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Case report

UNDIFFERENTIATED SARCOMA ARISING IN AN IMMATURE TERATOMA OF THE OVARY: A case report

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ABSTRACT

Introduction: The immature teratomas present less than 1 % of ovarian cancers and affect preferentially young women (1). We report a rare case of an ovarian teratoma harboring undifferentiated sarcoma highlighting the clinical and pathological characteristics.
Case presentation: The patient was a 24-year-old female who consulted for an abdominal mass. Pelvic ultrasound and magnetic resonance imaging (MRI) revealed an ovarian teratoma measuring 22 cm with no evidence of malignancy. She underwent an exploratory laparotomy. She had a unilateral adnexectomy as the ovarian parenchyma was damaged and the fallopian tube was involved in the mass. The final anatomopathological examination revealed the presence of a teratoma with a predominant component of undifferentiated sarcomatous. This component made the prognosis worse and led to a rapid unfavorable evolution with numerous metastases (lung, liver, and peritoneum). She refused all treatment and died within a week of diagnosis.
Discussion: Immature teratoma is a malignant tumor composed of tissues derived from the three embryonic cell lineages at different stages of maturation. This fact makes the determination of its histologic grade by radiologic examination very difficult (1) and consequently raises a challenge in the workup as the prognosis of immature teratomas is closely related to their histologic grade (16). Improved chemotherapy after surgery has increased the 5-year survival rate for these malignancies from 90 % to 100 % (18).
Conclusion: Young women are most commonly affected by immature teratomas. The prognosis and histologic grade are interrelated. To protect fertility, the trend now is treat them more frequently with conservative surgery.

1. Introduction

Ovarian teratomas are derived from pluripotent germ cells. The immature teratomas present less than 1 % of ovarian cancers and affect preferentially young women [1]. Data on immature ovarian teratomas are scarce because of their rarity. They have particular histologic features, and little is known about the different modalities of therapy and results [2]. Indeed, there is a considerable debate concerning their proper management at this age group. Unilateral salpingo-oophorectomy with a variety of peritoneal implants is considered as the standard treatment, the usefulness of chemotherapy, especially for higher grade and stage diseases, is highlighted by different researches [3]. Through a case report of an ovarian teratoma harboring undifferentiated sarcoma managed in our department of gynaecology and a literature review, we will study the epidemiologic, therapeutic, and prognostic aspects of immature teratoma.

2. Methods

The writing of this case report is consistent with the 2020 guidelines [4].

3. Presentation of case

This was a 24-year-old female student with no surgical or medical history. She was single. Her complaint was a progressive increase in abdominal volume since 4 months followed by the onset of pelvic pain. An abnormal uterine bleeding (Metrorrhagia) brought her to our gynaecology department.

The patient did not report any changes in her bowel habits nor in

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frequency of urination. She did not lose weight during this period. She had no family history of gynecological nor abdominal cancer. She denied being sexually active. On physical examination, she was pale with a relatively good general state. Her vitals were within normal limits. Her height was 1.68 m and her weight 62 kg, with a body mass index of 22 kg/m2. Abdominal examination revealed a huge mass exceeding the umbilicus without portal collateral circulation. It was firm and painless. Rectal examination and the rest of the physical exam were normal.

The ultrasound showed a solid cystic mass occupying the entire abdomen. Additional magnetic resonance images (MRI) exploration revealed the presence of a large well defined abdominopelvic mass measuring 24 cm of long axis with triple component evoking a teratoma in the left ovarian (Fig. 1). Biology exam found a severe microcytic and hypochromic anemia (7 g/d/VGM = 69.2/TCMH = 20.9), hyperplaquettose at 571000/mm3, a normal rate of white blood cells at 9900/mm3, LDH at 293 mmol/l and a CRP of 88.34 mg/l. This anemia could be due either to abnormal uterine bleeding or to an inflammatory phenomenon (elevated CRP). As her hemodynamics were stable, a treatment with venous ferrous sulfate was started. The tumor marker assay revealed an elevated CA125 level at 121 IU/ml, and a normal alpha-fetoprotein level at 1.14 IU/ml. Surgical resection was suggested as the first line of treatment. The patient accepted to proceed with an open surgical tumor resection after receiving the necessary counseling about potential risks of the procedure being explained. Given the young age and the absence of signs highly suggestive of malignancy, a large Pfanenstiel incision was made. An enormous solid cystic mass exceeding 30 cm of long axis and reaching the right hypochondrium. It was arising from the left ovary, being close to the left fallopian tube, and without adhering to any abdominal viscera. There were no visible cystic formations or lymphadenopathy. The extraction of the tumor entirely was impossible, and only the liquid component was aspirated (about 3 l). A left adnexectomy and peritoneal cytology were made with an extemporaneous examination that concluded to a macroscopically mature teratoma (Fig. 2) We were reassured and confided in this surgical gesture with meticulous control of the hemostasis without any transfusion. Her postoperative was uneventful. She was discharged to home on the second day. No adjuvant chemoradiotherapy was suggested until waiting for the final report from Histopathology. It came after six weeks, revealing a mature teratoma associated with a large undifferentiated high-grade sarcomatous component expressing vimentin (Fig. 3). Unfortunately, the patient was lost and trial to contact her failed. She came to OPD two months after the surgery complaining of an abdominal distention with an altered general condition. A full-body CT scan was done, showing multiple metastases (peritoneal carcinosis, nodules in the liver and lungs). The patient refused to be treated and died within a week of diagnosis.

4. Discussion

Degeneration is an uncommon side evolution of mature teratomas (3 %). These tumors' origin has not yet been identified. According to Linder et al., the abnormality that gives rise to the teratomas results from the initial meiotic division in a single, isolated germ cell [5]. This initial theory was confirmed by Ohama et al., who also demonstrated that for some tumors, the abnormality could develop later, during the second meiotic division [6]. They affect mainly young age groups with an average age of 19 years [7] and account for 20 % of malignant ovarian tumors and 1 % of all malignancies [8,9].This was the case in our 24-year-old patient.

The symptoms can vary from a basic cycle disorder and gravity-type pelvic pain to an abdomino-pelvic mass, occasionally they might be accompanied by disclosing complications including compression, torsion, bleeding, rupture, infection, or ascites. They may also present with a normal physical examination. Our patient presented with a pelvic pain, metrorrhagia, and a huge mass extending beyond the umbilicus.

Although useless in diagnosing malignant component, ultrasound remains a valuable tool for the positive diagnosis of mature teratomas [10]. Admittedly, MRI and CT scans are more accurate. On CT, for example, immature teratomas typically appear as large irregular tumors with heterogeneous aspects and are rarely cystic [11].

On MRI, they appear as a bulky tissue section with some scattered and fatty spots being hyperintense on T1. Several microcysts can be identified also in the mass.

In our patient, ultrasound revealed a multilocular cystic picture with a normal-sized uterus. MRI showed a significant abdominopelvic mass with a thick wall in the left ovarian. It was multiloculated with heterogeneous signal mainly cystic with some fat-liquid levels, associated with multiple thick vegetations and septa, enhancing after gadolinium injection with a signal of high diffusion. No invasion of the surrounding abdominopelvic structures laterally and posteriorly or any other sign of malignancy was reported and our patient was managed as a mature teratoma.

Studies have shown that radiological examinations are unable to determine the histological grade of the immature teratoma of the ovary as it contains tissues derived from the three embryonic celllines (the mesoderm, the endoderm, and the ectoderm) at different stages of maturation [1]. The level of immaturity is highly correlated to the degree of malignancy. In our case, the final histological result showed a tumor with necrotic growth, unusual nuclei and a high mitotic index. Vimentin, calretinin, and PS100 were all expressed by the cells of the tumor but the epithelial markers were not present. An increased blood level of more than 400 ng of alpha-fetoprotein is considered as a risk factor for a negative development and was found in 18 to 45 % of cases [12,13]. The presence of yolk components within the young teratoma has been documented as the cause of this aberrant alpha-fetoprotein

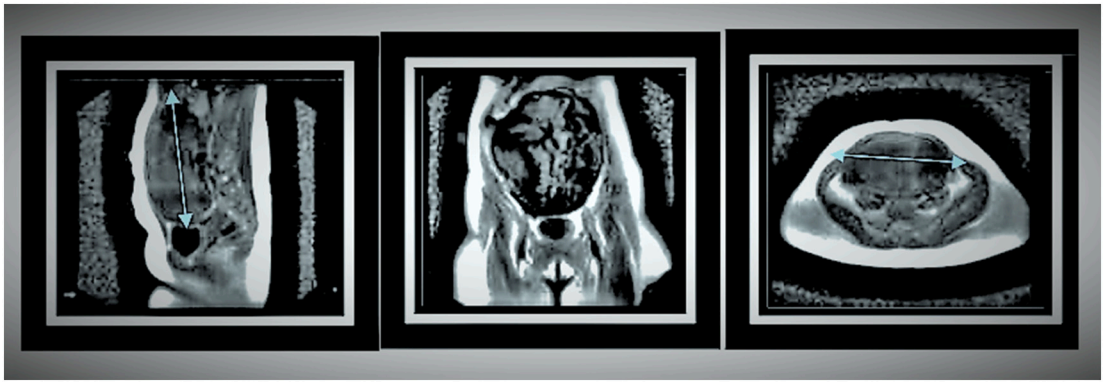

Fig. 1. MRI aspect of tumor: a large well defined abdominopelvic mass measuring 24 cm of long axis (↔); a = longitudinal section/ b = coronal section/ c = transverse section.

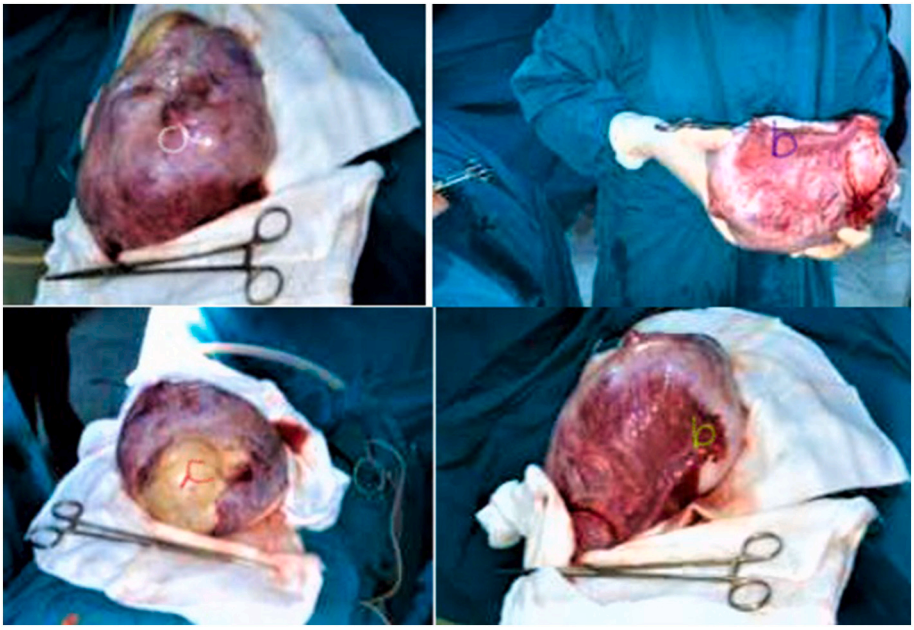

Fig. 2. The intra-operative aspect of the left adnexectomy: a/ ovarian mass; b/ fallopian tube; c/ ovarian parenchyma.

release. In our case, the level of alpha-foetoprotein was normal.

According to the French Cancer Society in 2013, depending on the histological grade, the treatment of immature teratomas is divided into two phases: surgery followed by chemotherapy. Surgery is always conservative and has two goals: removal of the tumor, determining the tumor's extent by histology [1]. A unilateral adnexectomy is usually advised, associated to a thorough abdominal cavity exploration, a peritoneal cytology and/or ascites removal, repeated systematic peritoneal biopsies including those at the omentum, and removal of any suspicious elements. In the absence of a lymphnode lesion, systematic pelvic and lumbaraortic lymphnode dissection is not recommended. There is no need for a systematic bilateral adnexectomy but a careful examination and, in some cases, a biopsy of the contralateral ovary are advised. There is no evidence of usefulness of performing a hysterectomy [14,15].

To maintain fertility, this conservative treatment is often used. An exploratory laparotomy and a left adnexectomy with peritoneal cytology were performed in our patient. An ovarian teratoma associated to an undifferentiated high-grade sarcoma was identified by final histopathology report.

The larger omentum and other organs, such as the liver, can occasionally be affected by the immature teratomas' rapid growth. A peritoneal invasion and metastatic lesions can be noted mainly in the abdominopelvic peritoneum [1].

These metastases usually occur with immature forms. Our patient who was lost after intial surgical treatment progressed rapidly: Numerous metastases were found, and the patient refused to be treated and died within a week of diagnosis. Li et al. Claimed that improvements in chemotherapy and the use of platinum salts made the prognosis of immature teratomas better. Before 1983, the survival rate was 40 %, and between 1994 and 1998, it was 95 % [12]. The histological grade of the underlying tumor and the prognosis of immature teratomas are closely correlated [16]. Grade 1 tumors have a favorable prognosis and the five-year survival rate is reported by the literature to be between 81 and 94 % [12,16]. However, Grade 3 immature teratomas have a high tendency for malignancy with a high rate of mortality, as was the case with our patient [12,16,17]. Chemotherapy has improved the 5-year survival rate of these malignancies from 90 % to 100 % [18].

5. Conclusion

Young women are most often affected by immature teratomas. The prognosis is correlated to histological grade. The radiological examination helps to suspect the diagnosis and istological examination confirms it. Multidisciplinary tmanagement involving oncologists, gynecologists, and pathologists is necessary. Although standard treatment tends to be conservative. Adjuvant chemotherapy can be suggested for high-grade malignancies.

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Ethical approval

Ethical rules were respected such as anonymous with no patient information or means of identifying.

The consent statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of the journal on request.

Research registration/guarantor

None.

CRediT authorship contribution statement

Karima Mekni: **Corresponding author** Writing- Reviewing and Editing.

Mona Mlika: Data curation, Writing.

Yoldiz Houcine: Data curation, Writing.

Chiraz ElFekih**:** Supervision

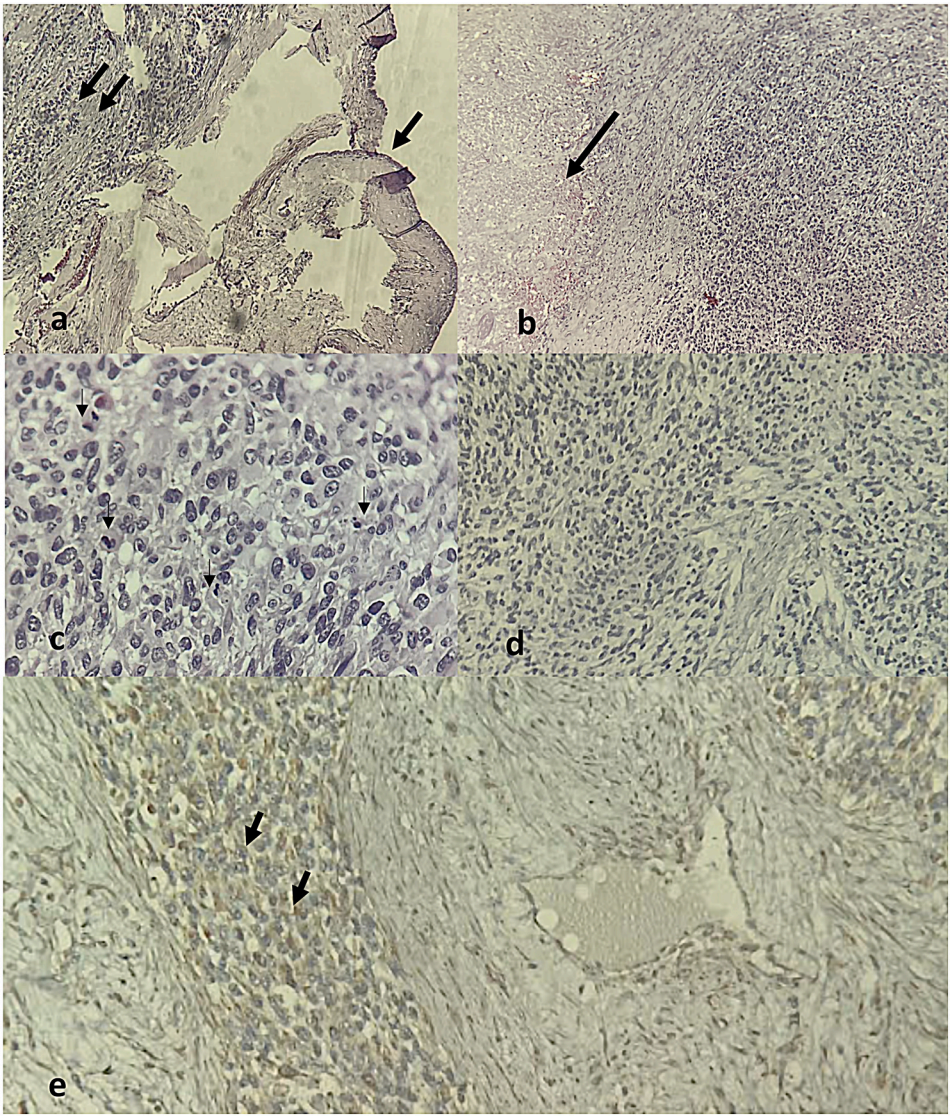

Fig. 3. a/ Cystic mass covered by a respiratory layer (arrow) with the presence of a spindle cell tumor within the wall (double arrow) (HEx250), b/ undifferentiated spindle cell tumor with large necrotic areas (arrow) (HEx250), c/ The tumor cells are pleomorphic with numerous mitotic figures (Arrow) (HEx400), d/ Negative immunostaining of the tumor cells with the cytokeratin, EMA, CD30, WT1, Calrétinine, Smooth muscle actin, Desmin, Myogenin, PS-100 antibodies (x250), e/ Positive immunostaining of the tumor cell with the vimentin antibody (x400).

Declaration of competing interest

The authors declare that they have no competing interests.

References

[1] W. Bouzoubaa, S. Jayi, Alaoui FZF, H. Chaara, M.A. Melhouf, Tératome immature de l'ovaire: à propos d'un cas., Disponible sur: Pan. Afr. Med J 27 (263) (2017) https://www.panafrican-med-journal.com/content/article/27/263/full.

[2] S. Jorge, N.L. Jones, L. Chen, J.Y. Hou, A.I. Tergas, W.M. Burke, et al., Characteristics, treatment and outcomes of women with immature ovarian teratoma, 1998–2012, Disponible sur: Gynecol Oncol 142 (2) (2016) 261–266 https://linkinghub.elsevier.com/retrieve/pii/S0090825816307533.

[3] J. Łuczak, M. Bagłaj, Ovarian teratoma in children: a plea for collaborative clinical study, J. Ovarian Res. 11 (1) (2018) 75, https://doi.org/10.1186/s13048-018-0448-2.

[4] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. Lond. Engl. 84 (2020) 226–230.

[5] Y. Rezk, J. Sheinfeld, D.S. Chi, Prolonged survival following salvage surgery for chemorefractory ovarian immature teratoma: a case report and review of the literature, Gynecol Oncol. 96 (3) (2005) 883–887.

[6] X. Wu, L.Y. Han, X. Xu, Z. Li, Recurrent immature teratoma of the ovary: a case report of radical secondary cytoreduction with replacement of the aortic bifurcation, Gynecol. Oncol. 95 (3) (2004) 746–749.

[7] F.F. Nogales, B.E. Favara, F.J. Major, S.G. Silverberg, Immature teratoma of the ovary with a neural component ("solid" teratoma). A clinicopathologic study of 20 cases, Hum. Pathol. 7 (6) (1976) 625–642.

[8] R.J. Kurman, H.J. Norris, Malignant germ cell tumors of the ovary, Hum Pathol. 8 (5) (1977) 551–564.

[9] R. Vajpeyi, WHO classification of tumours: pathology and genetics of tumours of the breast and female genital organs, Disponible sur: J. Clin. Pathol. 58 (6) (2005) 671–672 https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1770678/.

[10] Cancérisation d'un kyste dermoide de l'ovaire: à propos d'un cas [Internet], Disponible sur: http://www.research-journal.net/fr/Carcinogenesis-of-a-dermoid-ovarian-cyst.html, 2022.

[11] E.K. Outwater, E.S. Siegelman, J.L. Hunt, Ovarian teratomas: tumor types and imaging characteristics, Radiogr. Rev. Publ. Radiol. Soc. N. Am. Inc. 21 (2) (2001) 475–490.

[12] M. Kawata, S. Sekiya, R. Hatakeyama, H. Takamizawa, Neuron-specific enolase as a serum marker for immature teratoma and dysgerminoma, Gynecol Oncol. 32 (2) (1989) 191–197.

[13] Glioblastome péritonéal: récidive d'un tératome immature de l'ovaire (à propos d'un cas) - EM consulte, Disponible sur: https://www.em-consulte.com/article/88561/glioblastome-peritoneal-recidive-d-un-teratome-imm, 2022.

[14] B. Cushing, R. Giller, A. Ablin, L. Cohen, J. Cullen, E. Hawkins, et al., Surgical resection alone is effective treatment for ovarian immature teratoma in children and adolescents: a report of the pediatric oncology group and the children's cancer group, Am. J. Obstet. Gynecol. 181 (2) (1999) 353–358.

[15] MS1782021.pdf, Disponible sur: http://ao.um5.ac.ma/xmlui/bitstream/handle/123456789/19329/MS1782021.pdf?sequence=1&isAllowed=y, 2022.

[16] J.L.D. Trombelin, P. Deruelle, Évaluation de la prise en charge des tératomes immatures de l'ovaire: à propos de 3 cas. undefined [Internet] [cité 13 oct 2022]; Disponible sur: https://www.semanticscholar.org/paper/%C3%89valuation-de-la

-prise-en-charge-des-t%C3%A9ratomes-de-%C3%A0-trombelin-Deruelle/765702150105146cfe17b41a8b9180d172c6e5de, 2006.
[17] K. Sait, C. Simpson, Ovarian teratoma diagnosis and management: case presentations, J. Obstet. Gynecol. Can. JOGC 26 (2) (2004) 137–142.
[18] M.E. Beiner, W.H. Gotlieb, Y. Korach, A. Shrim, D. Stockheim, Y. Segal, et al., Cystectomy for immature teratoma of the ovary, Gynecol Oncol. 93 (2) (2004) 381–384.