Acute Pancreatitis as an Initial Presentation of Small-Cell Carcinoma of Ovary, Hypercalcemic Type in a Pediatric Patient

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Introduction

Small cell carcinomas account for 1% of all ovarian cancers[1] and are hypercalcemic type (SCCOHT) or neuroendocrine. SCCOHT is the most common undifferentiated ovarian malignancy in females younger than 40 years[2].

We report the first case of a pediatric patient with SCCOHT presenting with presumed gallstone pancreatitis and hypercalcemia with elevated PTHrp. Malignancy should be considered in adolescent patients presenting with hypercalcemia and elevated PTHrp. In early stages of SCCOHT, surgical resection followed by multi-agent chemotherapy and radiation may improve survival rates.

Case

A 15-year-old female with obesity, iron deficiency anemia, and menometrorrhagia presented with 2 weeks of constipation, vomiting, and acute onset severe right upper quadrant abdominal pain.

Potassium (3.5-5.0 mmol/L)	2.9
BUN (7-20 mg/dl), Creatinine (0.7-1.3 mg/dl)	12/0.8
Calcium (2.1-2.6 mmol/L)	4.57
Ionized calcium (1-1.4 mmol/L)	2.31
Mg (0.7-1.1 mmol/L), Phos (0.8-1.5 mmol/L))	0.6/0.7
Lipase (10-70 U/L)	2550
AST (0-30 U/L)	20
ALT (0-37 U/L)	23
Alk Phos (37-173 U/L)	73
Total bili (0-1 mg/dl), direct bilirubin (0-0.4 mg/dl)	1.5/0.3
Albumin (3.8-5.2 g/dl)	3.7
Hemoglobin (mg/dl)	9.9
PTHrP	20
PT (9.4-12.5 seconds), INR (08-1.1)	21.1/1.8
CA-125 (0-35 U/ml)	125
CEA, FSH, LH, prolactin, DHEA, estradiol, testosterone, Ca19-9, anti-mullerian hormone, inhibin B, AFP, 17-hydroxyprogesterone	Normal

Imaging

US abdomen showed cholelithiasis with no cholecystitis.

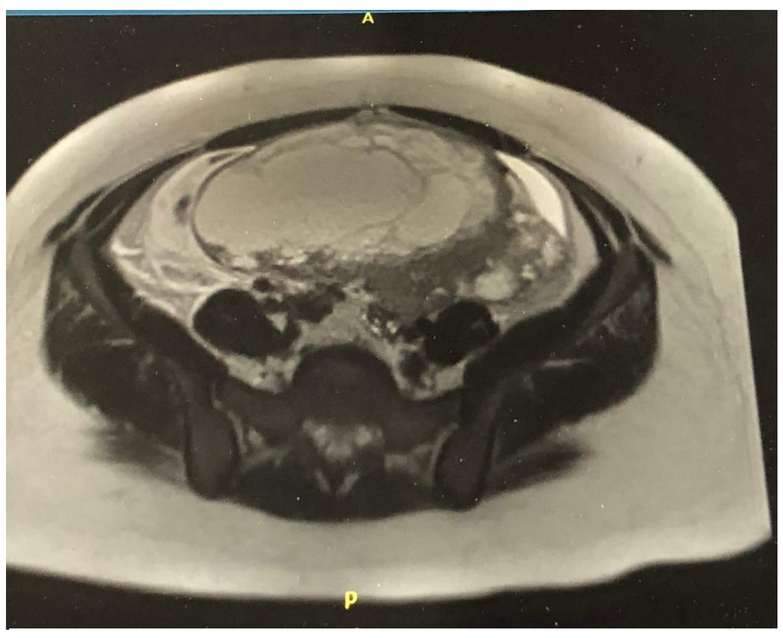

Figure 1 MRI pelvis right adnexal mass 10.0 (AP) x 18.3 (TV) x 19.4 (CC) cm with internal septations, complex cystic changes, and peripheral enhancing nodularity.

Treatment/Interventions

Patient underwent exploratory laparotomy for resection of right adnexal mass. The mass measured about 24 x 17 x 5.4 cm and had both solid and cystic components. Pathology report confirmed right sided small cell ovarian carcinoma, hypercalcemic type-stage 1C1. Genetic test for SMARCA4 was negative.

She completed 6 cycles of chemotherapy for SCCOHT (Cisplatin, Cyclophosphamide, Doxorubicin, and Etoposide) and is +100 days post autologous stem cell transplant.

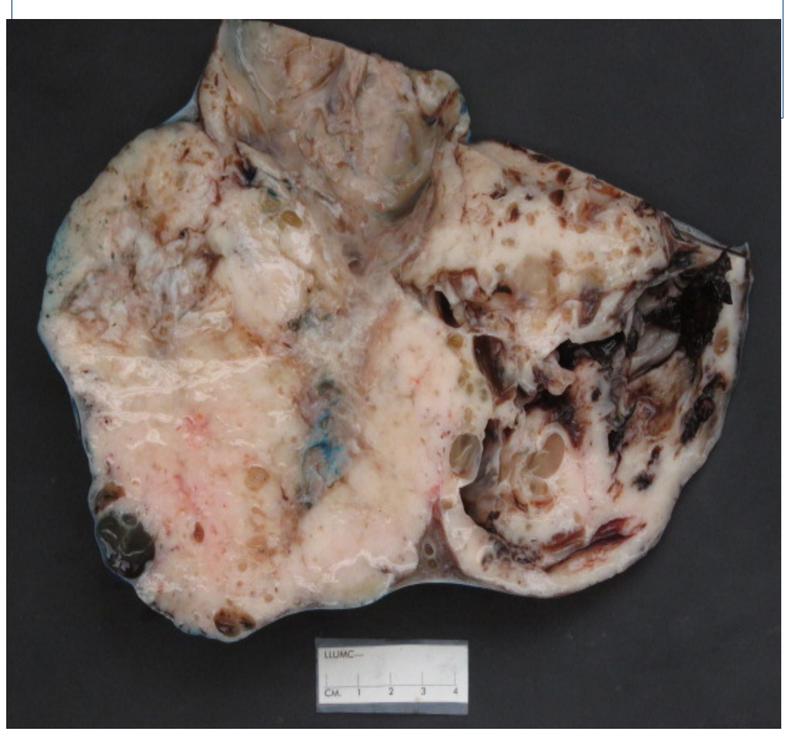

Figure 2 Gross specimen of right adnexal mass

Discussion

Hypercalcemia of malignancy occurs in about 20% of all cancer patients. Only 5% of gynecologic malignancies are associated with hypercalcemia, most commonly clear cell and small cell carcinomas. SCCOHT is associated with increased production of PTHrp. PTHrp promotes tumor growth and metastasis, resulting in poor prognosis of this type of malignancy.

A retrospective analysis showed that in earlier stages of SCCOHT, surgical resection followed by multi-agent chemotherapy and radiation may improve survival[3]. However, no standard therapy is yet available.

SCOOHT can be associated with mutation of the SMARCA4 gene, which is involved in regulating cellular transcription. Through improved understanding of the mechanism of SCOOHT, the hope is that a more targeted and effective therapy for patients will be developed.

Conclusion

The most common cause of hypercalcemia with elevated PTHrp in the pediatric population is malignancy. Thus, hypercalcemia in adolescent patients should prompt a consideration to workup for malignancy. Once the diagnosis of SCOOHT is made, genetic testing should be done in order to maximize the chance of a patient's survival through targeted therapy.

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