

subtle synovitis. Our patient had polyarthritis of both small and large joints and subsequently diagnosed to have seronegative RA according to the 2010 ACR-EULAR classification criteria of RA (score 7/10).

There are only few reports of RA with CVID. In one report a 38 yrs old lady with recurrent respiratory and gastrointestinal tract infections and pernicious anemia developed polyarthritis with damage of hand joints as evidenced by narrowing of the intra-articular space in the right metacarpophalangeal and radiocarpal joints with multiple bone cysts and erosions. Erosions were found in both humeral heads as well². Our patient is relatively young and developed polyarthritis at 17 years age and he had osteopenia without any features suggestive of joint destruction. In another report a 26 year old lady had a six-year history of chronic symmetric polyarthritis, three-year history of sicca syndrome and three episodes of pneumonia during previous three years. RA with secondary Sjögren's syndrome with CVID was the diagnosis. Polyarthritis and sicca syndrome improved with monthly administration of intravenous immunoglobulin (IVIG)³. Our patient had one-year history of polyarthritis that used to improve with NSAIDs and disease modifying anti-rheumatoid drugs (DMARDs) without immunoglobulin therapy. In one report a case of juvenile idiopathic arthritis (JIA) and JIA-associated uveitis develop infective features seven years after arthritic presentation and diagnosed as CVID at 23 years of age after 13 years of initial arthritic presentation.⁴ So arthritis may precede, may occur simultaneously or follow other features or complications of CVID. In our case infective disorders precede arthritis.

In our reported case, serum Ig G, Ig A and Ig E were reduced but Ig M was within normal range. Immunoglobulin levels ruled out the diagnosis of X-linked agammaglobulinemia because in that case Ig M would be undetectable. Normal WBC count ruled out severe combined immunodeficiency, as it occurs in infant with very low lymphocyte count.

Treatment of CVID includes intravenous immunoglobulin (IVIG) for passive protection. Generally, IVIG dose is 400 mg/kg at every three to four weeks. Patients with chronic sinusitis or chronic lung

disease require long-term treatment with broad-spectrum antibiotics. If bronchiectasis has developed, physical therapy and daily postural drainage are needed. In our case, polyarthritis used to improve with NSAIDs and DMARDs.

CONCLUSION

High level of awareness is needed for early diagnosis of CVID patients. CVID should be considered in arthritis patients especially in the background recurrent infections. Diagnosis of rheumatological disorders may also be problematic because of absent or low titre of antibodies and subtle clinical features due to inherent immunosuppression. Use of immunosuppressives should be restricted or to be used judiciously in associated rheumatological disorders because of risk of further immunosuppression. The case is reported because of rarity of association of RA with CVID and to make awareness among.

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Eosinophilic Granulomatosis with Polyangitis: A Case Report.

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Abstract: The aim of this paper is to report a case of very rare vasculitic syndrome i.e., eosinophilic granulomatosis with polyangitis (EGPA). A 47-year old lady developed rapidly progressive sensorimotor peripheral neuropathy with visual loss in one eye. She had history of allergic rhinitis and adult onset bronchial asthma. Examination showed healed vasculitic ulcers on left foot. Investigations showed eosinophilia (AEC: 6912/cmm). ANA was negative. P-ANCA was positive. But sural nerve biopsy was negative for vasculitis. Treated as EGPA, she is improving gradually with oral prednisone. **Conclusion:** This was a case of EGPA with mononeuritis multiplex and bronchial asthma but with negative nerve biopsy.

INTRODUCTION

Churg-Strauss syndrome was described in 1951 by Churg and Strauss as a syndrome characterized by asthma and a "strikingly uniform clinical picture"

with 'fever and eosinophilia, and symptoms of cardiac failure, renal damage and peripheral neuropathy resulting from vascular embarrassment in various systems'¹. The histological lesions observed by Churg and Strauss in most of the affected sites were extremely severe; most were the autopsy cases. The knowledge on EGPA has recently evolved: antineutrophil cytoplasmic antibodies (ANCA) were found in significant proportion of patients. Multiple attempts have been made to provide classification criteria². This entity has now been recognized by the 2012 revised nomenclature for

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vasculitides as eosinophilic granulomatosis with polyangiitis (EGPA)³. It is an uncommon disease with an estimated annual incidence of 1–3 per million, predominantly affecting females and is characterized by asthma, peripheral and tissue eosinophilia, extravascular granuloma formation and vasculitis of multiple organ systems⁴. Here we report a case of 47 year old female with features of peripheral neuropathy, retinopathy, ulcerative lesion over skin, history of bronchial asthma

CASE HISTORY

47 year old lady from Ponnala, Kerala, with childhood deafness presented with 2 months history of decreased power of left foot and paresthesias over distal left lower limb. Following this 1 week later she had weakness of right foot and paresthesias over distal right lower limb. There was no proximal muscle involvement. She also had 1 month history of left hand paresthesias and 3 weeks history of blurred vision in the left eye. She also had bronchial asthma and allergic rhinitis for 2 years, with frequent exacerbations.

She also had history of alopecia. Diabetes mellitus was detected 2 months ago. Her clinical examination revealed, healed ulcer of 1 cm x 1 cm size over plantar aspect of left foot and healed scar of 4 cm size just above left lateral malleolus (fig 1 & 2).



Fig 1: healed ulcer of 1 cm x 1 cm size over plantar aspect of left foot



Fig 2: healed scar of 4 cm size just above left lateral malleolus

Vitals were normal. All peripheral pulses were well felt. Vision was 6/9 in right eye and 6/60 in left eye. Left eye fundus showed paler optic disc and arteriosclerotic changes. There was no papilloedema. Bilateral sensorineural hearing loss was present. Other cranial nerves were normal. She had bilateral foot drop. Bulk of muscles was lower in left leg than right. There was atrophy of thenar muscles in left hand. Power was grade 1/5 in ankle-dorsiflexors, plantar flexors, evertors, invertors bilaterally. Power in upper limbs was normal. Plantar reflex and ankle jerks were absent bilaterally. Superficial sensations were decreased bilaterally below ankle and palmar aspect of lateral 3 and half digits in left hand. Vibration sense and proprioception were decreased till ankle joint bilaterally. Gait was high stepping. There was no peripheral nerve thickening. Examination of cardiovascular, respiratory and abdominal systems was normal. Investigations showed WBC count of 14,400/cmm, Neutrophils 49% and Eosinophils 48%. Absolute eosinophil count: 6912/cmm, ESR: 63mm/1hr Urine 24 hr proteins were 330mg/dl. Hepatic and renal function tests were normal. Chest X-ray, ECG were normal. Nerve conduction study done in median, ulnar, peroneal, tibial & sural nerves revealed moderate to severe, asymmetrical, predominantly motor, axonal polyneuropathy. MRI spine showed diffuse disc bulge at L4-L5 intervertebral disc causing indentation on anterior thecal sac. MRI brain was normal. ANA, anti ds-DNA, c-ANCA were negative but p-ANCA was positive (51.98; positive >12). But sural nerve biopsy showed no vasculitis or collections of eosinophils in the specimen sent. On the basis of clinical features and p-ANCA positivity diagnosis of EGPA was made. She was started on steroids and is improving gradually.

DISCUSSION

Churg-Strauss syndrome is predominantly small vessel vasculitis. It affects arteries and veins and sometimes median vessels. It affects predominantly upper respiratory tract characterized by the presence of allergic disease, typically asthma or allergic rhinitis which may last for months to several years⁵. This is considered as the prodromal phase.

There is an eosinophilia tissue infiltration phase, in which high peripheral eosinophilia may occur and tissue infiltration by eosinophils is observed in the lung, gastrointestinal tract and other tissues.

This is followed by a vasculitic phase, in which systemic necrotizing vasculitis affects a large number of organs ranging from the heart and lungs to peripheral nerves and skin.

In our patient, symptoms of upper respiratory tract involvement in the forms allergic Rhinitis and bronchial asthma were present for the past 2 years. She

had marked peripheral blood eosinophilia and her absolute eosinophil count was 6912/mm³ with elevated ESR but we could not demonstrate any tissue infiltration by Eosinophils. Since 2 months she developed features of moderate to severe asymmetrical predominantly motor axonal polyneuropathy. Her 24 hour serum protein was high indicating of glomerular involvement. Healed ulcers over the plantar aspect of the left foot and over the left leg above the lateral malleolus suggest the possibility cutaneous vasculitis. Though one can argue that the ulcer over the plantar aspect can also be trophic ulcer due to sensory neuropathy, the site of the plantar ulcer is not typical of trophic ulcer. The pattern of peripheral neuropathy is suggestive of a vasculitic neuropathy similar to mononeuritis multiplex. Sural nerve biopsy did not reveal any evidence of vasculitis or eosinophilic infiltration, possibly because that segment of the nerve may not be affected by the disease as the symptoms are predominantly motor.

Her vasculitis work up showed P-ANCA positivity; DNA, Anti DNA and C-ANCA were negative. Other hyper-eosinophilic disorders like LOEFFLER'S Syndrome, hypereosinophilic syndrome, eosinophilic gastroenteritis, chronic eosinophilic pneumonia and eosinophilic leukemia were excluded.

She also had pale disc with loss of vision in the left eye which was gradual in onset suggestive of optic neuropathy. In Churg-strauss syndrome, optic neuropathy is likely to be due to ischaemic optic neuropathy and damages nerve cells between the retinal ganglion cells and the lateral geniculate body (anterior visual system). It is difficult to state whether it is anterior ischemic optic neuropathy or posterior ischemic optic neuropathy. Apart from Churg-Strauss syndrome, both anterior and posterior arteritic ischemic neuropathy is seen in inflammatory diseases of the blood vessels like giant cell arteritis, polyarteritis nodosa, Wegener's granulomatosis and Rheumatoid arthritis^{6,7}. Now arthritic ischemic neuropathy is related to poor circulation in the optic nerve head, elevated intraocular pressure, high cholesterol levels, hypercoagulable states, in hypertension due to bleeding or cardiac arrest. Though she is diabetic, her respiratory symptoms with peripheral blood eosinophilia and P-ANCA positivity favour Churg-strauss syndrome.

CONCLUSION

In conclusion, this was a case of EGPA with characteristic features of mononeuritis multiplex, bronchial asthma

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