Thake J. et al. (2023). *Xjenza Online*, 11(1-2):49–52.

Xjenza Online: Science Journal of the Malta Chamber of Scientists
www.xjenza.org
DOI: 10.7423/XJENZA.2023.2.05

Research Report

Immature High Grade Teratoma in A 17-Year-Old Lady—a case report

J. Thake[1], **C. Zerafa**[1] **and S. Diacono**[*1]
[1] *Independent researcher, Msida, Malta*

Abstract. Immature teratomas are uncommon, malignant ovarian neoplasms that make up less than 1% of ovarian germ cell tumours. Immature teratomas are often larger and occur in younger women (in the first three decades of life). This case report discusses a 17-year-old lady was admitted to the emergency department in view of dyspnea, severe abdominal pain, abdominal distension and loss of appetite. On CTPA she had a large pelvic mass, and was referred to gynaecology. She underwent an elective right salpingo oophorectomy and cystectomy. Histology report indicated ovarian immature teratoma (high grade, grade 3) showing extensive neuroectodermal differentiation. The incidence of immature teratoma is highest in young adults. Most patients present with early-stage disease, are managed with fertility sparing surgery and chemotherapy with an excellent prognosis. Diagnosis requires a combination of clinical, radiological and laboratory findings.

Keywords: Ovarian neoplasms, immature teratoma, biomarkers, younger women.

1 Introduction

Immature teratomas are uncommon, malignant ovarian neoplasms that make up less than 1% of ovarian germ cell tumours. Immature and embryonic tissue from all three cell lines may be present within immature teratomas, and primitive neuroectodermal tissue is the most common malignant tissue subtype. Compared with mature teratomas, immature teratomas on average are larger, occur in younger women (in the first three decades of life), and show more solid components on imaging. The presence of small amounts of fat within an aggressive ovarian mass can suggest the diagnosis of malignant immature teratoma (Siegelman, 2005).

2 Case presentation

A 17-year-old lady presented to the emergency department with dyspnea, severe abdominal pain, abdominal distension and loss of appetite. Patient had no previous medical or surgical history, and was not taking any treatment. On admission she had a CT pulmonary angiogram (CTPA) done in view of dyspnea which showed no evidence of pulmonary embolism, however this showed large heterogeneous pelvic mass likely ovarian in origin causing right obstructive uropathy.

The CTPA report commented on a large mixed solid cystic mass (as seen in figure 1) arising from the pelvis measuring 32.4 cm (cc) × 13.3 cm (ap) × 25 cm (trans) with interspersed globules of fat and calcific components. The CT also noted that the ovaries were not distinguishable and fat plane with the uterus was also not respected. Due to the mass effect, there was resultant delayed nephrogram of the right kidneys with mild hydronephrosis in keeping with obstructive uropathy. The CT report noted that moderate ascites together with the size of the lesion suggest malignant nature and high risk of torsion. No aggressive bone lesions noted. Patient was referred urgently to gynecology. On examination patient had striae on her abdomen, soft abdominal distention otherwise unremarkable examination.

Tumour markers and routine blood tests were taken. She had elevated cancer antigen (CA125), Alpha-fetoprotein (AFP) and lactate dehydrogenase (LDH).

In view of the mild hydronephrosis present and the high blood pressure readings throughout the admission, the case was discussed with urologists and endocrinologists. She was started on amlodipine 5 mg daily and was referred to medical outpatients for further endocrinological tests. Urologists deemed necessary a bilateral Pollock catheter insertion, which was done during surgery. Urgent stent-

Correspondence to: S. Diacono (sheriseaned@hotmail.com)

ing was not needed in view of normal creatinine, stable parameters, as well as lack of pain and urinary symptoms.

She underwent an elective right salpingo oophorectomy and cystectomy.

Histology report indicated ovarian immature teratoma (high grade, grade 3) showing extensive neuroectodermal differentiation, 275 mm in the greatest dimension (pT1c pNx TNM classification of malignant tumours, 8th edition).

Post-operatively she was admitted to a high dependency unit (HDU) for further observation. She had sluggish bowel sounds thus remained nil by mouth and the Pollock catheter was kept in situ. She was prescribed ciprofloxacin 200 mg twice daily and Gentamycin 400 mg daily for a total of 9 days. Post-operatively the patient complained of right sided chest pain, and on CTPA there was bilateral features of pulmonary embolism involving the lower lobar segment of pulmonary arteries. Pneumoperitoneum was also noted on the report, in view of recent surgery. She was thus started on a therapeutic dose of clexane 80 mg twice daily.

She was transferred to the gynecology ward, started on a light diet and the catheters were removed. Her case was discussed with the oncologist and CT brain was suggested.

Non-contrast and contrast enhanced CT scan of the brain report showed preserved grey-white matter differentiation. No intra or extra axial haemorrhage or collections. No space occupying lesions, mass effect or midline shift. No aggressive bone lesions. In conclusion there were no acute intra-cranial pathology demonstrated and no evidence of metastatic disease.

Her discharge plan was a renal DTPA scan in 6 weeks (to assess kidney function), gynecology outpatients follow up in 6 weeks with ultrasound. She was discharged on Rivaroxaban 15 mg twice daily for 21 days and 20 mg daily for 6 months.

Renal scintigraphy commented on good percentage uptake of both kidneys and no significant tracer retention, in conclusion there was no scan evidence of complete obstruction as seen in figure 2.

She was followed up by oncology with chemotherapy and monitoring of tumour markers. CT liver was done to exclude metastasis. The report commented on a left adnexal cystic lesion measuring 4.8 (TR) × 4.4 (CC) × 4.1 (AP) cm with a tubular appearance, possibly dilated fluid-filled fallopian tube (hydrosalpinx). Note was also made of mild fat-stranding in the omentum anterior to ascending colon and adjacent to umbilicus likely omental infarction secondary to recent surgery.

Note was also made of a collection in the posterior pelvis (approx. 8 cm × 3 cm) which appeared mildly complex with areas of higher density represent a postop pelvic haematoma or residual disease. A repeat scan 4-6 weeks postop was advised for follow-up and reassessment, and to allow time for postop changes to resolve. Review of the images in bone window settings revealed no abnormalities. The report commented on minor atelectasis right lower lobe. Otherwise, the lung bases show normal appearances. In conclusion no hepatic metastasis demonstrated.

A repeat scan was done which commented on enlarge axillary lymph nodes on the left up to 1.6 cm. There was a small amount of residual pelvic fluid. There was fat stranding in the area of surgery compatible with postop changes.

Ultrasound-guided biopsy was performed using an 18-G core biopsy system, in view of multiple enlarged left axillary lymph nodes described on CT. The core biopsies showed well preserved lymph node parenchyma exhibiting reactive follicular hyperplasia in places. The Ki67 index was in keeping with a reactive lymph node. There was no expression of pancytokeratins (AE1/AE3) or of synaptophysin (performed on both tissue blocks) and there was no evidence of metastatic tumour in these biopsies. In conclusion there is lymph node parenchyma with no evidence of metastatic tumour.

3 Discussion

Immature teratoma is a germ cell tumour composed of tissues that can be traced to the three embryonic germ layers, with at least one of the them lacking full differentiation (Medeiros et al., 2018). Immature teratoma is a rare tumour and is of unknown aetiology (Busca et al., 2020). It makes up 1% of all teratomas, 1% of all ovarian cancers, and 35.6% of malignant ovarian germ cell tumours (Busca et al., 2020; Medeiros et al., 2018). Immature teratoma is the second most common type of ovarian malignant germ cell tumor. In the ovary, mature teratomas are considered benign, and immature teratomas are considered malignant (Medeiros et al., 2018). Immature teratoma of the ovary is almost always unilateral (Alwazzan et al., 2015).

Occurring primarily during the first 2 decades of life, with 26 years of age being the mean age of presentation (Alwazzan et al., 2015; Busca et al., 2020). The most common presenting symptom is abdominal distention (81%) (Alwazzan et al., 2015). Often associated with a rapidly growing painful abdominal mass. Torsion can also be part of the presentation (Busca et al., 2020).

Diagnosis requires a combination of clinical, radiological and laboratory findings. If a young female presents with rapidly enlarging adnexal mass, with mildly elevated AFP and typical radiological findings, this raises high suspicion

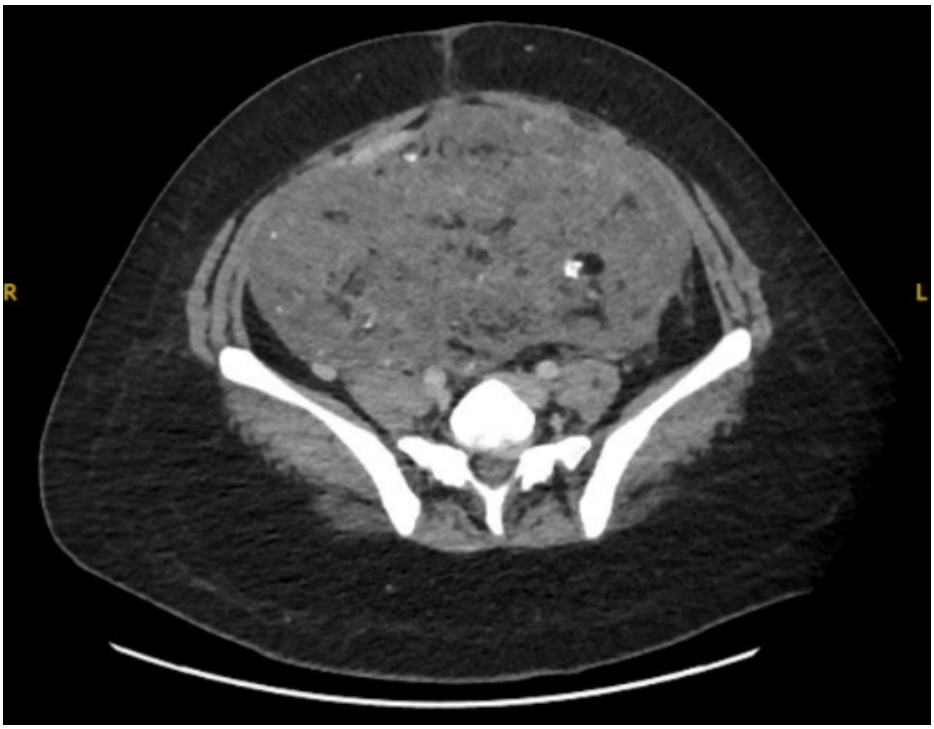

Figure 1: CT scan showing large mixed solid cystic mass within the abdomen.

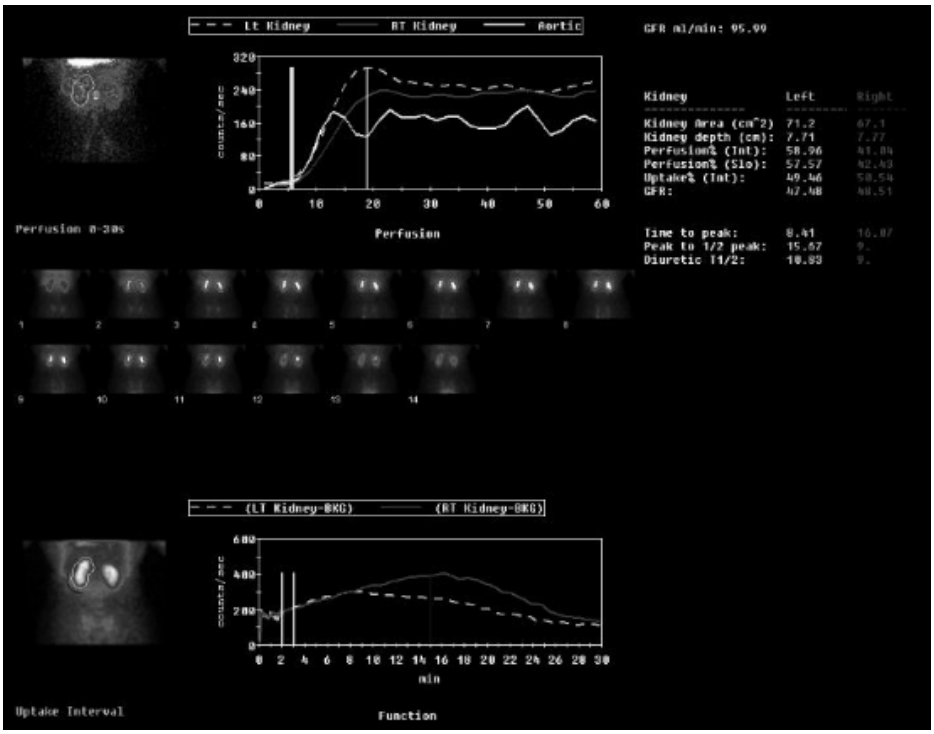

Figure 2: Renal scintigraphy of right and left kidney showing good percentage uptake of both kidneys and no significant tracer retention.

of immature teratoma. To confirm the diagnosis as well as for grading, histological examination is required (Busca et al., 2020). On CT imaging, the solid component appears large and irregular, with coarse calcifications and small foci of fat scattered throughout (Choudhary et al., 2009).

Different tumor markers are elevated depending on the component tissues. Elevated serum tumor markers, can aid in initial diagnosis, therapy monitoring, and post-treatment surveillance. For the screening of malignant germ cell tumors, CA125 is useful. CA19-9 is high in immature teratoma, and mature cystic teratoma with malignant transformation. Dysgerminoma and yolk sac tumor have high levels of LDH (Kawai et al., 1992; Shaaban et al., 2014).

Elevated α-fetoprotein or β-hCG levels are diagnostic of ovarian malignant germ cell tumors and should be measured in all young women who present with a pelvic mass (Kawai et al., 1992; Shaaban et al., 2014). Alpha-fetoprotein is found in all patients with a tumor that contains components of a yolk sac tumor. Serum α-fetoprotein concentrations are significantly elevated at diagnosis in most ovarian immature teratomas with foci of yolk sac tumor. Mixed germ cell tumors may secrete β-hCG, α-fetoprotein, or both, depending on the components (Kawai et al., 1992; Shaaban et al., 2014).

Prognosis improves overall after chemotherapy. Older age at diagnosis, advanced stage and high grade histology confers worse prognosis (Jorge et al., 2016). Adjuvant chemotherapy has improved overall survival after surgery (Alwazzan et al., 2015). Grade is the most important risk factor for relapse (Busca et al., 2020).

Patients with stage I, grade 1 tumor may be treated through surveillance, whilst for grade 2 or 3 lesions, surgery and chemotherapy is done (Busca et al., 2020). Most of the patients present with early-stage disease, and are managed with fertility sparing surgery and chemotherapy. These have an excellent prognosis (Jorge et al., 2016).

4 Conclusion

The incidence of immature teratoma is highest in young adults aged 18 to 39. Most patients present with early-stage disease, are managed with fertility sparing surgery and chemotherapy and have an excellent prognosis. Later age at diagnosis, advanced stage, and high-grade histology confer a worse prognosis. Diagnosis requires a combination of clinical, radiological and laboratory findings (Jorge et al., 2016).

5 Acknowledgement

The author would like to thank the patient and her family for their kind informed consent in publishing of this case. Mr John Thake who is the caring consultant and Ms Christine Zammit Zerafa Senior registrar within the firm. Mater Dei hospital for kindly providing the necessary approvals to be able to carry out this case report.

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