

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

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Background

Neuroendocrine neoplasms (NENs) in the presacral region are rare and often represent direct extension from hindgut or genitourinary NETs or metastases.

The presacral space here is defined as a region bounded by the rectum anteriorly, the sacrum and coccyx posteriorly, the peritoneal reflection superiorly, and inferiorly by the pelvic floor muscles. The lateral margins are defined by the ureters and the iliac vessels. The presacral space contains tissues derived from all three germinal layers, which can lead to the development of a variety of tumor types.

However, primary NETS and NENs of the presacral space are rare and infrequently described. Herein, we report a case series of primary presacral NEN's from a single institution.

Methods

With approval from our Institutional Review Board, patient data were retrieved from a database of electronic medical records. The catchment period was from January 1, 2000 to April 1, 2016. Primary presacral NETs were defined as arising from presacral space and with no associated gastrointestinal, pancreatic, or urogenital lesion. Metastasis from other sites were excluded. The final determinations were made in multi-disciplinary conferenc. All cases were reviewed by a pathologist with experience in this disease.

Results

| Case # | Gender | Age at presentation/Dx | Co-morbidity | Symptoms | Tumor Size at Presentation | Metastasis at Presentation | Clinical diagnosis | Treatment regimen | Follow-up (years) | Outcome |
|--------|--------|------------------------|--------------------------------|--|----------------------------|--|--|---|-------------------|------------------------------------|
| 1 | F | 30/30 | None | Rectal pain | NA | no | Well differentiated NET, grade 1, associated with tailgut cyst | -Tumor cytoreduction -Radiation -Somatostatin analog | 10 | Alive, active disease |
| 2 | F | 20/33 | Hypothyroidism | Perirectal pain, flushing, diarrhea | NA | no | Well differentiated NET arising from teratoma | -Surgery -Somatostatin analog | 10 | Alive, disease free |
| 3 | F | 25/38 | Anterior sacral meningocele | Left-sided sciatica, pelvic pain, constipation | 10 cm | no | Well-differentiated NET, grade 1, associated with tailgut cyst | -Surgery -PRRT -Somatostatin Analog | 9 | Alive with active disease |
| 4 | M | 39/39 | Crohn's disease | Pelvic pain | 7 x 5 x 7 cm | Liver | Well differentiated NET, grade 2 | -Somatostatin analog -Chemotherapy -Radiation therapy | 2 | Alive with liver, bone, orbit mets |
| 5 | M | 71/71 | Hypothyroidism Hypertension | Right femoral vein DVT incidentally | 7 cm | Lymph nodes Bone | Well differentiated NET, grade 1 | -Surgery -CyberKnife therapy -Somatostatin Analog -Radioembolization | 6 | Alive with liver, bone mets |
| 6 | F | 36/40 | None | Chronic constipation | 11 x 9.6 x 9.5 cm | Liver | Well differentiated, grade 2 | -Surgery -Hepatic artery -Radioembolization -Somatostatin analog | 5 | Alive with liver and pancreas mets |
| 7 | F | 39/39 | Morbid obesity | None, incidental | 3.2 x 4.5 cm | Liver, lung, | Well differentiated NET, grade 2 | -Somatostatin analog -Interferon alpha | 2 | Alive with liver, lung, bone mets |
| 8 | F | 50/50 | None | Abdominal pain | 8.3 cm right pelvic tumor | Liver, pancreas, mesentery, lymph node | Well differentiated NET, grade 2 | - Chemotherapy | 1 | Alive with active disease |
| 9 | F | 45/45 | no | Hip pain | 3 cm | Lymph node | Poorly differentiated NET, grade 3 | -Chemotherapy -Radiation therapy | 1 | alive |
| 10 | F | 77/77 | Fallopian tube adenocarcinoma | Night sweats, weight loss, fatigue | 12 cm | Liver | Large Cell NET, grade 3 | -Chemotherapy -Somatostatin analog | 1 | expired |

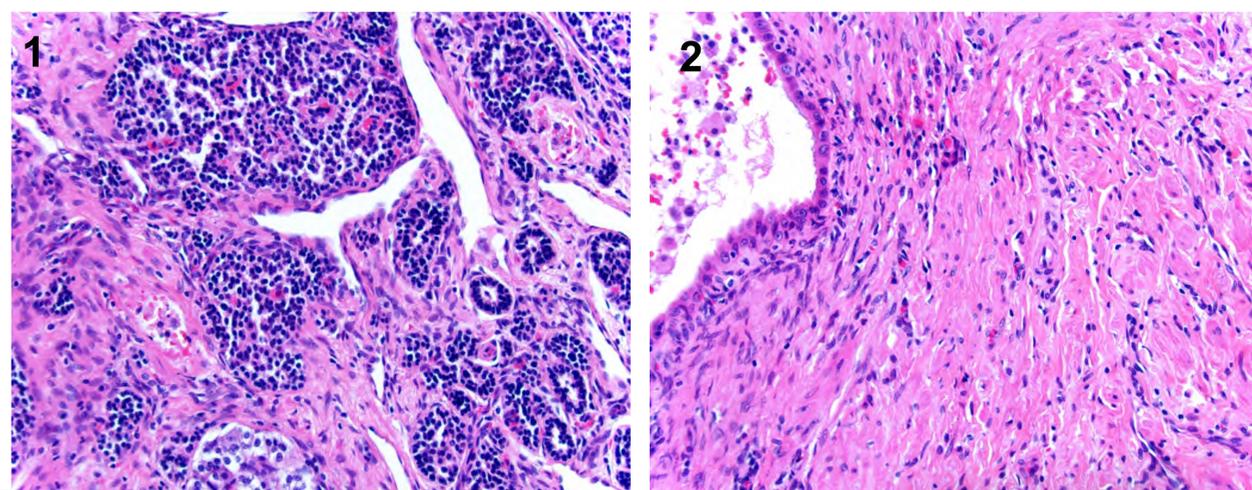


Figure 1. Case #2

Presacral well-differentiated NET:
1 - The tumor cells are uniform and are arranged in nests and pseudoglandular structures.
2 - Cystic spaces lined by ciliated columnar epithelium are present adjacent to the neuroendocrine tumor, suggestive of NET arising in association with a teratoma

