MIXED OVARY TUMOR WITH MALIGNANT GERMINAL COMPONENT AND SEX CORD-STROMAL, UNCLASSIFIED: PRESENTATION OF AN UNUSUAL CASE

Por el control del cáncer

Co-authors J. Saenz¹, M. Medina², M.I.Beltran³, O. Suescun², A. Almeciga², P. Calderon²

¹Especialista en entrenamiento-Universidad Militar Nueva Granada-Instituto Nacional de Cancerología, Department of Gynecologic Oncology, Bogota D.E., Colombia.

²Instituto Nacional de Cancerología, Department of Gynecologic Oncology, Bogota D.E., Colombia.

³Instituto Nacional de Cancerología, Department of Pathology Oncological, Bogota D.E., Colombia.

Background and Aims:

It is an Ovarian neoplasm composed by germ cells and elements of the sexual cords, in genetically and phenotypically normal women without the morphology of gonadoblastoma. In 1972, Talerma introduced the term for these neoplasms. The objective of this work is to report an unusual case of a 14-years-old patient, with precocious puberty who presented this tumor.

Clinical case:

A 14-year-old patient with a history of abdominal pain. Ultrasound evidenced of solid abdominopelvic mass with areas of cystic degeneration, diameters 22 x 13 x 10 cm. Antecedent of precocious puberty and menarca at 8 years old. Phenotypical without alterations. She was taken to surgery, evidence of right ovarian tumor, predominantly solid, smooth surface and multilobed. It weighed 2460 grams and sized 24 x 18 x 11 cm. No pelvic or para-aortic adenomegalies were found. Pelvic cavity without metastatic involvement. The histological report showed mixed tumor of ovary with malignant germinal component and stromal-unclassified sexual cords: endodermal sinus tumor and dysgerminoma (70%) and sexual cord tumor with annular tubules (30%). It was classified as Stage IA. Immunohistochemistry is Positive in an endodermal sinus component: AFP, Oct3-4, Sall4, PLAP, Cd117. In sexual cords component: Inhibin, Ck, Calretinin. Negative: Synaptophysin, Chromogranin. Prior to tumor resection Alfetoprotein levels > 1000 ui / ml (vr: children 91 days to 18 years: less than 9.96 ui / ml), LDH 15312 u / l, Bhcg positive (vr: 0 to 5.0). At the tenth postoperative week Alfetoprotein: 2.1, LDH: 459 u / l, Bhcg <1.0 mui / ml (vr: 0 to 5.0). She received adjuvant chemotherapy with Bleomycin-Etoposide-Cisplatin scheme for 3 cycles. One year after surgical resection in disease-free period.

40x Dysgerminoma component: abundant clear or slightly eosinophilic cytoplasm, large round nuclei with prominent nucleoli and energetic mitotic activity.

10x Endodermal Sinus component channels forming microcysts coated by atypical epithelial cells. Focally similar to Schiller-Duval body-like fibrovascular structures.

10x Component of sex cords with annular tubules. Where reverse polarity is recognized.

Discussion

This is an infrequent neoplasm reported in the literature. Approximately 10% of these tumors have malignant germ cell components compared to 60% of gonadoblastomas. It differs from gonadoblastoma by its macroscopic appearance, histological pattern, absence of regressive changes and occurrence in normal gonads of phenotypic and genetically normal women. Its presentation includes abdominal pain, abnormal uterine bleeding, ascites and abdominal and pelvic masses. Disease-free periods vary from 1 to 15 years, and only three cases reported recurrence or metastasis. They are less frequent cause of sexual precocity in girls, but should be taken as a differential diagnosis by its etiology. The prognosis is favorable.

Conclusions

This is a very rare neoplasm, the management being the resection of the gonad that contains the tumor and the conservation of the opposite side gonad that is normal. It should be considered in the differential diagnosis of a girl with an abdominal mass and precocious puberty.

• Keywords

Ovarian neoplasms, Germinal, Sex Cord-Stromal Tumor.

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