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Wolffian Tumor and Peutz-Jegher Syndrome: A Case Report

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1. Introduction

The Female Adnexal Wolffian Origin (FATWO) is an extremely rare gynecological neoplasm first described in 1973, and with less than 90 cases described in English. [1,2] The currently preferred terminology, according to the World Health Organization Tumor Classification, is Wolffian tumor.3 It is a neoplasm with low malignant potential and is presumed to be derived from mesonephric (Wolffian) duct remnants in the upper female genital tract, and the most frequent location of is in the broad ligament, but they can also be found in the fallopian tube, paravaginal area, ovary or the retroperitoneum. [2,4,5] The rarity of reported cases, nonspecific clinical manifestations, ill-defined radiological features and with variable morphology make the diagnosis of wolffian tumor very challenging. [2] We reported a case of Wolffian tumor in a young woman with clinical Peutz Jeghers Syndrome.

2. Case Report

A 21-year-old woman presented to the emergency department complaining of diffuse abdominal pain. She was being treated for a urinary tract infection due to a previous complaint of dysuria. Computerized tomography showed a tumor in the right fallopian tube suggesting ruptured ectopic pregnancy. A laparoscopy was performed and showed a mixed, heterogeneous and multilocular tumor, with 10 cm, located in the region of the right annex and attached to the ipsilateral fallopian tube. The patient underwent surgery with complete resection of the tumor.

The anatomopathological examination demonstrated malignant epithelioid proliferation characterized by high-grade atypical cells that formed masses and microcysts with a myxoid background. The exam questioned a juvenile granulosa tumor. Therefore, an immunohistochemical exam was requested as diagnostic propaedeutic. Immunohistochemistry concluded that it was a wolffian tumor, with the following antigens tested (Figure 1). The patient was referred for oncological follow-up and genetic counseling given the recent diagnosis and family history of the syndrome - her mother had a clinical diagnosis of Peutz Jeghers syndrome with mucocutaneous pigmentation, polyposis in the gastrointestinal tract and breast cancer (Figure 2) (Table 1).

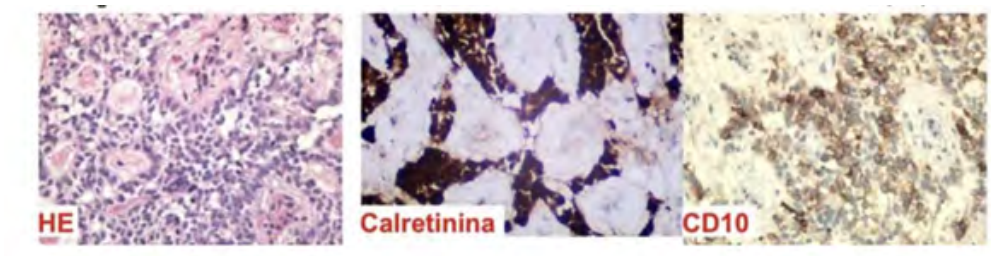

Figure 1: Wolffian Tumor

Table 1: Antigens Tested

Sequence	Antigen (clone)	Result
1	Cytokeratin AE1/AE3 (AE1/AE3)	Positive
2	Cytokeratin 7 (CK7) (OV-TL 12/30)	Positive
3	CD56 (123C3)	Positive
4	Calretinin (DAK-Calret 1)	Positive
5	WT1 (6F-H2)	Positive
6	INI-1 (MRQ-27)	Positive (preserved expression)
7	SMARCA4 (BRG1)	Positive (preserved expression)
8	Estrogen Receptor protein (EP1)	Positive in areas
9	Progesterone Receptor protein (PgR 636)	Positive in areas
10	Androgen Receptor (AR441)	Positive in areas
11	CD99 (12E7)	Positive in areas
12	Beta-catenin (ß-catenin-1)	Positive at the cytoplasmic membrane
13	Inhibin (R1)	Positive in foci
14	CD10 (56C6)	Positive in foci
15	CD117 (c-kit) (policlonal)	Positive in foci
16	P53 (DO-7)	Positive in foci
17	EMA (epithelial membrane antigen)	Positive in rare foci
18	Ki-67 (MIB-1)	Positive in 20% of neoplastic cells
19	Glypican-3 (1G12)	Negative
20	CDX2 (DAK-CDX2)	Negative
21	SALL4 (6E3)	Negative
22	PAX8 (MRQ-50)	Negative
23	Cyclin D1 (EP12)	Negative
24	GATA3 (L50-823)	Negative
25	P16 (CINTEC)	Negative

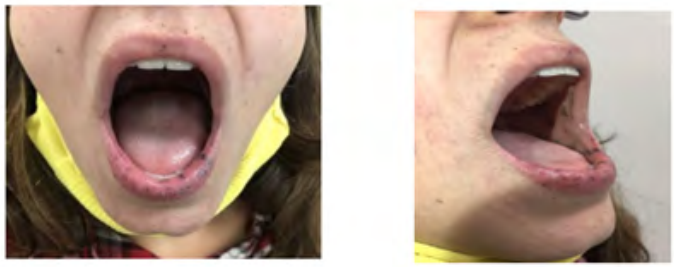

Figure 2: Clinical characteristic of the patient with mucocutaneous pigmentation

3. Discussion

Wolffian tumor is a rare gynecological neoplasm that affects women of different ages, from 15 to 83 years old, with a mean age at diagnosis of 50 years. [2] Many patients remain asymptomatic, and the tumors are discovered incidentally during physical examination or abdominal surgeries performed for other pathologies. [6,7] Peutz-Jeghers syndrome is an autosomal dominantly inherited syndrome characterized clinically by mucocutaneous pigmentation and gastrointestinal polyposis. [8,9] An elevated risk of gastrointestinal cancers, an increased risk of cancers at other sites, such as breast, ovary, uterus, cervix, lung, and testis, has been described [8-12] and rare tumors have also been attributed to Peutz-Jeghers syndrome. [8,9] A recent series cases report showed association adnexal tumor with Peutz-Jeghers syndrome and nearly 50% of cases harbores STK11 alterations. [13] The histogenesis of these tumors remains an enigma, and further studies, especially those focusing on transcriptomic, epigenetic, and proteomic analysis will hopefully aid in answering this question.

4. Conclusion

Overall, this case adds to the literature the report of a rare gynecological tumor, which affects women of different ages and presents a nonspecific clinical and radiological presentation. In addition to presenting morphological variety that complicated the pathological diagnosis and pathogenesis and molecular changes are still

not well known. There is no consensus on the management of this neoplasm, but the importance of genetic counseling must be emphasized in patients with Peutz-Jeghers syndrome.

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