andrew Hendifar²

¹Chinese PLA General Hospital; ²Cedars-Sinai Medical Center

Background: Presacral carcinoid tumors are rare entities that are found at the presacral space and have been characterized as both benign or similar to hind-gut tumors. We report our experience in the diagnosis, management, and outcomes of primary presacral neuroendocrine tumors (NETs).

Methods: This was an IRB approved retrospective review of medical records and surgical pathology specimens of patients (pts) with a diagnosis of neuroendocrine tumor at Cedars-Sinai Medical Center between Jan 2000 and Jan 2016.

Results: Ten pts were identified. The median age at presentation was 38 years (20 - 77 years), and 8 female patients. One patient presented with carcinoid-like symptoms. Two presented without symptoms and were diagnosed incidentally. Seven presented with symptoms related to mass effect. The median size of the tumor was 7.0 cm (3 - 12cm). Of the 10 cases, 2 were associated with a presacral teratoma. On pathologic review, 6 were grade 1/2 and well differentiated and 2 were grade 3 and poorly-differentiated. 5 cases were metastatic on presentation with liver, lung or skeletal metastasis. CT, MRI, PET-CT and Scintigraphic imaging were used routinely for all cases. 7/8 cases were detectable using Octreoscan. 9 pts were treated with a somatostatin analog. 6 patients were treated surgically.

Conclusion: Presacral neuroendocrine tumors are clinically and histologically similar to GEPs and infrequently present with flushing or diarrhea. Octreoscan imaging and somatostatin analog therapy were frequently applied. Further biologic characterization of this rare subtype is needed.