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Immature Ovarian Teratoma: A Case Report

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Abstract **Case Report**

We report the case of a 21-year-old single woman who presented with a high-grade immature teratoma revealed by a progressive increase in abdominal circumference, operated on immediately, and in whom we opted for active surveillance instead of adjuvant chemotherapy, Through this observation and the review of the literature, we will be able to discuss and discover what an immature teratoma is and mainly see the indications for adjuvant chemotherapy in high-grade cases, the type of chemotherapy and the modalities of active surveillance.

Keywords: Immature ovarian teratoma, grades, Chemotherapy, Active surveillance.

INTRODUCTION

Ovarian teratomas are tumors that develop from pluripotent germ cells. A distinction is made between mature and immature teratomas. Immature teratomas account for less than 1% of ovarian cancers, and mainly affect young patients. They are non-seminomatous germ cell tumors containing a variable amount of immature embryonic tissue, usually neuro-ectodermal tissue.

OBSERVATION

This is a young woman (F.M) aged 21, single with no particular pathological history. She presented with a progressive increase in abdominal circumference. Abdominal-pelvic MRI revealed a left latero-uterine mass measuring 135 mm in diameter, hemorrhagic and multi-loculated, with thick enhanced septa and a small peritoneal effusion of organic appearance. The patient underwent direct peritoneal cytology, left adnexectomy (Figure 1), appendectomy, right parietocolic gutter biopsy and left parietocolic gutter biopsy. Pathological examination (Figure 2) showed: absence of tumour cells on direct peritoneal cytology, a solid cystic tumour of the left ovary measuring 17 cm in large diameter, whose morphological features were primarily suggestive of a high-grade immature teratoma (grade 3). or a primitive neuroectodermal tumor (PNET). Immunohistochemical analysis confirmed the diagnosis of a poorly differentiated central PNET neuroectodermal tumor of the left ovary. Post-surgical tumor markers were negative, as was the follow-up TAP scan. The tumor was classified as pT1a N0 M0, corresponding to stage I according to the FIGO classification. Two options were proposed at the multidisciplinary consultation meeting (RCP): adjuvant BEP-based chemotherapy or active surveillance. Both options were discussed with the patient, and because of the high cost of oocyte conservation, we opted for active surveillance.

Figure 1: Adnexectomy specimen

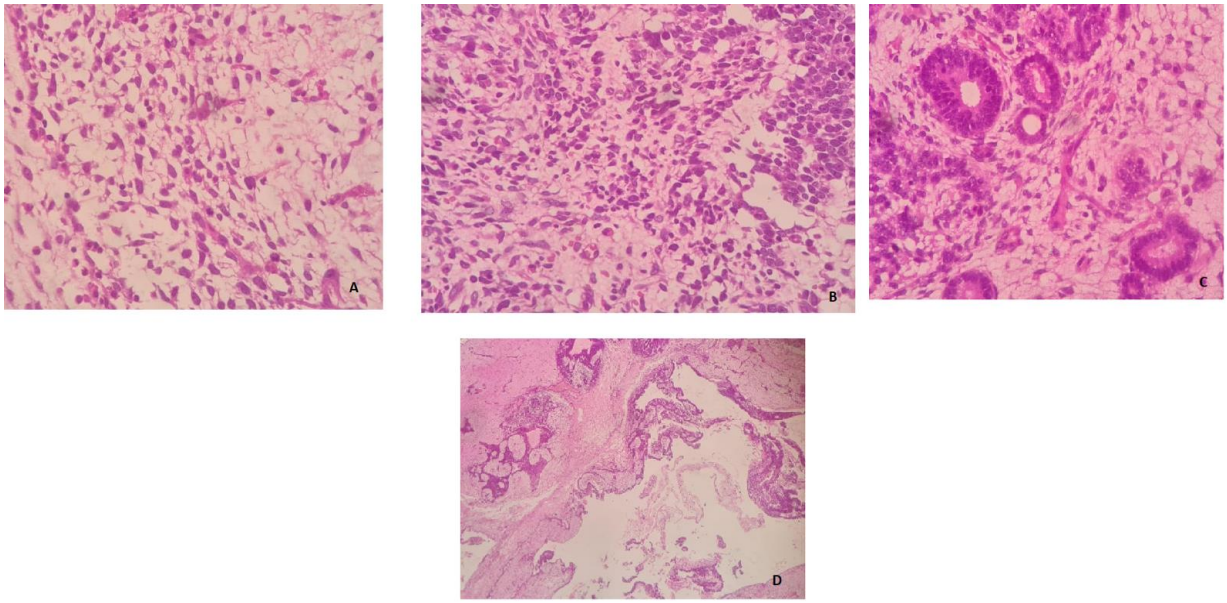


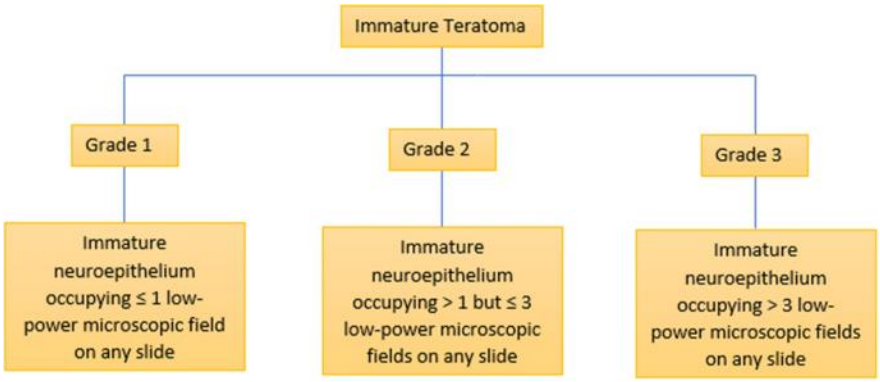
Figure 2:(A +B+C+D) microscopic appearance of immature neuro-ectodermal tissue proliferation, mixed with an immature mesenchymal background, containing immature cells, and embryonic, striated muscle differentiation

DISCUSSION

Immature teratoma of the ovary, a malignancy originating from germline cells, was initially documented by Thürlbeck and Scully in 1960 [1]. These tumors typically contain various amounts of immature embryonic tissues, often neuroectodermal in nature [2]. Immature teratomas constitute a minority of teratomas, comprising about 3% of all teratomas, 1% of ovarian cancers, and 20% of ovarian malignancies arising from germline cells [3-5]. The exact etiology of these tumors remains a subject of debate. Linder et al. proposed that teratomas originate from a single germ cell, with abnormalities occurring during the first meiotic division [6]. Subsequent research by Ohama et al. supported this notion, suggesting abnormalities might also arise during the second meiotic division or from a mature oocyte [7].

Clinically, immature teratomas typically manifest in younger individuals, with an average onset age of around 19 years [8]. However, these tumors can occur across various age groups. Symptoms are diverse, mirroring those of other ovarian masses, ranging from menstrual disturbances to pelvic pain, transit issues, and abdominal distension. Clinical examination findings are also varied, from normal to tenderness or the presence of an abdominal or pelvic mass. Imaging studies, including ultrasound, CT, and MRI, provide insights into the tumor's characteristics. Immature teratomas typically present as irregular masses with mixed tissue components and fatty areas, often containing calcifications and enhancing with contrast [9-10]. MRI may reveal voluminous tissue portions with sparse fatty areas and microcysts [9]. However, radiological findings do not reliably predict histological grade, as immature teratomas comprise tissues from all three embryonic cell lineages at various stages of maturation. Histologically, immature teratomas are classified based on Norris classification into three grades, reflecting their degree of immaturity and neuroectodermal presence [11]. However, current practice trends towards a simplified classification into low and high grade. Surgical exploration often reveals solid cystic tumors with features suggestive of high-grade immature teratoma or primitive neuroectodermal tumor (PNET). Immunohistochemical analysis helps confirm the diagnosis, with elevated alpha-fetoprotein levels indicating a higher risk of recurrence [12, 13]. In summary, immature teratoma of the ovary presents diagnostic and therapeutic challenges due to its diverse clinical and histological features. Understanding its varied presentations and potential for recurrence is crucial for effective management and patient outcomes. The progression of immature teratomas is characterized by rapid tumor growth, with primarily locoregional extension often involving peritoneal invasion necessitating capsular breach. Secondary lesions are described as superficial granulations, typically greyish or yellowish, scattered throughout the abdominopelvic peritoneum, occasionally extending to structures like the greater omentum or liver. Therapeutic approaches for immature teratomas, as delineated by the expert center for rare gynecological malignancies in 2022 and the French Society of Oncology in 2013, entail a two-pronged strategy: surgical intervention followed by chemotherapy, tailored to the histological grade [14, 15]. Surgical intervention aims at diagnosis, therapeutic tumor removal, and staging. The initial surgery often includes unilateral adnexectomy, complete pelvis and abdominal cavity exploration, peritoneal lavage, and/or ascites removal, alongside multiple peritoneal biopsies and excision of suspicious elements. Lymph node dissection is not warranted unless abnormalities are detected on imaging or palpation. The conservative

surgical approach aims to preserve fertility, emphasizing the need for meticulous surgical techniques. Our patient underwent laparotomy revealing a 17 cm ovarian mass, leading to unilateral adnexectomy with multiple biopsies and appendectomy. Histopathological examination confirmed a grade 3 immature ovarian teratoma, dictating subsequent treatment decisions. Grade 1 tumors typically warrant clinical surveillance post-surgery, given their favorable prognosis. In contrast, adjuvant chemotherapy, often employing BEP protocol, is reserved for grade 2 or 3 tumors, significantly improving prognosis [15]. Advances in chemotherapy, particularly platinum-based regimens, have notably enhanced survival rates for immature teratomas. Overall, prognosis for immature teratomas correlates with tumor grade, with grade 1 tumors exhibiting excellent 5-year survival rates, while grade 3 tumors present higher recurrence and mortality rates [2, 12, 17]. In our case, the tumor was classified as stage I, aligning with literature recommendations advocating for surgical management, with subsequent discussions regarding adjuvant therapies tailored to the patient's preferences and clinical considerations.



tiered grading system.

	Histological criteria
Grade 1	Tumors with rare foci of immature neuroepithelial tissue that occupy < 1 low power field (40 x) in any slide (**low grade**).
Grade 2	Tumors with similar elements, occupying 1-3 low power fields (40 x) in any slide (**high grade**).
Grade 3	Tumors with large amount of immature neuroepithelial tissue occupying > 3 low power fields (40 x) in any slide (**high grade**).

	High grade/low grade classification	FIGO Stage
Ovarian tumor gade 1	Low grade	Ia
Ovarian tumor grade 2-3	High grade	Ia
Implants grade 2 or 3	High grade	>II
Grade 0 peritoneal implants regardless of the grade of the ovarian tumor		>II

The grading of immature ovarian teratomas into high grade/low grade and their correspondence

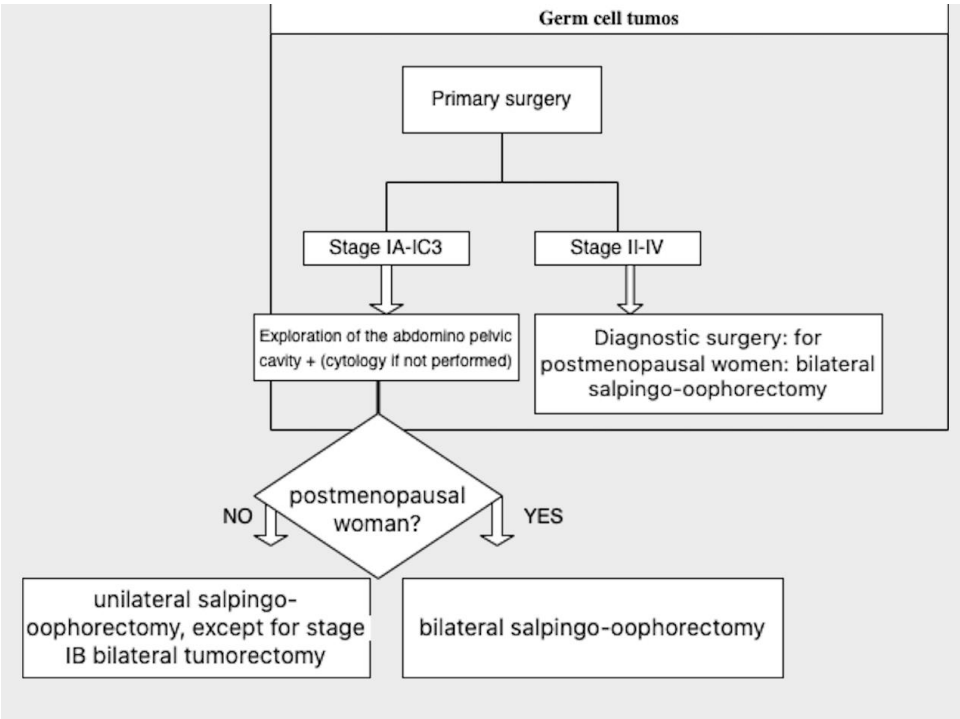


Figure 3: Decision tree for treatment

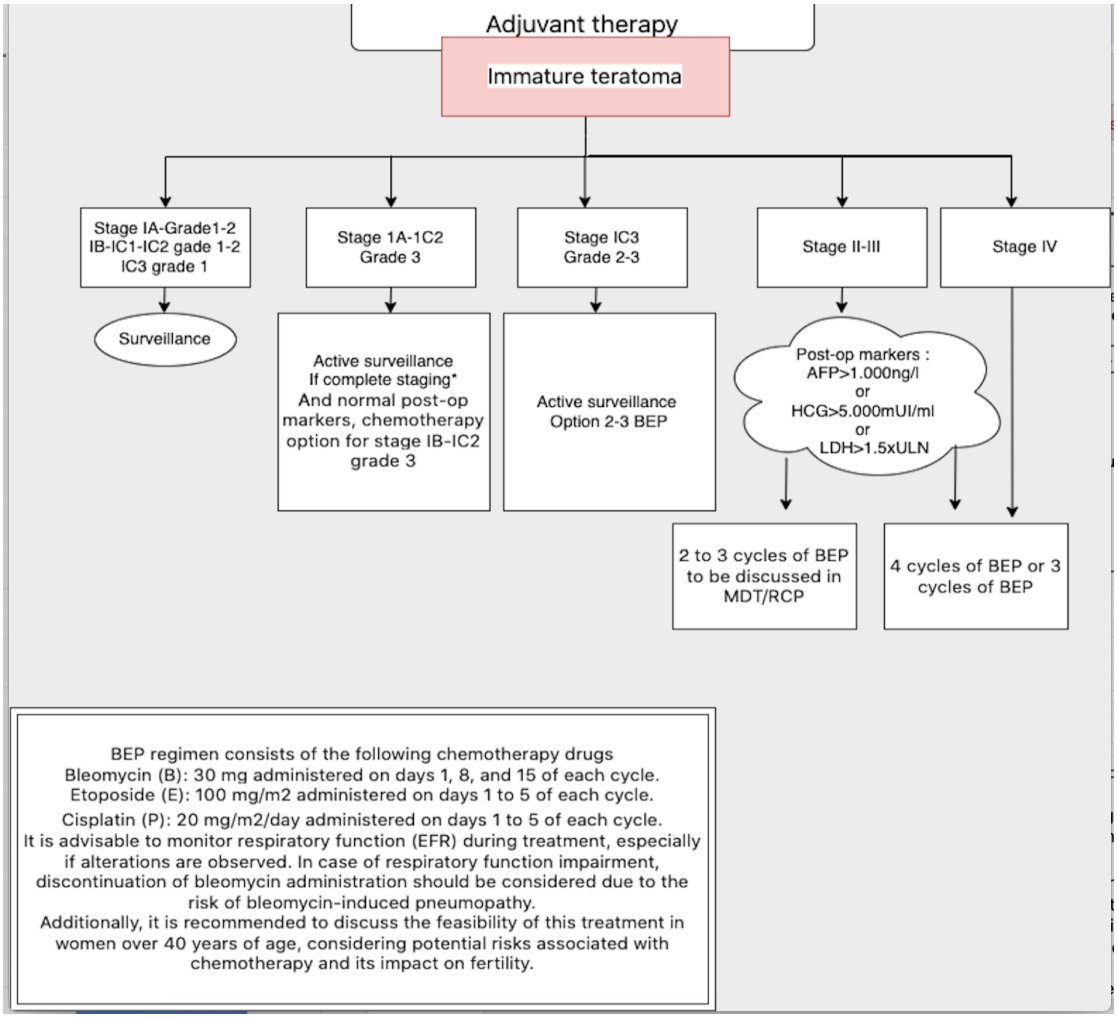


Figure 4: Adjuvant Treatment (Chemotherapy/Active Surveillance)

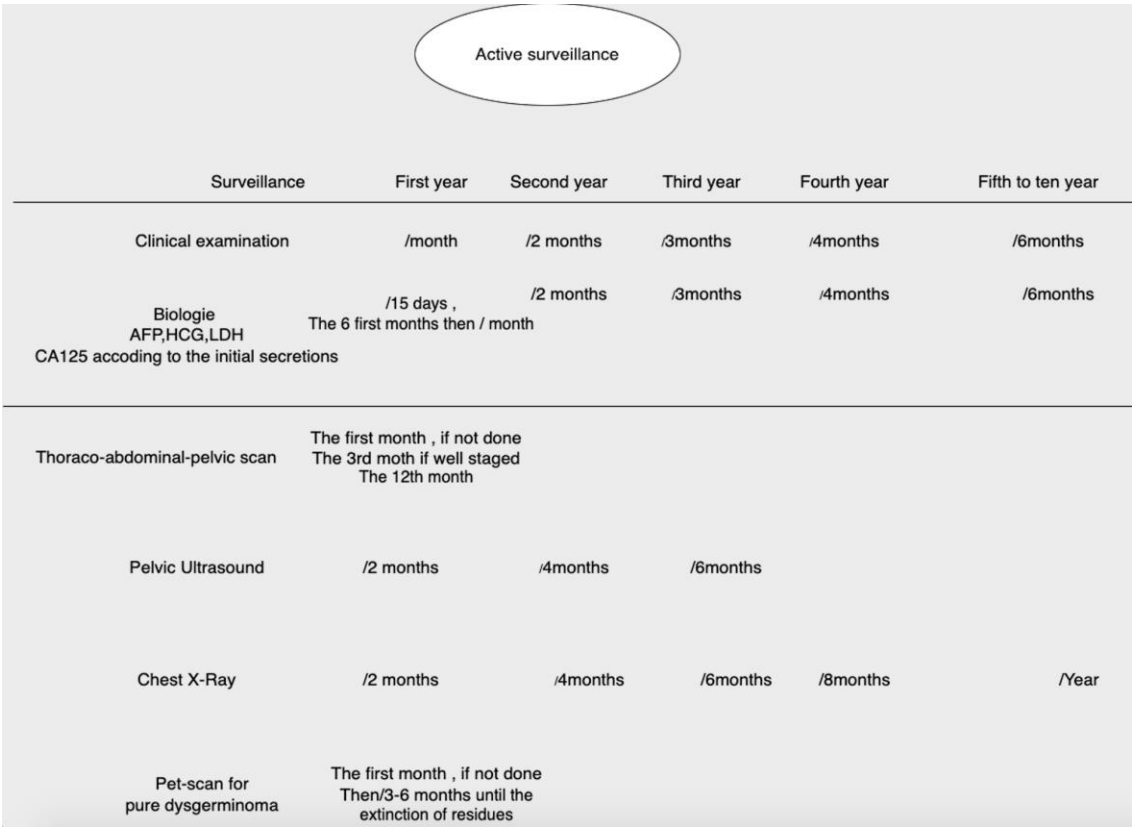


Figure 5: Active surveillance

CONCLUSION

The prognosis of immature teratoma is intricately linked to its histological grade. Predominantly affecting young women, its diagnosis is typically prompted by radiological findings and definitively confirmed through histopathological analysis. Management strategies often favor a conservative surgical approach aiming to preserve fertility, complemented by chemotherapy tailored to the tumor grade. Effective therapeutic planning necessitates multidisciplinary collaboration among oncologists, gynecologists, and pathologists.

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