Case report

Immature Teratoma of the Ovary: A Case Report and LiteratureReview

Abstract:

Immature teratomas are rare malignanttumorsrepresentinglessthan one percent of ovarianteratomas, occurring in youngwomen and adolescents. Theirdevelopment arises fromcellsderivedfrom the threeembryonicgermlayers (endoderm, mesoderm, and ectoderm). They are classifiedintothree grades of increasingseveritybased on the proportion of immature neural cells. The prognosisisdirectlycorrelatedwith the histological grade. The therapeutic management isprimarilysurgical, possiblycombinedwith adjuvant chemotherapy, whilemaximizing the preservation of fertilitypotential.

Keywords :Immature teratoma; Ovary;Tumor; Chemotherapy; Surgery

Introduction :

Immature teratomais a non-seminomatousgermcelltumor first describedin 1960 by Thürlbeck and Scully[1]. Immature teratomas are very rare, accounting for lessthan one percent of malignantovariantumors. Their classification intothree grades of malignancywasinitiallyproposed by Thurlbeck and Scully[1] and modifiedin 1976 by Norris and O'Connor to betterdefinetherapeutic indications[2]. However, the management of immature teratomaremainscontroversial due to the rarity of thesetumors, whichlimitssufficientstudiescomparingdifferenttherapies. We report the case of a 17-year-old patient admitted to the gynecological and breast unit of the National Institute of Oncology CHU-Ibn Sina for the management of an abdominopelvic mass. Sheunderwent an abdominopelvic CT scan and pelvic MRI, followed by surgicaltreatmentinvolving a right adnexectomy and several biopsies. The histologicalexaminationconfirmed an immature ovarianteratoma. The aim of thisstudyis to highlight the diagnostic challenges, the evolution of this type of ovariantumor, and to discuss management modalities.

Observation :

A 17-year-old patient with no medical or surgicalhistory, unmarried,

wasadmitted to ourhospital for the management of pelvic pain in the context of a general state alteration. Examinationrevealed a round, non-tender abdominopelvic mass reaching the umbilicus. The pelvic CT scan showed a complexcysticabdominopelvic mass containing a non-enhancing tissue component aftercontrast injection, calcification, and a fattycompartment, measuring 109 x 104 x 143 mm in itslargest dimensions (Figure 1). MRI revealed a large mixed abdominopelvic mass containing a fleshy portion, roughlyoval in shape, withlobulated contours, surrounded by a capsule withseptations, measuringapproximately 144 x 117 x 190 mm (Figure 2). The CA 125 levelwaselevated at 78.9 UI/ml (0–35 U/ml); BHCG wasnegative; LDH was 162 U/L (125–220 U/L); and AFP was 5.29 ng/ml (0.89–8.78 ng/ml). The patient underwentlaparotomy, whichrevealed a large ovarian mass on the right ovary, leading to right adnexectomy, peritonealcytology, peritonectomy, omentectomy, and removal of a polypfrom the Douglas pouch (Figure 3). The pathologicalresultsconfirmed a grade 2 immature teratoma of the ovary and a secondarylocalization in the Douglas pouchThe omentectomy and peritonectomydid not containany cancer cells. Peritonealfluidcytologywasnegative. The case waspresented to the multidisciplinaryoncology team, and the decision for adjuvant chemotherapy and oocyte cryopreservationwas made.

Discussion:

Immature teratoma of the ovaryis a malignanttumorcomposed of tissues derivedfrom the threeembryoniccelllineages (mesoderm, endoderm, and ectoderm) present at various stages of maturation within the tumor [5]. Immature teratomaspreferentially affect youngwomen in their second decade of life;however, cases have been described in the literatureacross all ages, includingpostmenopausalwomen[3,4]. In our observation, the patient was 17 yearsold. Immature teratomarepresents 3% of teratomas, 1% of all ovarian cancers, and 20% of malignantgermcelltumors of the ovary [5]. Typically, immature teratomaspresent as a rapidlyenlargingabdominopelvic mass associatedwithincreased abdominal volume and a sensation of pelvicheaviness, as seen in our case. There isusually no alteration in generalhealth or menstrual cycle disturbances. In our observation, however, therewas a generalhealthalterationwithsignificantweightloss. It can alsobediscovered as a complication related to tumorvolume: torsion or compression of neighboringorgans [6]. This tumorisgenerallyunilateral, large, rounded, unilobulated or multilobulated, and has a mixed liquid and solid component [7].

The contralateralovarypresents a mature teratomain 26% of cases and another immature teratomain 10% of cases[8].

In imaging, immature teratomais a large tumor (12—25 cm) [9]. The ultrasoundappearance of immature teratomasisnonspecific [10]. CT and MRI images are more characteristic. Generally, on the scan, immature teratomas are rarelycystic and present as a large, irregulartumor of mixed tissue and fattyappearance, with the solid part consisting of numerouscoarse and amorphous calcifications associatedwithscatteredfattyislands and some rare microcysts [11,12]. In our case, the CT showed a complexcysticabdominopelvic mass containing a non-enhancing tissue component, calcifications, and a fattycompartment, while MRI highlighted the presence of a large mixed abdominopelvic mass containing a fleshy portion withlobulated contours and a capsule emittingseptations. Imaging plays an important role in orienting the diagnosis, but the anatomical-pathologicalstudyconfirms the diagnosis.

The immature teratoma of the ovaryconsists of a mixture of mature and immature tissue in varying proportions, and the amount of immature neural tissue allows for classification intothree grades of increasingmalignancy. This classification wasinitiallyproposed by Thurlbeck and Scully and modifiedin 1976 by Norris and O'Connor to betterdefinetherapeutic indications[1,2] (Table 1). Currently, this classification tends to besimplifiedintolow and high grade [12]. Tumorswith a betterprognosis are grade 1 tumors, for which the five-yearsurvival rate isestimatedbetween 81 and 94% [6,13,14]. Grade 3 immature teratomas have a highlymalignantpotential and theirrapidevolution, bothlocally and distally, results in higher rates of recurrence and mortality [6,13,14].

The measurement of serumtumor markers iscommon in the discovery of an ovariantumor. Tumor markers can assist in diagnosis, prognosisevaluation, and post-therapeutic follow-up. A nonspecificelevation of CA 125 can befound in cases of immature teratoma, as in manypelvic pathologies[15]. Alpha-fetoproteinmaybeelevatedin 18 to 45% of cases[15,16]. A serumlevelabove 400 ngisconsidered a risk factor for poor progression. Lactate dehydrogenase (LDH), oftenused as a tumor marker to evaluatetumorburden in malignantgermcelltumors, can reflectrapidcellproliferation and high metabolismwithin the tumor. This maybeobserved in immature teratomas, although LDH islessspecificthan AFP. CA 19.9 isalsobeingevaluated, and very high levels have been found in some cases of immature teratomas, but the role of this marker isstill to bedetermined [6]. Human chorionicgonadotropin (β-

hCG) can alsobeslightlyelevated in some immature teratomasthatcontaintrophoblasticelements [15,16]. In our patient, CA 125 waselevated at 78.9 UI/ml, and the othertumor markers werenegative.

In terms of treatment, according to the French Society of Oncologyin 2013, the management of immature teratomasisdividedintotwoparts:surgicaltreatmentfollowed by chemotherapydepending on the histological grade. Surgicaltreatmentgenerallyconstitutes the first therapeuticstep, involvingunilateralovariectomy or initial adnexectomythrough a midlinelaparotomy, allowing for simultaneous exploration of the abdominal cavitywithperitonealfluid collection for cytologicalstudy and systematic multiple peritoneal biopsies, including at the epiploon and collection of anysuspiciouselement. Systematicpelvic and lumbar-aorticlymphadenectomyis not recommended in the absence of lymphnodeabnormalities. A samplewillbetaken if an anomalyisdetected by CT or duringsurgical exploration [17,18]. A meticulous inspection of the contralateralovaryisnecessarysince the contralateralovarypresents a mature teratomain 26% of cases and another immature teratomain 10% of cases[8], althoughsystematicbilateraladnexectomyis not indicated. However, itis certain that more aggressivetreatmentsuch as hysterectomy and contralateraladnexectomy, whichwoulddefinitivelyeliminateanypossibility of future pregnancy, isunnecessary [19]. The coloscopicapproachisrarelyused due to the size of the tumor;treatment must takeintoaccount the preservation of fertility, as immature teratoma of the ovaryisgenerally a tumor of youngwomen.

In our observation, weopted for conservative treatment to preservefertility in our 17-year-old patient, withperitoneal biopsies. Weperformedlaparotomy, whichrevealed a large ovarian mass on the right ovary, leading to right adnexectomy, peritonealcytology, peritonectomy, omentectomy, and removal of a polypfrom the Douglas pouch. The pathologicalstudyconfirmed a grade 2 immature teratoma of the ovary and a secondarylocalization in the Douglas pouch.

Adjuvant chemotherapyisdetermined by the histological grade. For grade 1 tumors, clinical follow-up after conservative surgerysuffices, with no long-termrecurrenceobservedaccording to a study by Carinelli on 245 cases of immature ovarianteratomasafterunilateraladnexectomy [20]. Adjuvant chemotherapyisusuallyreserved for grade 3 tumors or even grade 2 tumors. The

mostcommonlyusedprotocolincludesthree agents borrowedfromtesticular cancer chemotherapy: BEP, consisting of bleomycin, etoposide, and a platinumsalt (cisplatin) [18,20,21]. For our patient, during the multidisciplinary meeting and considering the histological grade and the location on the polypremovedfrom the Douglas pouch, adjuvant chemotherapywasindicatedaccording to the BEP protocolwith oocyte cryopreservation. The cryopreservationwas not performed due to the patient'slack of means.

A recentstudy by Cushing et al. contradicts the necessity of chemotherapy, showingthatsurgeryaloneprovides a curative treatment in the majority of cases, regardless of the initial tumor grade, with a four-yearsurvival rate of 100% and 97.7% of patients withoutrecurrence. The authorsconcludethatchemotherapyshouldbereserved for post-surgicalrecurrences to avoid the adverse effects of chemotherapy in patients who are oftenveryyoung[17,18].

Conclusion:

Immature teratoma of the ovaryis a malignanttumorwhoseseverity and prognosisdependdirectly on the histological grade, indicating the tumor'saggressiveness. It primarily affects youngwomen, and diagnosisisusuallysuspectedthroughradiologicalexaminationsbeforebeingconfirmed by pathologicalexamination. The recommendedtreatmentfavors a conservative surgicalapproach to preservefertility as much as possible. Depending on the tumor grade, chemotherapymaybenecessary to completetreatment.

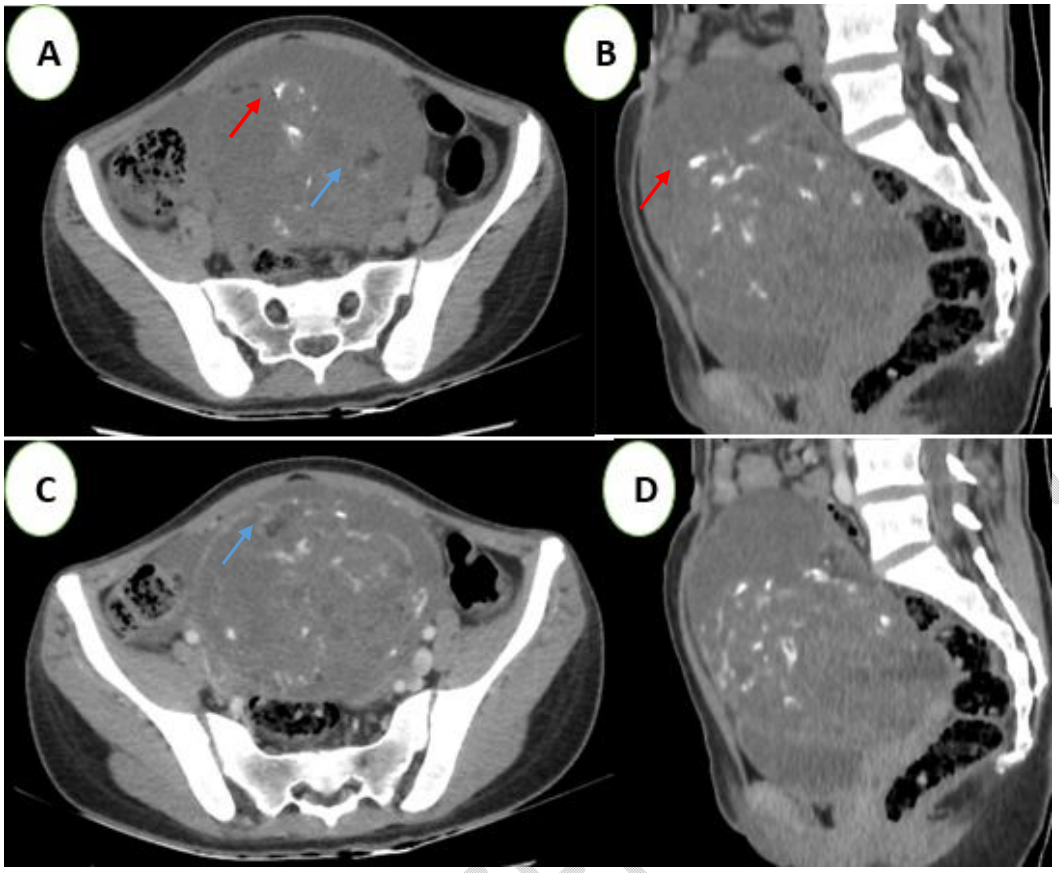

Figure 1 : Abdominopelvic CT Scan without (A and B) and withcontrast injection (C and D), in axial (A and C) and sagittal (B and D) slices: A large heterogeneousabdominopelvic mass, predominantlyfluid-filled and non-enhancingaftercontrast injection, containingfatty areas (bluearrow) and calcifications (redarrow) consistent with a teratoma.

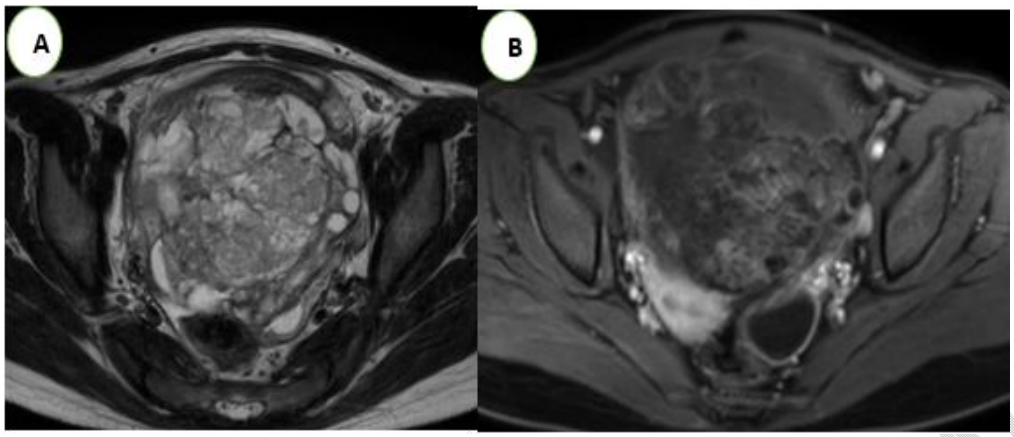

Figure 2 Pelvic MRI T2 axial sequences (A) and T1 FATSAT with gadolinium injection (B) of the same patient confirming the ovarianorigin of the teratoma**.**

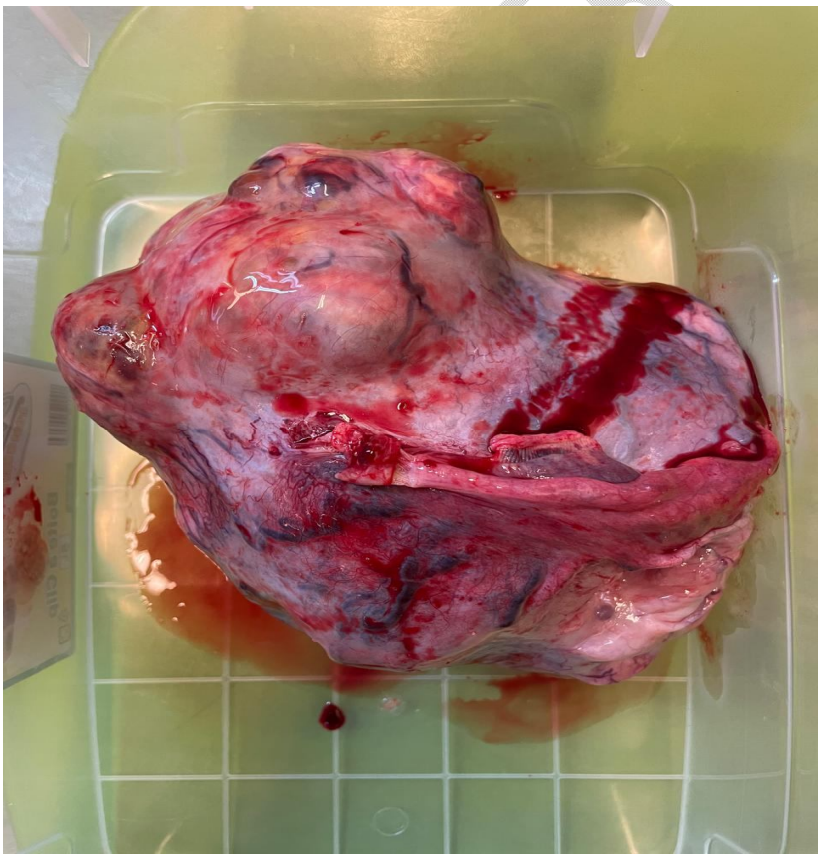

Figure 3:Operativespecimen of an immature teratomafromour patient.

TABLE 1 : Grading of immature ovarianteratomas by Norris and O'Connor (in 3 grades)

Grade 0	Fully mature tissue with rare mitoticactivity.
Grade 1	Tumorcontaining rare areas of immature neuroepithelial tissue occupyinglessthan one field per slide at 40x magnification.
Grade 2	Tumorcontaining rare areas of immature neuroepithelial tissue occupying 1 to 3 fields per slide at 40x magnification.
Grade 3	Tumorcontaining large areas of immature neuroepithelial tissue occupying more than 3 fields per slide at 40x magnification.

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