INTERNATIONAL JOURNAL OF CURRENT ADVANCED RESEARCH
ISSN: O: 2319-6475, ISSN: P: 2319-6505, Impact Factor: 6.614
Available Online at www.journalijcar.org
Volume 12; Issue 08(A); August 2023; Page No. 2447-2449
DOI: http://dx.doi.org/10.24327/ijcar.2023.2449.1532

Research Article

DENTAL MANAGEMENT OF PATIENTS WITH EVANS SYNDROME

Saniya Firdose, Mamatha NS and Ankesh Jain

Oral and Maxillofacial Surgery

ARTICLE INFO

Article History:
Received 6th May, 2023
Received in revised form 15th June, 2023
Accepted 12th July, 2023
Published online 28th August, 2023

Key words:
Evans syndrome, dental treatment, Auto-immune hemolytic anemia, Immune thrombocytopenia.

ABSTRACT

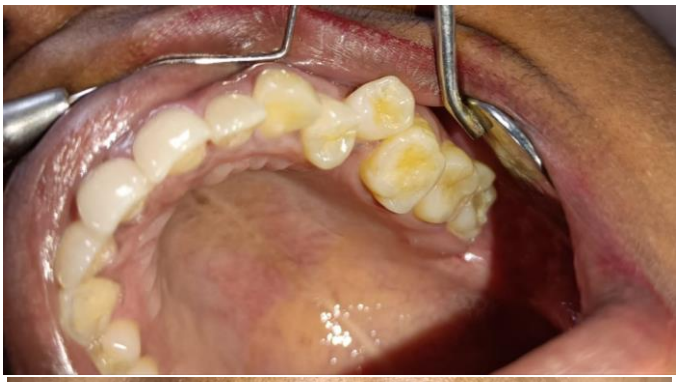
Evans syndrome (ES) is a rare hematologic disorder characterized by the concomitant or sequential association of warm Auto-immune hemolytic anemia (AIHA) with Immune thrombocytopenia (ITP) and less frequently autoimmune neutropenia. Patients with combinations of autoimmune diseases carry a worse prognosis and are often difficult to treat as they are at risk of post-operative bleeding also the drugs that are routinely used in treatment carry a potential adverse reaction. Knowing this medical condition is paramount to providing safe and effective dental treatment and preventing undesirable events. This article aims to report a case of management of a patient with Evans syndrome, treatment of Evans syndrome, and suggests a guide for dental practitioners when treating patients with the condition.

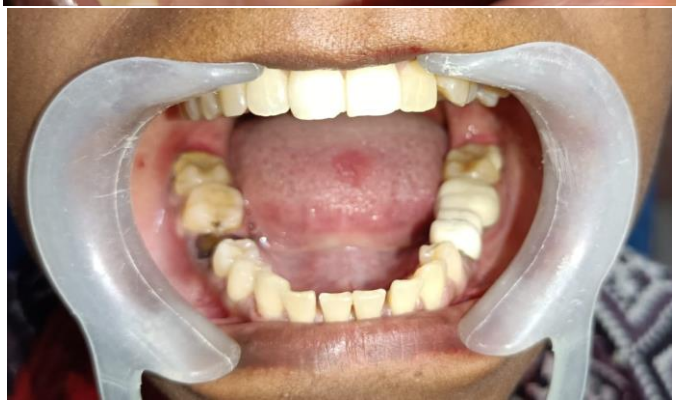
INTRODUCTION

Evans syndrome was first described by Robert Evans in the year 1951.[1] It is defined by the simultaneous presentation of autoimmune hemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP) and sometimes autoimmune neutropoenias, which may appear concomitantly or one may occur earlier than the other by several months.[2] Evans described the first diagnosis criteria as the presence of anemia, reticulocytosis, increased serum bilirubin levels, fecal urobilinogen, evidence of antibodies against erythrocytes at 37^{o}c, hemolysis of transfused erythrocytes, the presence of purpura, prolonged bleeding time, bone marrow aspiration with normal or increased number of megakaryocytes, with no other etiology like the presence of exogenous toxins or any family history of hemolytic diseases.[3] Several studies have reported that adult patients with Evans syndrome have been linked to other diseases, such as systemic lupus erythematosus and aplastic anemias.[4] Dental management of patients with ES requires knowledge of clinical manifestations and treatment modalities to avoid uneventful experiences and deliver the right treatment**.**

CASE REPORT

A female patient aged 42 years reported to the Department of Oral and maxillofacial surgery RRDCH with the chief complaint of pain in the upper left back region of the mouth for 5 days. The patient was diagnosed with ES at the age of 27 years. She had been hospitalized multiple times requiring blood transfusions and had also undergone a splenectomy six years back. She was asymptomatic for the last 5 years and is closely monitored with hematologic screening once every 3 months. The patient discontinued her regimen of medications in the month of January 2023 as per her physician's advice, which included – Azathioprine 50mg and Hydrocortisone 100mg for 13 years. Currently, she is on Hydroxychloroquine 200mg one tablet per day.





Upon physical examination, the patient was well-nourished and moderately built. Her vital signs were within normal

Corresponding author:* **Saniya Firdose
Oral and Maxillofacial Surgery

limits. Intraoral examination revealed multiple carious teeth and the presence of a prosthesis in the 3rd quadrant.

Laboratory values at the presentation were within normal limits.

	Patient	Reference
WBC (flow cytometry)	**11330 cells/cumm**	4500-11000 cells/cumm
RBC (hydrodynamic DC detection)	4.75 mill/cumm	3.8-4.8 mill/cumm
Hb (sodium lauryl sulphate)	13.5 g/dl	12-15 g/dl
PLATELETS (hydrodynamic DC detection)	233000/µL	150000-300000/µL
WBC differential (Auto analyzer/ flow cytometry)		
Lymphocytes	**40.9 %**	20-40%
Neutrophils	47.0%	40-75%
Eosinophils	2.8%	<7%
Monocytes	8.8%	2-10%
Basophils	0.5%	0-1%

Owing to multiple carious lesions 4 teeth were indicated for extraction. The treatment plan was described to the patient in detail. After consultation with the physician and the recent record of hematologic stability, extraction under Local anesthesia was considered.

The extractions were performed without complications in 3 appointments each week apart. Hemostasis was achieved. Postoperative instructions were given in written and verbal format, antibiotics and analgesics (paracetamol) were prescribed, and the patient was discharged home. The patient returned after a month for a follow-up. The clinical examination of the surgical areas revealed adequate healing.

DISCUSSION

ES is a rare form (1%) of immune thrombocytopenic purpura and associated Autoimmune hemolytic anemia. It was first described by Fisher and Evans. The combination of the above two conditions can be associated with underlying autoimmune diseases, lymphoproliferative diseases, or primary immunodeficiencies.[5] This condition's median age of presentation ranges between 5.5 and 7.7 years, with an overall range of 0.2-26.6 years. No sex predilection is known, and Evans syndrome can be found in any ethnic group.

The etiology of ES is unknown and is defined as idiopathic. Recent studies suggest that ES is a state of profound immune dysregulation and a coincidental combination of immune cytopenias. The clinical presentation of ES is consistent with a generalized immune failure, with cellular and humoral immunity abnormalities. Neutropenia occurs in up to 55% of patients or pancytopenias.[1]

The patient presents with symptoms of hemolytic anemia – pallor, lethargy, jaundice, and symptoms of thrombocytopenia – petechiae, frequent bruising, mucosal bleeding, cutaneous bleeding, hematuria, menorrhagia, fatigue and in several cases cardiac complications. An examination may reveal lymphadenopathy, or organomegaly (hepatic or splenic) which could be chronic or intermittent and only be apparent during acute exacerbations of the disease. There are high chances of recurrence and patients are also at high risk of developing malignancies.

The diagnosis of ES is mainly by the exclusion of other conditions with similar clinical presentations. A full blood count will confirm the presence of cytopenias and a blood film should be examined for features like polychromasia, spherocytes, raised reticulocyte count, unconjugated hyperbilirubinemia, and decreased haptoglobins. A decreased number of T-lymphocytes and abnormal CD4/CD8 ratios have been described.[1]

Management strategies in ES attempt to decrease the clinical expression of the syndrome and control the autoimmune response. Corticosteroids are commonly used as the initial line of therapy, alone or in combination with other agents. Prednisolone 2-4 mg/kg/day has been effective. Intravenous immune globulin (IVG) first described in 1985, is an alternative to patients for whom high doses of long-term corticosteroids are required, and whose side effects are severe. It is given at a total dose of IVG of 2 g/kg, combined with other therapy forms, including cyclosporine, cyclophosphamide, vincristine, danazol, and azathioprine.[6] Recently Rituximab, an anti-CD20 monoclonal antibody that depletes B-cells through cellular antibody toxicity and cell apoptosis, is considered an effective line of treatment if corticosteroids do not provide the desired pharmacological effect.[7]

Splenectomy is another mode of treatment when other medication therapy fails. Although the patients undergoing splenectomy might require long-term antibiotic coverage, apart from complications such as sepsis, etc.

Dental management of patients with ES comes with different challenges as there is always the risk of complications. There are increased risks of bleeding and life-threatening hemorrhages due to reduced platelet count caused by antiplatelet antibodies. The clinician should record a detailed medical history along with consultation from the patient's hematologist. A complete blood count has to be done 24-48h before extractions. A decreased platelet count of <50000/µL indicates the need for intravenous immunoglobulin infusion 24 h before the extractions. A Neutrophil count <1000/mm^3 requires antibiotic prophylaxis with post-operative antibiotic coverage for 7 days.

Patients on long-term steroid therapy might be at risk of developing acute adrenal crisis during the time of the procedure hence it might necessitate steroid coverage before a moderate or invasive procedure. Also, Patients who have undergone splenectomy require antibiotic prophylaxis as they are at high risk of developing systemic infections.

References

1. Norton A, Roberts I. Management of Evans syndrome. Br J Haematol 2006;132:125-37.
2. Pinto A, Lindemeyer RG, Alawi F. Management of a young patient with combined autoimmunity: Evans syndrome: a case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2007 Apr;103(4):505-11. doi 10.1016/j.tripleo.2006.07.022. Epub 2006 Nov 7. PMID: 17095266.
3. Jaime-Pérez JC, Aguilar-Calderón PE, Salazar-Cavazos L, Gómez-Almaguer D. Evans syndrome: clinical perspectives, biological insights, and treatment modalities. J Blood Med. 2018 Oct 10;9:171-184. doi 10.2147/JBM.S176144. PMID: 30349415; PMCID: PMC6190623.

4. Miescher PA, Tucci A, Beris P, Favre H. Autoimmune hemolytic anemia and/or thrombocytopenia associated with lupus parameters. Semin Hematol 1992;29:13-7
5. Savasan S, Warrier I, Ravindranath Y. The spectrum of Evans' syndrome. Arch Dis Child 1997;77:245-8.
6. Pui CH, Williams J, Wang W. Evans' syndrome in childhood. *J Pediatr* 1980; 97:754-8.
7. Shanafelt TD, Madueme HL, Wolf RC, Tefferi A. Rituximab for immune cytopenia in adults: idiopathic thrombocytopenic purpura, autoimmune hemolytic anemia, and Evans syndrome. Mayo Clin Proc 2003;78:1340-6

How to cite this article:

Saniya Firdose, Mamatha NS and Ankesh Jain (2023) 'Dental Management of Patients with Evans Syndrome', *International Journal of Current Advanced Research,* 12(08), pp. 2447-2449.
DOI: http://dx.doi.org/10.24327/ijcar.2023.2449.1532

****