AUTOIMMUNE HEMOLYTIC ANEMIA ASSOCIATED WITH MATURE TERATOMA : A RARE PRESENTATION

Mathew J Kottarathara M.D. , Yousef B A Mohamed M.D. , Rahma Anchimbon M.D. , Aizaaz Khan M.D. , Zulekha Atif M.D. , Jose Cervantes M.D.
Jamaica Hospital Medical Center, Jamaica, NY , 11418 - USA

Introduction :

Autoimmune hemolytic anemia (AIHA) occurs when antibodies are directed against one's own red blood cells (RBCs). It is usually associated with viral syndromes, lupus or drug reactions. As described in the case below, teratoma should be considered in AIHA.

Case Description:

A 19-year-old female presented with jaundice and lethargy of 1-week duration and 1 episode of dark urine. The examination was benign except for generalized icterus.

Admission labs revealed hemoglobin of 6.7 g/dL, HCT of 19.3%, MCV 128.6 fl, leukocytosis 17 K/uL, haptoglobin <15, LDH 4726 U/L, reticulocyte count of 36.4 %, ALT 59 U/L, and AST 172 U/L. B12, folate and TSH levels were within normal limits. Coombs test revealed acute warm hemolytic anemia. Peripheral smear showed numerous microspherocytes and polychromasia. Viral and mycoplasma serologies, ds-DNA and ANA were negative.

The patient was started on methylprednisone but hematocrit did not improve. Abdominal sonogram (Figure 1) incidentally noted a 12.0x9.4x13.7 cm structure in the right adnexa with echogenic components. The CT scan (Figure 2) showed a large septated mass in the posterior pelvis with fat fluid level and soft tissue density. A cluster of large dense calcification was noted within this mass consistent with a dermoid cyst from the right ovary. A decision was made to remove the dermoid cyst laparoscopically.

Follow-up hematocrit a week after surgery showed an improvement to 31.9%. The patient never required blood transfusions.

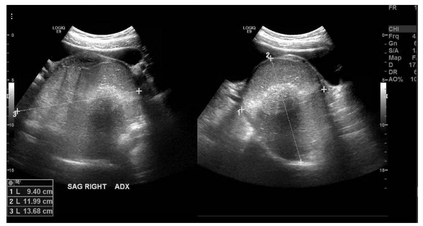


Figure 1: Abdominal sonogram showing a 12.0x9.4x13.7 cm structure in the right adnexa with echogenic components.

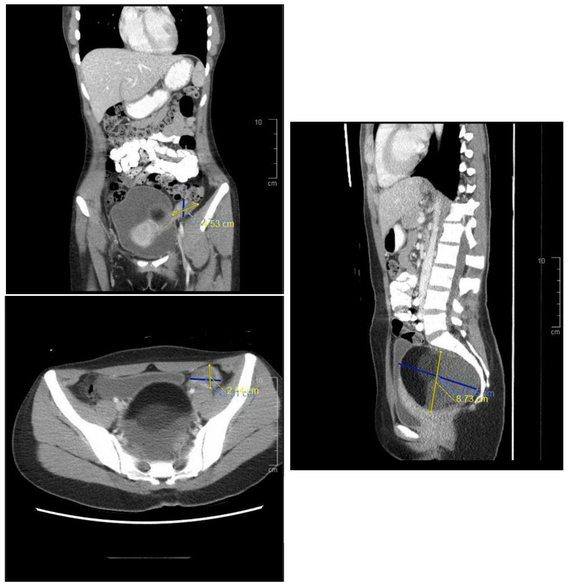
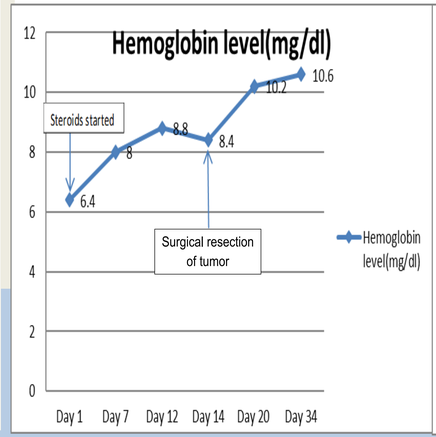
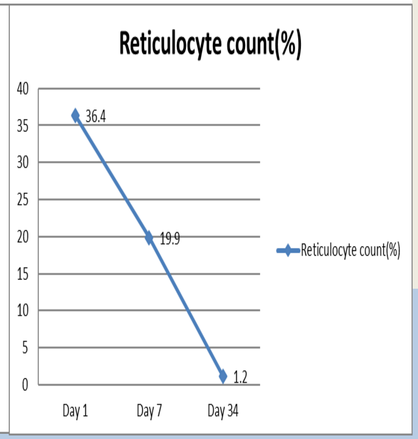


Figure 2: Computed tomographies of pelvis and abdomen showing a large septated mass in the posterior pelvis with fat fluid level and soft tissue density. A cluster of large dense calcification was noted within this mass consistent with a dermoid cyst from the right ovary.





Discussion :

Mature teratomas constitute around 10-20% of all ovarian tumors among 20-40 year olds. AIHA due to mature teratoma is seen in less than 1% of cases. Glucocorticoids and splenectomy are the mainstay treatment of AIHA but are not effective in the case of ovarian teratoma. Mechanisms of AIHA in such cases have been explained by a number of hypotheses.

Some propose that the tumor produces a substance that coats the RBCs making them antigenic to the host, while others suggest the tumor itself may be antigenic and antibodies produced in response to it cross-react with host cells. The fact that removal of the tumor is curative supports the latter hypothesis. The half-life of IgG is roughly 23 days and antibodies have shown to persist from 2 weeks to 7 months after removal of the tumor

Conclusion :

In patients with steroid resistant hemolytic anemia, mature teratomas should be ruled out. If present, management should include surgical removal as it is the only curative therapy and harmful procedures such as splenectomy can be avoided.

References:

1.Payne D, et al. Autoimmune hemolytic anemia associated with ovarian dermoid cyst: case report and review of the literature. Cancer 1981; 43: 721–4
2.Rathore R, Sharma S, Arora D. Clinicopathological Evaluation of 223 Cases of Mature Cystic Teratoma, Ovary: 25-Year Experience in a Single Tertiary Care Centre in India. Journal of Clinical and Diagnostic Research : JCDR. 2017;11(4):EC11-EC14. doi:10.7860/JCDR/2017/23909.9612.
3.Sahin, H., Abdullazade, S. & Sanci, M. Insights Imaging (2017) 8: 227. https://doi.org/10.1007/s13244-016-0539-9
4. Puthenparambil J, Lechner K, Kornek G. Autoimmune hemolytic anemia as a paraneoplastic phenomenon in solid tumors: a critical analysis of 52 cases reported in the literature. Wien Klin Wochenschr 2010;122:229-36. 3
5. Park SB, Kim JK, Kim K, Cho K (2008) Imaging findings of complications and unusual manifestations of ovarian neoplasms. RadioGraphics 28:969–983
6.Raimundo PO, Coelho S, Cabeleira A, Dias L, Gonçalves M, Almeida J. Warm antibody autoimmune hemolytic anemia associated with ovarian teratoma. BMJ Case Reports. 2010;2010:bcr06.2009.1971. doi:10.1136/bcr.06.2009.1971.
7.Kim I, Lee JY, Kwon JH, et al. A Case of Autoimmune Hemolytic Anemia Associated with an Ovarian Teratoma. Journal of Korean Medical Science. 2006;21(2):365-367. doi:10.3346/jkms.2006.21.2.365