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Steroid Cell Tumours Not Otherwise Specified – A Review of A Rare Condition

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Introduction:

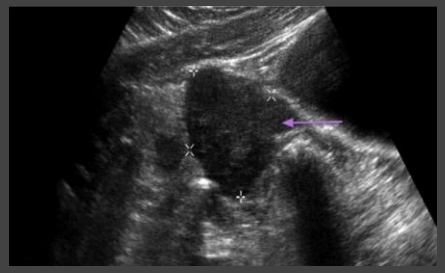
Steroid cell tumours not otherwise specified (SCT NOS), one of the three subtypes of steroid cell tumours, are sex cord-stromal tumours of the ovary. SCT NOS is a rare condition with the potential of malignancy albeit most of them are benign. There are quite a few published case reports regarding this condition, however, only a limited amount of studies have attempted to identify the aetiology, risk factors, and treatment. The purpose of this review is to summarize the existing evidence which may throw light on avenues for future studies.

Common presentation

It was found the average age of diagnosis on SCT NOS is 47 but it may present at any age. The clinical presentations of SCT NOS vary but most cases presented with characters of hirsutism and virilisation. The common clinical presentations are not specific, commonly including abdominal pain, distention, and bloating. However, most of the significant presentations are those associated with the hormonal activity and virilizing properties of the tumor, which accounts for 56%-77% of patients. Signs and symptoms of masculinizing tumors usually present in two definite phases, an early phase of defeminization and a subsequent phase of masculinization. Typically, the first noticed symptom is oligomenorrhea or amenorrhea. Common signs of masculinization include hirsutism, acne, clitoral enlargement, increased libido, sterility, enlargement of the larynx, deepening of the voice, and temporal alopecia.

Diagnosis

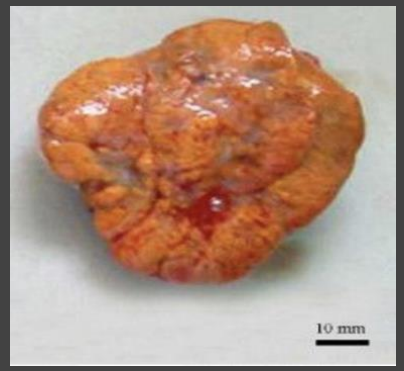
The diagnosis of SCT NOS, should be made on the basis of the clinical virilizing syndromes, the microscopic pictures, as well as immune reactivity to some immunohistochemical markers. Inhibin and calretinin were thought to be sensitive and robust markers in differentiating sex cord-stromal from non-sex cord-stromal tumors.



Treatment

SCT NOS, are also associated with malignancy. It has been reported that 43% of SCT, NOS cases were malignant. Malignancy is generally associated with pathological features: tumour size, mitotic figures, nuclear atypia, necrosis and/or haemorrhage. Surgery is essential for the management of SCT NOS. For women who have completed their families, total hysterectomy with bilateral salpingo-oophorectomy and complete surgical staging is recommended. For women desiring future fertility, unilateral salpingo-oophorectomy is adequate if the tumour is unilateral and the pathologically malignant features are negative. Tumour resection has been reported, however, information on the possibility of recurrence is limited.

Adjuvant chemotherapy is recommended to be based on the histologic appearance of the tumor and on its surgical stage. However, there are no well defined chemotherapy guidelines for clinical management of steroid cell tumor



Conclusion

The diagnosis of SCT NOS is challenging. It tends to affect women in childbearing age, but can occur at any age. It should be a differential diagnosis in patients presenting with secondary amenorrhoea and virilising symptoms with elevated testosterone levels. More studied will be needed to identify the risk factors, methods of diagnosis, as well as the optimal management of treatment and follow up.

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