

Steroid Cell Tumor

Abdul Al-Farsi, *Ayman Al-Talib

ورم الخلية الستيرويدية

عبدول الفارسي، أيمن الطالب



Figure 1: Clitoromegaly

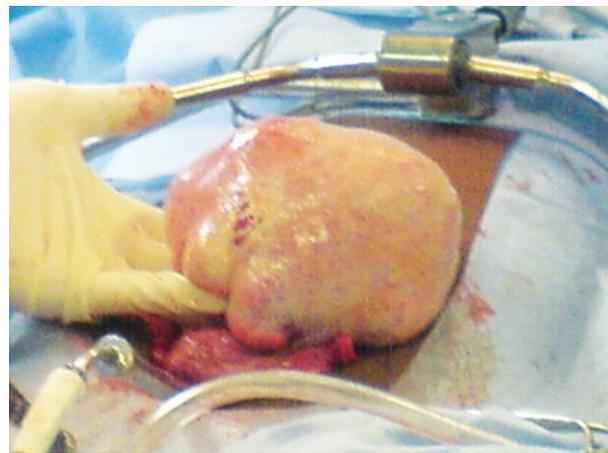


Figure 2: Steroid cell tumor gross appearance

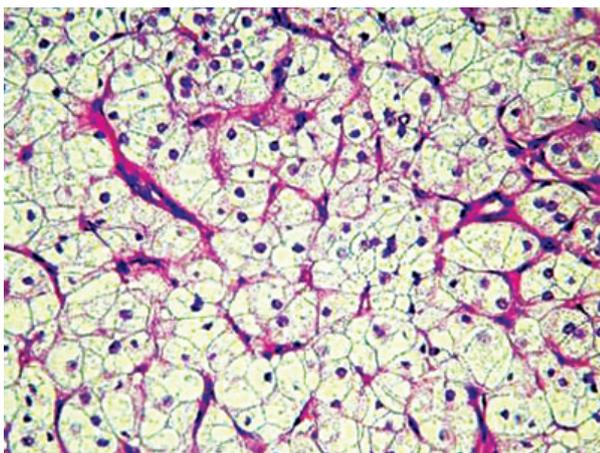


Figure 3: Histological appearance of steroid cell tumor

A 42-YEAR-OLD WOMAN PRESENTED WITH abdominal pain, increasing hirsutism and clitoromegaly [Figure 1]. Her plasma testo-

sterone levels were very high (total testosterone: 33.3 ng/mL, free testosterone: 912 pg/mL) and the cancer antigen 125 (CA-125) test was 120 U/mL. The computed tomography showed a large, 12.7 x 7.2 cm, lobulated, enhancing mass with central necrosis originating from the right ovary with a moderate amount of ascites. The preoperative diagnosis was Sertoli-Leydig cell tumor. The patient had an exploratory laparotomy where a total hysterectomy, bilateral salpingo-oophorectomy, lymph nodes dissection and omentectomy were performed [Figure 2]. The tumour, which was removed, was confined to the right ovary and there was no evidence of adjacent structures involvement. No ascites was seen. During the surgery, the patient's blood pressure went up to 240/140 mmHg. Postoperatively, the patient was doing well though her blood pressure was uncontrolled so was started on

Micardis® (telmisartan). Serum levels of testosterone returned to normal a few weeks after the surgery. The histopathological diagnosis was a steroid cell tumor, not otherwise specified [Figure 3].

Steroid cell tumours of the ovary account for approximately 0.1% of all ovarian tumours and are subdivided into 3 subtypes: stromal leuteomas, Leydig cell tumours, and steroid cell tumours, not otherwise specified (NOS). The steroid cell tumour (NOS) is the most common of the 3 subtypes, accounting for approximately 60% of these tumors and the majority of them show virilisation,^{1,2} 25-45% of which are clinically malignant.^{2,3} Estrogen secretion occurs in 6% to 23% of the tumors, which may be associated with menorrhagia, postmenopausal bleeding, or even endometrial adenocarcinoma. Cushing's syndrome occurs in 6% to 10% of the cases. Approximately 25% of the cases of steroid cell tumours (NOS) are not associated with hormonal disturbances.⁴ They can occur at any age, but usually develop in adults with an average age of 43 years.¹ Steroid cell tumours often present as unilateral solid tumours and occasionally as cystic tumours. Necrosis or calcification is frequently associated.⁵ Ovarian steroid cell tumours are characterised by cells with abundant intracellular lipids, which are similar to adrenocortical cells.⁶ The classical gross finding of steroid cell tumor (NOS) is a solid, well-circumscribed ovarian mass. Microscopic findings include diffusely arranged cells, although tumor cells may be present in nests, clusters, cords, or columns. The stroma is most commonly scant, but may be prominent and can oc-

asionally be fibromatous, edematous, or myxoid. The cells are polygonal to round, with distinct cell borders and central nuclei, and they often have prominent nucleoli. The cytoplasm varies from spongy in lipid-rich cells to granular and eosinophilic in lipid-poor or lipid-free cells [Figure 3]. The mainstay of ovarian steroid cell tumor is surgery. Careful follow-up evaluation should include a measurement of sex hormone levels, particularly for those patients who demonstrated elevated levels before removal of the primary tumor.

REFERENCES

1. Kim YT, Kim SW, Yoon BS, Kim SH, Kim JH, Kim JW, et al. An ovarian steroid cell tumor causing virilization and massive ascites. *Yonsei Med J.* 2007; 48:142-146.
2. Veras E, Deavers MT, Silva EG, Malpica A. Ovarian steroid cell tumor, not otherwise specified: a clinicopathologic study. *Zhonghua Bing Li Xue Za Zhi* 2007; 36:516-520.
3. Liu AX, Sun J, Shao WQ, Jin HM, Song WQ. Steroid cell tumors, not otherwise specified (NOS), in an accessory ovary: a case report and literature review. *Gynecol Oncol* 2005; 97:260-262.
4. Malaika W, Sathima Natarajan. Pathologic Quiz Case: A Rare Tumor of the Ovary. *Arch Pathol Lab Med* 2003; 127:890-892.
5. Faraj G, Di Gregorio S, Misiunas A, Faure EN, Villabrile P, Stringa I, et al. Virilizing ovarian tumor of cell tumor type not otherwise specified: a case report. *Gynecol Endocrinol* 1998; 12:347-352.
6. Hayes MC, Scully RE. Ovarian steroid cell tumors (not otherwise specified). A clinicopathological analysis of 63 cases. *Am J Surg Pathol.* 1987; 11:835-845.