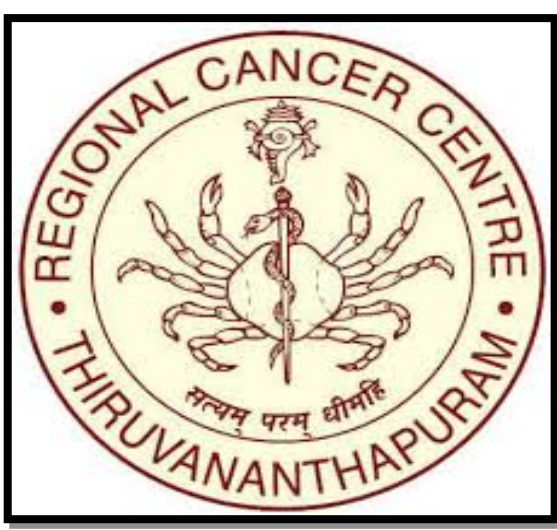


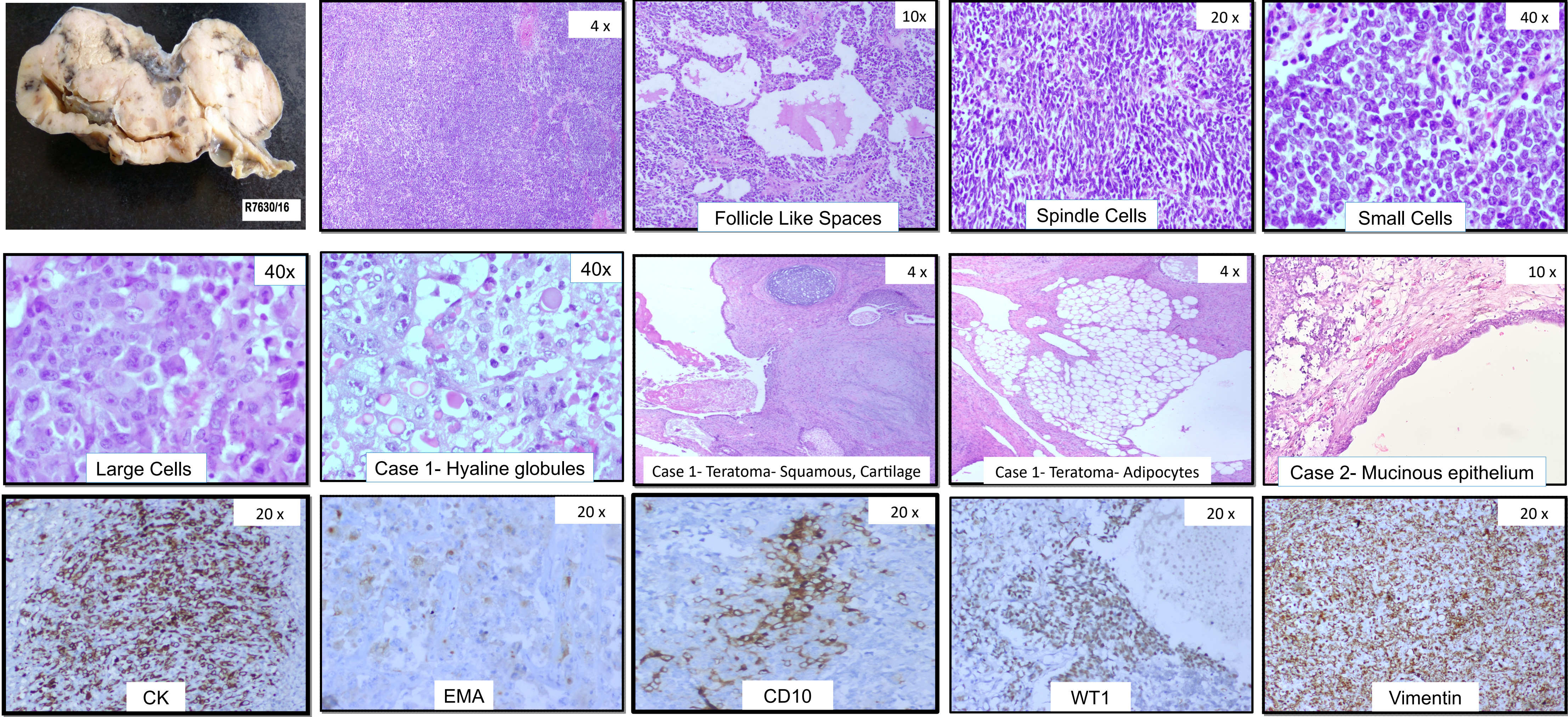


SMALL CELL CARCINOMA OVARY HYPERCALCEMIC TYPE (SCCOHT): A SERIES OF 4 CASES INCLUDING ONE WITH TERATOMATOUS COMPONENT

Dr. Ram.G.Gopal, Dr. Preethi.T.R, Dr. Anitha Mathews, Dr. Sindhu Nair P, Dr. John Joseph, Dr. Priyakumari T, Dr. Jayasree K.

REGIONAL CANCER CENTRE, KERALA, INDIA

	CASE 1	CASE 2	CASE 3	CASE 4
Age	30	10	25	13
Clinical findings	Abdominal pain, menorrhagia-3 months	Abdominal pain, vomiting-1 month	Abdominal pain and distension-1 month	Abdominal pain and distension-1 month
Calcium level pre-op	Not done	Not done	Not done	Not done
Calcium level recurrence	9.5 mg/dL	10.08 mg/dL	9.9 mg/dL	9.5 mg/dL
Type of surgery	Unilateral Salphingo-ooporectomy	Unilateral Salphingo-ooporectomy	Unilateral Salphingo-ooporectomy	Unilateral Salphingo-ooporectomy
Gross findings	18x18x13 cm- Nodular grey-white with irregular shaggy cysts	13x12x15 cm- Solid brown to black with haemorrhagic and cystic areas	15x10.5x2.5 cm- Solid grey-white, focally friable, partly lobulated with cystic areas,	3x1x0.5 cm- Solid grey-white to brownish
Microscopy	Sheets of small cells with follicle like spaces mixed with large cell areas, focal hyaline globules and a minor mature teratomatous component.	Small cell and large cell areas. An occassional focus showed a few cysts lined by mucinous epithelium.	Small cell and large cell areas. Focally cells showed nuclear grooves . Spindle cell areas. Hyalinised and myxoid stroma	Small cells in reticular pattern, in sheets with follicles like spaces and in strands in a myxoid stroma.
Differential diagnosis	Mixed Germ cell tumour	- Adult granulosa cell tumour - Neuroendocrine tumour	- Adult Granulosa Cell Tumour - Neuroendocrine tumour	- Mixed germ cell tumour -Yolk Sac Tumour and embryonal carcinoma - Neuroendocrine tumour
Positive IHC	CK, EMA (focal paranuclear) CD10, WT1, Vimentin,	CK, WT1, EMA (focal paranuclear)	CK, EMA (focal paranuclear), CD10, WT1, Vimentin	CK, Vimentin
Negative IHC	OCT3/4, PLAP, CD 30, CKIT, CK7, Synaptophysin, Chromogranin, Desmin.	Inhibin, Calretinin, Synaptophysin Chromogranin, OCT 3/4, CKIT	Inhibin, Calretinin, Synaptophysin, Chromogranin, Desmin, CK7	PLAP, CD30, AFP, WT1, Inhibin, Calretinin, Synaptophysin, Chromogranin. Desmin



DISCUSSION

- Rare highly aggressive undifferentiated neoplasm of young women.
- Of uncertain histogenesis.
- 60% cases develop Hypercalcemia. 50% cases show large cells and 80% cases show follicle like spaces.
- Exhibits loss of SMARCA4 gene
- Loss of BRG1 on IHC is a sensitive marker for diagnosis of SCCOHT

FOLLOW UP

- Cases 1, 2 and 3 recurred in 2-3 months after surgery , case 4 recurred in 6 months. All patients succumbed to the disease despite chemotherapy.
- **Hypercalcemia was not present at recurrence in any of the 4 cases**

REFERENCES

1. Young RH, Oliva E. Small cell carcinoma of the ovary, hypercalcemic type: a clinicopathologic analysis of 150 cases. Am J Surg Pathol. 1994; 18: 1102–1116
2. Foulkes WD, Clarke BA et al. No small surprise - small cell carcinoma of the ovary, hypercalcaemic type, is a malignant rhabdoid tumour. J Pathol. 2014 Jul;233(3):209-14
3. W G McCluggage; E Oliva et al . An Immunohistochemical Analysis of Ovarian Small Cell Carcinoma of Hypercalcemic Type International Journal of Gynecological Pathology. 23(4):330-336, October 2004
4. Conlon N; Silva A. Loss of SMARCA4 Expression Is Both Sensitive and Specific for the Diagnosis of Small Cell Carcinoma of Ovary, Hypercalcemic Type, American Journal of Surgical Pathology: March 2016 - Volume 40 - Issue 3 - p 395-403
5. Karanian-Philippe M, Velasco V. SMARCA4 (BRG1) loss of expression is a useful marker for the diagnosis of ovarian small cell carcinoma of the hypercalcemic type (ovarian rhabdoid tumor): a comprehensive analysis of 116 rare gynecologic tumors, 9 soft tissue tumors, and 9 melanomas. Am J Surg Pathol. 2015 Sep;39(9):1197-205.