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Bilateral Ovarian Dermoid Cysts with Persistent Anemia and Thrombocytopenia in an Adult Female: A Rare Case Report

Nidhi Chaturvedi [a#], Neema Tiwari [a*#] and Rani Bansal [a¥]

[a] Department of Pathology, Subharti Medical College, Meerut, India.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Autoimmune phenomena are commonly encountered in the female population either as the primary disease or in association with conditions like pregnancy or as a part of paraneoplastic syndromes. Exact pathogenesis of such immune reactions may be clearly defined in cases like Grave's disease, SLE etc or may be secondary to unexplained phenomena .These patients however may manifest as hematological symptoms like anemia and thrombocytopenia, skin manifestations or co existing thyroid issues .Tumors commonly present with autoimmune paraneoplastic syndromes. These can be non-hematological and hematological. Common hematological manifestations we encounter are either ITP or autoimmune hemolytic anemias. Ovarian tumors rarely occur with autoimmune features. There have been documented cases of bilateral dermoid cyst with associated Immune thrombocytopenia (ITP) as well as autoimmune hematological anemias in recent literature. Here we discuss a case of a 26-year-old female with bilateral ovarian dermoid cyst, who presented with transfusion dependent anemia and thrombocytopenia. Her anemia responded to immunosuppression therapy but thrombocytopenia responded to lesser extent.

Keywords: Ovarian; dermoid; autoimmune; anemia; thrombocytopenia.

[#] *Assistant Professor;*
[¥] *Professor and HOD;*
**Corresponding author: E-mail: nehaneemat@yahoo.co.in;*

1. INTRODUCTION

Autoimmune phenomena are commonly encountered in the female population as the primary disease or in association with conditions like pregnancy, or as a part of paraneoplastic syndromes. Exact pathogenesis of such immune reactions may be clearly defined in cases like Grave's disease, SLE etc or may be secondary to unexplained phenomena .These patients however may manifest as hematological symptoms like anemia and thrombocytopenia or skin manifestations or co existing thyroid issues .Tumors may present with autoimmune paraneoplastic syndromes which can be non hematological and haematological [1]. Common hematological manifestations we encounter are either ITP or autoimmune hemolytic anemias. Rarely ovarian tumors may occur with autoimmune features. There have been documented cases of bilateral dermoid cyst with associated Immune thrombocytopenia (ITP) as well as autoimmune hematological anemias in recent literature.

Autoimmune hemolytic anemias may occur in several conditions and one of the rarer conditions for it to occur in is ovarian tumors. The mechanism for this phenomenon is not well understood, however some authors hypothesize about role of lymphocytes in dermoid tumors causing autoimmune reactions [1-4]. "Autoimmune hemolytic anemia is IgG Mediated disease and is a serious condition for mother and fetus [5]. Similarly ITP, another manifestation of immune reactions in ovarian tumors is a hematologic disorder involving accelerated platelet destruction with compromised platelet production" [2]. "It is divided into primary ITP, an isolated thrombocytopenia, and secondary ITP, where disorders such as infectious (eg, Hepatitis C virus, HIV), or autoimmune (eg, systemic lupus erythematosus, chronic lymphocytic leukemia, antiphospholipid syndrome) disorders are causing the thrombocytopenia" [1]. It is ultimately a diagnosis by exclusion, made upon assessment of the patients' response to treatment. Corticosteroids are used as first line therapy agents and may be supplemented by , intravenous immunoglobulin, and anti-Rh(D) immune globulin.

Here we discuss a case of a 26-year-old female with bilateral ovarian dermoid cyst, who presented with transfusion dependent anemia and thrombocytopenia. Her anemia responded to immunosuppression therapy but thrombocytopenia persisted.

2. CASE REPORT

We present a 26-year-old female who presented to the OPD with complaints of fatigue and shortness of breath of 2 weeks. Her relevant examination findings were within normal limits. Her Investigations revealed RBC count of 0.97 million, Hb-3.5gm/dl, Hct-12.5%, MCV 128.4fl, MCH 36.5pg, MCHC28.4gm/dl, RDW-CV 16.5%,Platelet count-50000/mm3. DLC showed 68% polymorphs 30%lymphocytes and 2% monocyte with a TLC of $4.6x10^3$/mm3.Her reticulocyte count was 3.5%, her peripheral smear showed macrocytosis with thrombocytopenia. Bone marrow aspiration reported erythroid hyperplasia with megaloblastoid features and increased iron stores. Bone marrow biopsies revealed erythroid hyperplasia with megakaryocytopenia. Her DCT was negative but ANA was 1+. Urine examination was unremarkable, serum ferritin 269ng/ml, LFT, KFT Thyroid function test were within normal limits. Vitamin B12 was within normal limits. Her USG whole abdomen revealed bilateral dermoid cyst. The patient is currently under treatment for immune mediated cytopenias. Her hemoglobin levels improved within a week of corticosteroid therapy though her thrombocytopenia persisted. Hence the patient has been planned for bilateral cyst removal surgery.

3. DISCUSSION

In our case the patient had persistent transfusion dependent anemia and thrombocytopenia. She had been transfused with 6 units of RBCs and 2 units of platelets (SDP). In view of immune mediated cytopenias she was started on steroids (1mg/kg body weight), her hemoglobin has showed recovery (with upto 1.5gm/dl increase in 10 days) and her thrombocytopenia responded to some degree with her last platelet counts being 35000/mm^3(previous counts were 24000/mm^3). We hypothesize that since there has been some response to steroid therapy in this case ,post bilateral oophorectomy her platelet counts and hemoglobin should recover completely. The response to steroid favours a diagnosis of autoimmune mediated suppression of hemoglobin and platelets. "There are very few papers published regarding the frequency of occurrence, the mechanisms of association, and the clinical behaviour of hematologic abnormalities in patients with ovarian teratoma. There is one case of ITP in association with ovarian teratoma with associated pregnancy

described in the literature while some other cases of ITP occurring as paraneoplastic syndrome in extragonadal germ cell tumors, embryonal cancers with mediastinal masses have also been reported" [6,7].

Initially due to her clinical condition a diagnosis of aplastic anemia was suspected but as mentioned in the case the bone marrow findings were not supporting the diagnosis. The mechanism of association between cytopenias and ovarian teratoma is still unknown. It is believed that cytopenias like thrombocytopenia starts with antibodies against a platelet glycoprotein leading to the degradation of antibody-coated platelets in the spleen. This degradation results in the release of more peptides from the platelet constituents, with production of new antibodies against the platelet-derived peptides [8].

The substance secreted by the teratoma may be toxic to the normal platelets with direct platelet membrane damage, affecting the survival of the platelets. It is likely that the effect is not only on platelet destruction but also on platelet production. If the effect is on platelet production, then the use of thrombopoietin receptor agonists might be of use to increase platelet count prior to surgery in such patients [9].

"Agarwal et al in 2003 reviewed 20 cases of AIHA associated with dermoid cyst, 85% of them responded to cystectomy after failed corticosteroids and one underwent cystectomy shortly after diagnosis who showed some response to steroid management" [6]. "Our patient has been started on steroid therapy and is under observation. Persistent thrombocytopenia may be a poor prognostic factor in such cases" [6-8].

The patient was on steroid therapy and has slowly responded to treatment with anemia and thrombocytopenia slowly getting better. Other Other hematologic abnormalities co existing with non hematological malignancies may include myelodysplasia, idiopathic thrombocytosis, acute lymphocytic leukemia, or acute myeloid leukemia.

Association between dermoid ovarian cyst and AIHA is still a rare phenomenon with a limited number of cases reported in the literature. Ovarian teratomas are relatively common, but the incidence of associated hemolytic anemia is low. This phenomenon was reported by West-Watson and Young [8] but reported cases are still limited especially in pregnancy. There are different hypothesis describing the immunological reaction triggered by the tumor. Several reports support the hypothesis that the tumor produces antibodies against red blood cells/platelets. Antibody production seems to cease immediately after tumor removal in almost all reported cases [9].

"Payne et al. [10] in 1981 have carried out a literature review of cases reported as dermoid cysts with AIHA, and collected around 19 cases. Sixteen patients responded favourably to tumor removal alone. Negligible response to steroids was reported in 3 patients who required dermoid removal to achieve full response" [11,12].

There are few case reports where ovarian dermoid cyst cases have been reported with no apparent gynaecological/obstetric complaints but only with cytopenias on CBC as presenting complaint. Our case is similar however there is suboptimal response to immunosuppression although the clinician gave a her provisional diagnosis of Autoimmune cytopenias.

4. CONCLUSION

Hence, we conclude that any young patient with cytopenias and suboptimal response to standard therapeutic measures should be investigated for co-existing tumors as cytopenias in many such cases gets resolved once the tumor is removed.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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