Derby Gynaecological Cancer & Laparoscopic surgery Centre

Gynae Oncology Symposium 2016

26th February 2016

Derby Hospitals NHS
NHS Foundation Trust

Small Cell Cancer of the Ovary

Case presentation & Review of Literature

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Taking pride in caring

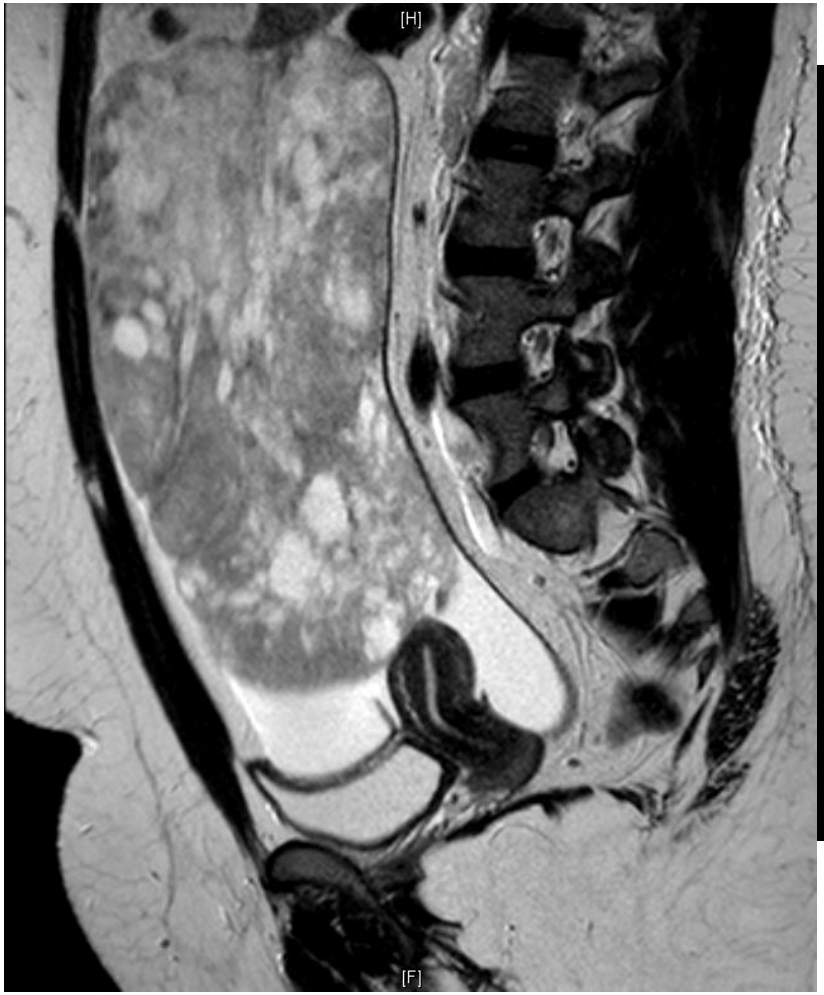
DELTA Centre
Derby Endoscopic & Laparoscopic Training & Academic Centre

Contents

- Recent Case at RDH
- Epidemiology
- Characteristic Features
- Treatment options
- Prognosis

Case: HW

- 16yr old
- PC: 4/52 hx of RIF pain- admitted with severe pain
- PMH: PCOS (Pelvic US 4/12 normal)
- FHx: Mother died (unknown cause) Dec 2014
- Ix: -
- USS/MRI - 20cm complex pelvic mass
- Raised Ca 125 72, LDH 682- AFP, Bhcg, Ca 19-9 - all normal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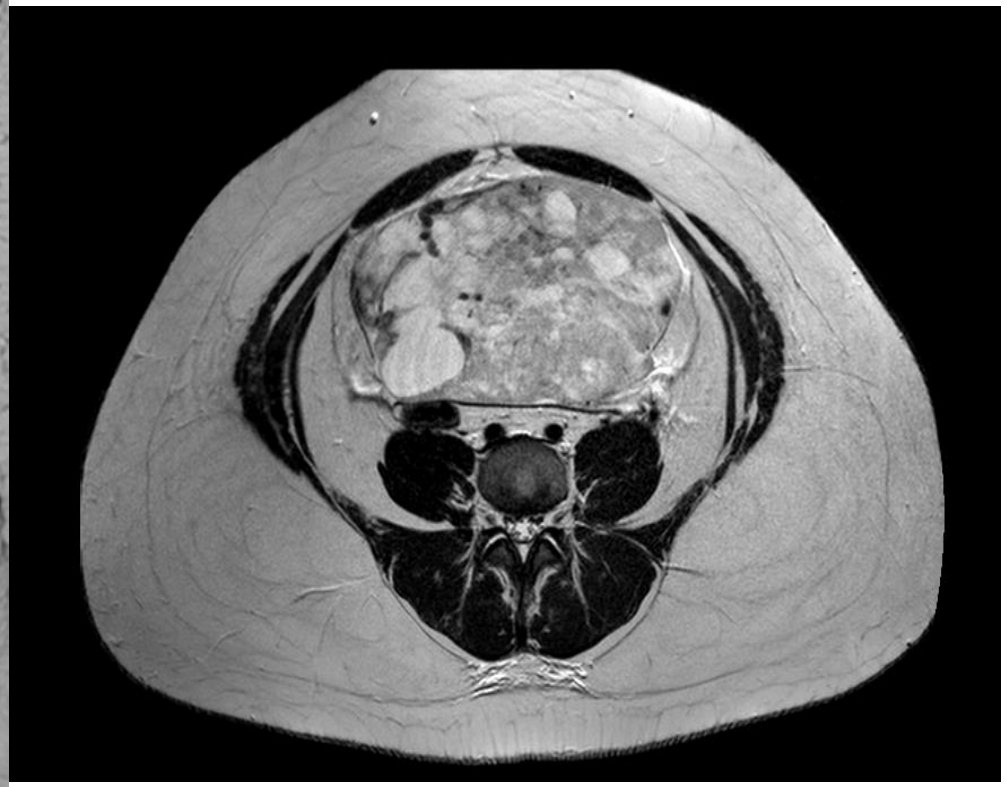

Case: HW

- Discussed at Gynae MDT on 24/12/15
- Rx- Midline Laparotomy, left salpingo-oopherectomy, peritoneal washings and infracolic omentectomy
- Findings at Laparotomy: - 20cm solid left ovarian mass
- Discharged well

- Histology showed a primary small cell carcinoma of the hypercalcaemic type, Figo 1c

- Chemotherapy commenced Feb 2016

Epidemiology

- Rare and biologically very aggressive
- Unknown histogenesis
- Mean age 24 years
- 85% Caucasian race
- 96% are unilateral tumours
- >40% present at Stage 3

Features

- Strong association with germline or somatic mutations in SMARCA4 gene
- Similarities to malignant rhabdoid tumours
- ~60% asymptomatic paraneoplastic hypercalcaemia
- Raised Ca125

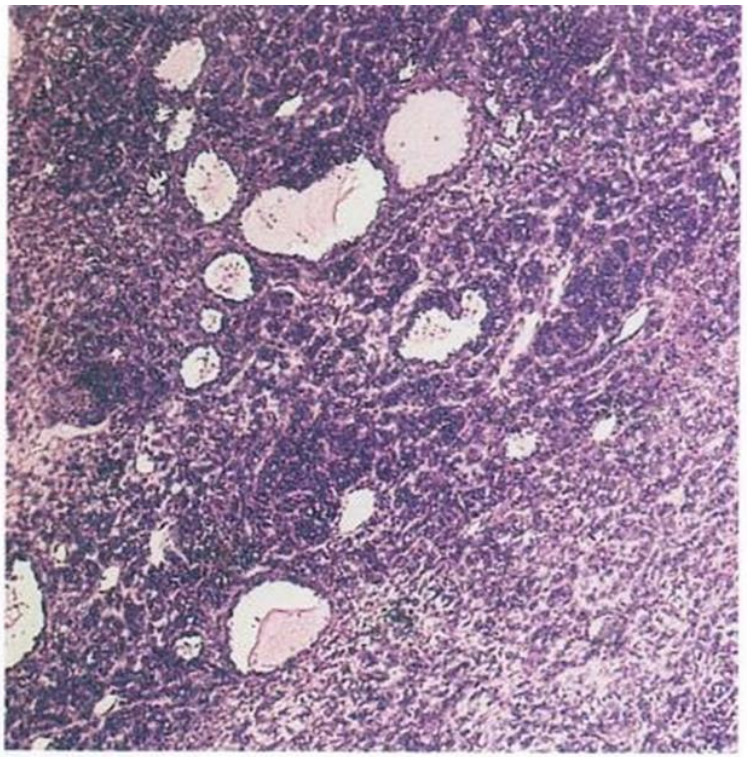

Figure 17-10
SMALL CELL CARCINOMA, HYPERCALCEMIC TYPE
Follicle-like structures of varying sizes containing eosinophilic material are present within a densely cellular tumor.

Treatment

- No definitive treatment regime established
- Surgery → Resection/debulking
- Multi-agent chemotherapy
- Radiotherapy
- ? autologous bone marrow transplantation

Prognosis

- 1 year survival rate 50%
- 5-year survival rate approximately 10%
- Almost all patients with Stage 1b disease or higher died of their disease
- ~75% develop recurrence
- Better prognosis in Stage 1a:-

- Age > 30
- Normal pre-op Calcium
- Tumour size <10cm
- Absence of large cells on histology

Summary

- Small cell ovarian cancer (hypercalcaemic type) is a rare and highly aggressive tumour predominantly affecting young women
- Many present with a paraneoplastic hypercalcaemia and a raised ca125
- The tumours resemble rhabdoid masses with new research to suggest causative mutations in the SMARCA4 gene
- Mainstay of treatment continues to be Surgery + multi-agent chemotherapy +\- radiotherapy, with a potential survival benefit with autologous bone marrow transplantation
- Prognosis despite treatment remains grim due to high recurrence rates, with few living more than two years

References

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