Original Research Paper

Volume-10 | Issue-1 | January - 2020 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Medicine

LUPUS - AN OLD WOLF WITH A NEW FRIEND

Dr. Suhas. Murali Resident, Department Of General Medicine, Jawaharlal Nehru Medical College, Belgaum, Karnataka

Dr. Archana Khanagavi* Assistant Professor, Department Of General Medicine, Jawaharlal Nehru Medical College, Belgaum, Karnataka *Corresponding Author

Dr. Santosh Hajare Professor, Department Of Gastroenterology, Jawaharlal Nehru Medical College, Belgaum, Karnataka

ABSTRACT Systemic Lupus Erythematosus (SLE) is common amongst the various rheumatological conditions noted in clinical practice. But rarer entities like Cronkhite-Canada syndrome are reported very rarely in literature specially from the Indian subcontinent. Hence the knowledge available of such rare conditions are also sparse. This is an extremely rare case of a combination of these two conditions which may help researches provide some insight into the disease processes of this condition.

KEYWORDS : Systemic Lupus Erythematosus, Cronkhite-Canada Syndrome

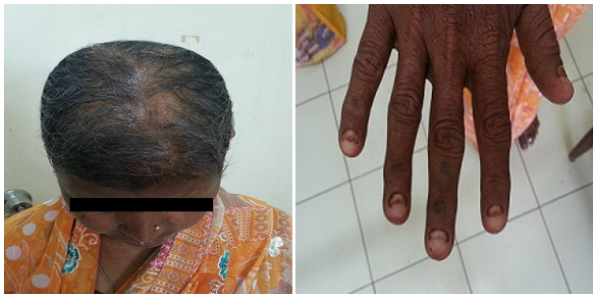
INTRODUCTION :

Systemic Lupus Erythematosus is fairly common among rheumatological disorders, with an incidence of 10-400 per 100,000 population in the United States[1]. Even in a developing country like India where the actual numbers are not fully accounted for, few studies do indicate an incidence of around 3.2 per 100,000 population[2]. As we know for a fact that this auto-immune condition is known to affect fairly a large spectrum of tissues in the body, the clinical manifestations of the same is quite varying. Hence it is important to thoroughly investigate for SLE when other autoimmune diseases are being worked up for. We present one such interesting case.

CASE REPORT :

A 42 year old female patient was referred to our hospital with a 6 months history of diarrhoea. She had severe loss of weight and loss of appetite. The patient had been worked up for most common causes chronic diarrhoea like infective, osmotic, dysmotility, malabsorptive causes over the last few months by her primary care physician. She had not responded to any symptomatic treatment either.

General examination was significant because it directed us to think towards an auto-immune pathology. She was found to have alopecia, malar rash, but significantly dystrophic nails at admission as shown in the Figure-1,2. The rest of the clinical examination was unremarkable.



Figures -1 & 2 : Shows the presence of alopecia (left) with dystrophic nails (right)

On routine investigations, she was found to have proteinuria. But rest of the haematological, renal, hepatic functions were all within normal limits.

She then underwent an Gastro-Duodenoscopy which showed the entire stomach and duodenum studded with polyps as shown in Figure-3. Later to complete the workup she also underwent a colonoscopy which showed that the colon was not spared either. Biopsy of both duodenum and colonic polyps were taken which showed villous atrophy along with minimal plasma cell infiltrate.

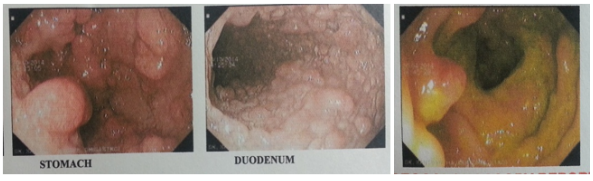


Figure-3: Endoscopic images of the Stomach (left), Duodenum (middle) and Colon (right) showing the presence of numerous short sessile polyps, from which biopsy was taken.

Now, based on the endoscopic findings in correlation with the skin and ectodermal changes she was suspected to have the rare Cronkhite-Canada syndrome. But the presence of malar rash, protienuria made us think she might be harbouring one more auto-immune condition like SLE. Hence a workup for SLE was undertaken. Indeed she turned out to be positive for Anti-Nuclear, Anti-Smith and Anti-Rho antibodies. Thus a diagnosis of Systemic Lupus Erythematosus along with Cronkhite-Canada Syndrome was made.

FOLLOW UP :

The patient was started on prednisolone and azathioprine and was discharged. She was followed up and one year later she underwent a repeat endoscopy which showed a marked regression of polyps as shown in Figure-4. Symptomatically too the patient had remarkable improvement of symptoms with markedly reduced hair loss no reported flares of SLE either as shown in Figure-5.

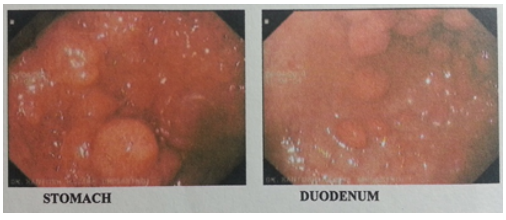


Figure-4 : Repeat Endoscopic images show the presence of marked reduction of the number of polyps in the stomach (left) and duodenum (right)

Figure-5: Shows the patient had marked reduction in hair loss after 1 year

DISCUSSION :

Cronkhite-Canada syndrome is an extremely rare condition characterised by the presence of ectodermal changes like onychodystrophy, hyperpigmentation, alopecia along with gastrointestinal polyps, diarrhoea, weight loss, abdominal pain. Details about this disorder is mostly from studies done in Japan among 110 cases. It pegs the incidence of this syndrome to be about one per a million population3. But so far the etiopathogenic mechanisms of the condition have not been clearly elucidated.

It is known to be associated with other auto-immune conditions like hypothyroidism and scleroderma4. But its association with SLE is an extremely rare entity so much so that the last few reported case of such a combination probably dates back to around 30 years ago in 19865. The importance of this association is the fact that, this may give a clue regarding the pathogenic mechanisms involved in Cronkhite-Canada syndrome. The association with SLE and the subsequent marked improvement seen in our patient with steroids and azathioprine, makes us think that an auto-immune mechanism similar to SLE has an etiological role along with probably other genetic and environmental factors, whose roles need to be looked into.

REFERENCES :

1. Hannahs Hahn B., Systemic Lupus Eryhthematosus.In;Kasper, Fauci, Longo et al, editor. Harrison's Principles of Internal Medicine, 18TH ed. USA, McGraw Hill publications;2012.p.2724.
2. Joshi V.R., Systemic Lupus Eryhtematosus.In; Munjal Y.P, editor.API Textbook of Medicine, 9th ed. India, Jaypee Publishers;2012.p.1853.
3. A. Goto, "Cronkhite-Canada syndrome: epidemiological study of 110 cases reported in Japan," Nihon Geka Hokan, vol. 64, no. 1, pp. 3–14, 1995.
4. Qiao M, Lei Z, Nai-Zhong H, Jian-Ming X. Cronkhite-Canada syndrome with hypothyroidism. SouthMed J. 2005;98:575–6. doi: 10.1097/01.SMJ.0000157528.71614.C4.
5. Kubo T, Hirose S, Aoki S, Kaji T, Kitagawa M. Canada-Cronkhite Syndrome Associated With Systemic Lupus Erythematosus. Arch Intern Med. 1986;146(5):995-996.