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Case Report

A pure and bizarre ovarian malignancy - choriocarcinoma: case report

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ABSTRACT

Germ cell malignancies represent 15% of ovarian cancers in Asians. Choriocarcinoma is a malignant tumor of the ovary with trophoblastic differentiation can be gestational or non-gestational in origin the study is aimed to increase awareness of rare malignant cancer in children that can present in uncommon symptoms and is diagnosed only if the doctor is vigilant. It is a prospective observational case report. Though uncommon a cancer, choriocarcinoma cases have been reported several times in the literature with no definitive treatment plan. The neoadjuvant chemotherapy followed by surgery has proven too successful in the above case. A multidisciplinary approach to juvenile cancer can have a fruitful outcome with high cure rates. The rarity of this cancer makes it difficult to have clear cut diagnosis fixed treatment plan. A good history taking and easy availability of ultrasound with fast-track referral can save many lives from the deadly disease like cancer. Due to paucity of data in regard to pure choriocarcinoma this case report may help in gathering more attention to the research towards this cancer.

Keywords: Pure choriocarcinoma, Beta human chorionic gonadotropin, Chemotherapy

INTRODUCTION

Germ cell malignancies represent 15% of ovarian cancers in Asians.[1] Choriocarcinoma is a malignant tumor of the ovary with trophoblastic differentiation. Pure non-gestational choriocarcinoma is an extremely rare primary germ cell neoplasm, with reports of only a few cases.[2] It can be a gestational or non-gestational choriocarcinoma in origin and may be a primary tumor or a metastasis from other organs. Non-gestational choriocarcinoma occurs mainly as a component of a mixed germ cell tumor.[3,4] The estimated incidence of gestational ovarian choriocarcinoma is 1 in 369 million pregnancies.[5] Non-gestational ovarian choriocarcinoma account ≤0.6% of all ovarian neoplasms; the pure type is extremely uncommon.[6] Generally, majority of the non-gestational type have been clinically diagnosed in patients who were sexually immature, unable to conceive or had never had sexual intercourse.[3] Non-gestational choriocarcinoma consisting of cytotrophoblastic and syncytiotrophoblastic cells that secretes beta-human chorionic gonadotropin (β-hCG), arises spontaneously, and is primarily located within midline structures such as the retroperitoneum or mediastinum. We hereby present a case report of a pure ovarian choriocarcinoma and discuss the diagnosis and treatment together with a brief review of the literature. Consent was obtained from the father of the patient.

CASE REPORT

A 13-year-old girl presented with acute pain abdomen, fever, and vomiting for one day. The patient had been experiencing on and off pain abdomen and low-grade fever for one week. There was a history of loss of appetite and weight loss of approximately 3-4 kg over past 2 months. She had her menarche 8 months ago; had been having menstruation irregularly every 2-3 months since then. At a local hospital she was diagnosed to have an adnexal mass

of ovarian origin, high β-hCG and anemia and was referred for the same.

On arrival the patient appeared cachexic, had high temperature. Physical examination revealed pallor, body mass index (BMI)- 13 kg/m^2; there was lower abdominal tenderness and a palpable solid mass of 10×8 cm. Ultrasound showed a normal sized uterus with thin endometrium and a complex ovarian mass measuring 10×7.1 cm with irregular cystic areas. The investigation reports were as haemoglobin: 7.6 g/dl (>11 g/dl); β-hCG 192631.0 mlU/ml (<5.0); alpha fetoprotein (AFP) 0.9 ng/ml (<20); carcinoembryonic antigen (CEA) 2.5 ng/ml; lactate dehydrogenase (LDH) 554 IU/l (<220 IU/l). Computed tomography (CT) scan showed a large heterogeneously hypodense lesion measuring ~ 10.7×8.4×10.1 cm (CC×AP×TR) in the pelvis showing peripheral nodular hyper enhancement on post contrast images. Few enhancing incomplete internal septae noted. There was no evidence of metastasis to other organs.

In view of poor functional status of the girl, preoperative neoadjuvant chemotherapy was planned instead of upfront surgery. An ultrasound guided biopsy confirmed choriocarcinoma following which she was started on bleomycin-etoposide-cisplatin (BEP) regimen. The patient did well and tolerated 3 cycles of chemotherapy. Following the first chemotherapy β-hCG was 3930 mIU/ml, second chemotherapy 193.6 mIU/ml third chemotherapy 58.9 and fourth chemotherapy was 23. Following this a decision of Surgery was taken by the multidisciplinary of oncologist and gynecologist. Patient withstood the surgery well and recovered well. She had 3 monthly follow-ups for 1 year in which she remained disease free (with history, clinical, radiological assessment and beta hCG levels). Currently she is at annual follow-up in the joint oncology and gynecology department at KMC Manipal and doing well.

Histopathology

Figures 1 and 2 shows histopathology images.

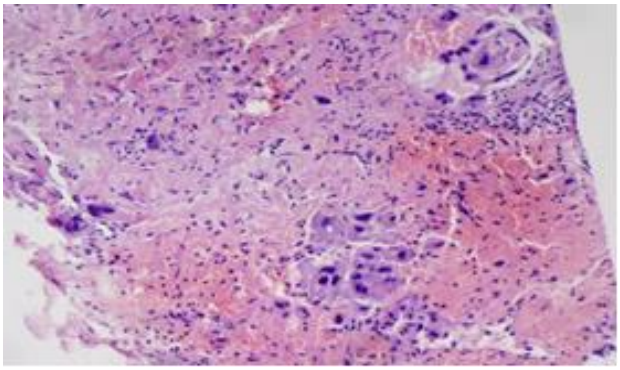

Figure 1: Photomicrograph showing clusters of malignant trophoblasts (H&E, X20).

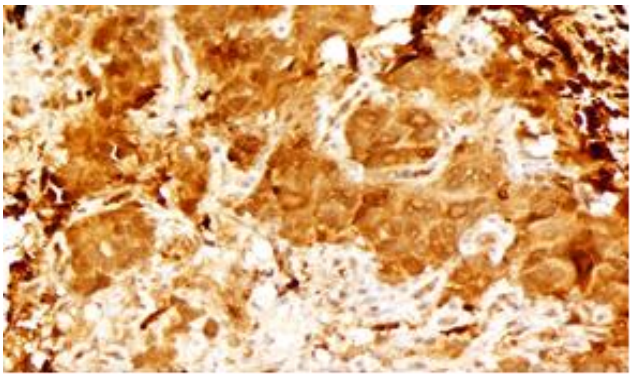

Figure 2: Tumor cells showing b-hCG immunoreactivity (X40).

Radiology

Computed tomography scan done pre-procedure for evaluation.

Following the neoadjuvant chemotherapy (NACT) treatment she underwent laparotomy salphingo-oopherectomy and histopathology reported as choriocarcinoma right ovary and reported to no residual tumor.

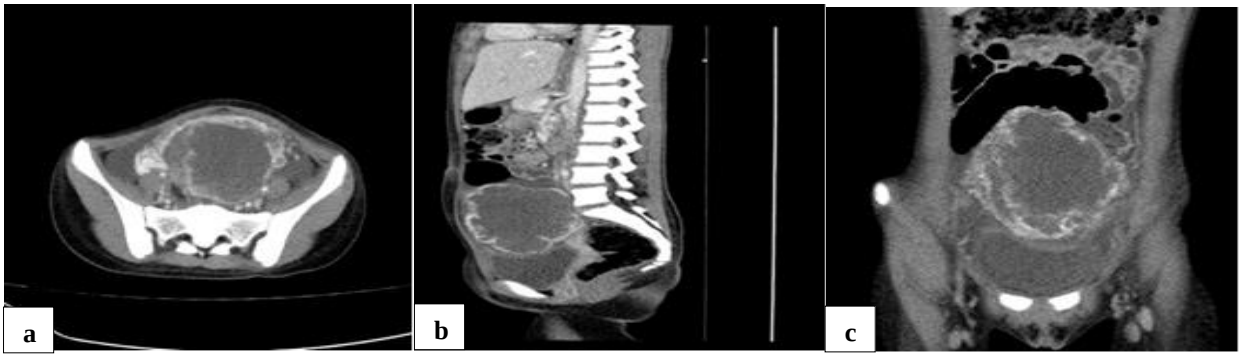

Figure 3: Radiology images (a) axial view; (b) sagittal view; and (c) coronal view.

T- Choriocarcinoma tumor

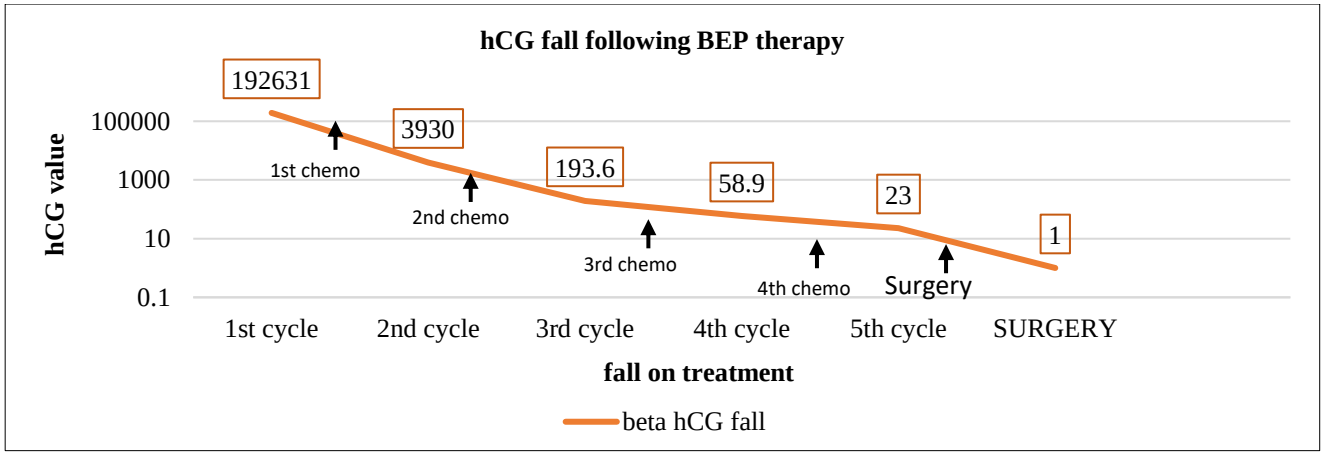

Figure 4: β-hCG regression curve.

DISCUSSION

Pure choriocarcinoma of ovary is a rare but highly malignant tumor of germ cell origin. It is characterized by presence of pathological trophoblastic malignant tissue and biochemical production of β-hCG in the absence of ongoing pregnancy. Early metastasis is encountered through blood and lymphatics.

Choriocarcinoma has a penchant for vascular invasion which is in keeping the embryological function of the normal trophoblast. It's so rare a malignancy that prognosis, chemo-sensitivity and genetics for non-gestational choriocarcinoma is not well established when compared to the gestational type.

The diagnosis of pure ovarian choriocarcinoma is very difficult and mainly in child bearing age group. In presence of high β-hCG and adnexal mass is most often confused for an ectopic pregnancy. There is no ultrasound and immune-histochemical differences between the gestational or non-gestational choriocarcinoma. The presence of a well-developed corpus luteum of pregnancy adjacent to the tumor may be indicative of gestational origin of the tumor. Several studies have investigated high expression of β-2 microglobulin (BMG) in choriocarcinoma.[7] Tanaka et al reported 11 different cases of gestational choriocarcinoma with lack of expression of messenger RNA for BMG and its presence in non-gestational choriocarcinoma.[8] Studies done by Kato et al and Norman et al indicated use of serum BMG as a serum tumor marker for non-gestational choriocarcinoma.[9] Presence of paternal DNA is a definitive distinction of gestational and non-gestational origin of the tumor. Non gestational choriocarcinoma shows the presence of only maternal origin without any alleles for paternal DNA.[10]

Non gestational has not shown association with factors responsible for other germ cell tumor except for dysgenetic gonads.

Goswami et al analyzed 30 cases of pure choriocarcinoma in 2001 and other studies concluded that pure choriocarcinoma responds well to combination of surgical ablation and post –operative chemotherapy with BEP regimen but till date there is no set definitive treatment modality.[2] In our patient direct chemotherapy with BEP regimen was started considering the poor performance status of the girl. Gestational choriocarcinoma is highly chemo-sensitive and responds well to methotrexate-based chemotherapy as compared to non-gestational counterpart for which BEP regimen is found to be effective. There was progressive fall in β-hCG.[11-13]

The study highlights several features, treatment and emphasizes the rarity of pure ovarian choriocarcinoma. The disease concerned is so rare that a stricter diagnostic criterion should be used in order to correctly categorize non-gestational origin from gestational origin, as unless confirmed by DNA analysis or the disease occurs in a patient who is pre-menarchal, one cannot with absolute certainty classify an ovarian choriocarcinoma.[2,7] Recently, DNA polymorphic analysis has been used to successfully determine non-gestational versus gestational origin of ovarian choriocarcinoma. This technology will allow for appropriate classification of this disease process which will ultimately lead to improved therapeutic strategies as more information is learned about pure non-gestational ovarian choriocarcinoma.[14]

Diwei et al described a 20-year male diagnosed with a rare case of metastatic testicular choriocarcinoma with pulmonary metastases which showed sustained and complete response to adjuvant chemotherapy with BEP regimen.[15] Another case of metastatic choriocarcinoma of the testis that presented as a cholestatic jaundice due to mets to liver but patient died one month later, despite general chemotherapy with BEP regimen.[16]

A case report of a young women with a clinical suspicion of ruptured ectopic pregnancy underwent emergency laparotomy with an intra op finding of ovarian mass with

bleeding later diagnosed as primary pure non-gestational choriocarcinoma of ovary.[17]

The only case report of a 13-year-old girl with pure choriocarcinoma in turkey managed with triple chemotherapy consisting of metho-trexate, actinomycin and cyclophosphamide that was started 2 weeks following the surgery at an oncology center and was followed with β-hCG and the patient received one additional course of chemo-therapy.[18]

CONCLUSION

There is no standard therapy of treatment established yet for the disease. Treatment is often extrapolated from treatment strategies for gestational choriocarcinoma and other germ cell tumors, thereby leading to significant heterogeneity. In reviewing the clinical outcomes of those with pure ovarian choriocarcinoma, it is difficult to make definitive treatment recommendations secondary to heterogeneity and inconsistent reported of, relevant clinical factors, including disease classification, patient age, stage, surgery, adjuvant therapy and outcomes, combined with the rarity of pure ovarian choriocarcinoma. Commonly used adjuvant combinational treatment regimens include BEP and EMA. A multidisciplinary approach to juvenile cancer can have a fruitful outcome with high cure rates.

Clinical significance

As far as our knowledge is concerned this is the 2nd only case report of pure choriocarcinoma to be diagnosed at such an early age of 13 and the only case report from Indian subcontinent. The rarity of this cancer makes it difficult to have clear cut diagnosis and fixed treatment plan. Due to paucity of data in regard to pure choriocarcinoma this case report may help in gathering more attention to the research towards this cancer.

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REFERENCES

1. Corakci A, Ozeren S, Ozkan S, Gurbuz Y, Ustun H, Yucesoy I. Pure nongestational choriocarcinoma of ovary. Arch Gynecol Obstet. 2005;271(2):176-7.
2. Goswami D, Sharma K, Zutshi V, Tempe A, Nigam S. Nongestational pure ovarian choriocarcinoma with contralateral teratoma. Gynecol Oncol. 2001;80:262-6.
3. Russell P, Farnsworth A, editors. Surgical Pathology of the Ovaries. 2nd. Churchill Livingstone; Edinburgh. Non-gestational choriocarcinomas. 1997;263-4.
4. Scully RE, Young RH, Clement PB. Tumors of the Ovary, Maldeveloped Gonads, Fallopian Tube, and Broad Ligament. Atlas of Tumor Pathology. 3rd series. Fascicle. Volume 23. Armed Forces Institute of Pathology; Washington DC. Choriocarcinoma. 1998;258-60.
5. Axe SR, Klein VR, Woodruff JD. Choriocarcinoma of the ovary. Obstet Gynecol. 1985;66(1):111-4.
6. Vance RP, Geisinger KR. Pure nongestational choriocarcinoma of the ovary report of a case. Cancer. 1985;56(9):2321-5.
7. Jacobs AJ, Newland JR, Green RK. Pure choriocarcinoma of the ovary. Obstet Gynecol Surv. 1982;37:603-9.
8. Tanaka K, Nabeshima Y, Takahashi H, Takeuchi S, Nabeshima Y, Ogata K.. Lack of effective messenger RNA for beta 2-microglobulin in a gestational human choriocarcinoma cell line (GCH-1) Cancer Res. 1981;41:3639-41.
9. Kato M, Ohashi K, Saji F, Wakimoto A, Tanizawa O. Expression of HLA class I and β2-microglobulin on human choriocarcinoma cell lines: induction of HLA class I by interferon-γ. Placenta. 1991;12(3):217-26.
10. Koo HL, Choi J, Kim KR, Kim JH. Pure non-gestational choriocarcinoma of the ovary diagnosed by DNA polymorphism analysis. Pathol Int. 2006;56(10):613-6.
11. Lv L, Yang K, Wu H, Lou J, Peng Z. Pure choriocarcinoma of the ovary: a case report. J Gynecol Oncol. 2011;22(2):135-9.
12. Hayashi S, Abe Y, Tomita S, Nakanishi Y, Miwa S, Nakajima T, Nomoto K, Tsuneyama K, Fujimori T, Imura J. Primary non-gestational pure choriocarcinoma arising in the ovary: A case report and literature review. Oncol Lett. 2015;9(5):2109-11.
13. Mood NI, Samadi N, Rahimi-Moghaddam P, Sarmadi S, Eftekhar Z, Yarandi F. Pure ovarian choriocarcinoma: report of two cases. J Res Med Sci. 2009;14(5):327-30.
14. Jia N, Chen Y, Tao X, Ou E, Lu X, Feng W. A gestational choriocarcinoma of the ovary diagnosed by DNA polymorphic analysis: a case report and systematic review of the literature. J Ovarian Res. 2017;10(1):1-8.
15. Lin D, Tan AJ, Singh-Rai R. A Literature Review and Case Report of Metastatic Pure Choriocarcinoma. Case Rep Oncol Med. 2015;345018.
16. Ahsaini M, Tazi F, Mellas S, Elammari J, Khalouk A, Wieringa RS, et al. Pure choriocarcinoma of the testis presenting with jaundice: a case report and review of the literature. J Med Case Rep. 2012;6:269.
17. Rawal G, Ahluwalia C, Yadav AK, Dhawan I. Primary pure non-gestational choriocarcinoma of ovary: a rare case report. Int J Reprod Contracept Obstet Gynaecol. 2017;6(9).
18. Ozdemir I, Demirci F, Yucel O, Demirci E, Alper M. Pure ovarian choriocarcinoma: a difficult diagnosis of an unusual tumor presenting with acute abdomen in a 13-year-old girl. Acta obstetricia et gynecologica Scandinavica. 2004;83(5):504-5.

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