Case Report

A rare case of Rhupus syndrome with Hashimoto's thyroiditis, associated adverse effect of drugs and incidental findings

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Abstract

The term Rhupus is coded for the individuals who have rheumatoid like arthritis with erosions and fulfil the criteria for both rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). Abnormal Th2 cell plays a central role in SLE while Th1 participate in RA. Thus the overlap of SLE and RA has a very low incidence (0.01%-0.2%) in patient with arthritis. This 40-year-old male patient presented with complains of severe multiple joint pain with progressive deformities, diminished vision from last 2 months, redness on back, headache and swelling over scalp. As this patient had history of severe trauma that was considered as a precipitating factor for ongoing chronic inflammatory disorder. Posterior subcapsular cataract was explained by prolong use of systemic steroid and rashes on the lower back due to sulfasalazine. Radioimaging study revealed arachnoid cyst and calcified projection arising from outer table of skull, which was kept under observation.

Keywords: Rheumatoid arthritis, Rhupus syndrome, systemic lupus erythematosus

Introduction

The term Rhupus is coded for the individuals who have rheumatoid like arthritis with erosions and fulfil the criteria for both Rheumatoid arthritis (RA) and systemic Lupus Erythematosus (SLE).[1] The definition of Rhupus syndrome remains controversial, as the immunopathological process of SLE is considered to be the exact opposite of RA. Abnormal Th2 cell plays a central role in SLE while Th1 participate in RA.[2] Thus the overlap of SLE and RA has a very low incidence (0.01%-0.2%) in patient with arthritis.[3] Rhupus syndrome is a rare disorder and very less number of cases have been reported. We hereby report a case of Rhupus syndrome with hashimoto's thyroiditis, associated adverse drug reactions and incidental findings of large arachnoid cyst in the posterior fossa of brain with calcified projection arising from the outer table of the skull.

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Case Report

A 40-year-old male patient presented with complaints of severe multiple joint pain with progressive deformities, diminished vision from last 2 months, redness on back, headache and swelling over scalp from last 2 weeks. This patient was apparently well 4 years before this presentation, when he first developed the symptoms of pain in the multiple joint. His RA FACTOR and ANTI Cyclic Citrulinated Peptide (CCP) was negative with raised TSH, ESR and CRP. A diagnosis of inflammatory polyarthritis, hashimoto's thyroiditis was made and he was treated with prednisolone and thyroxin. One year before this presentation he was admitted to this hospital; with complains of fever 4-5 days, cough and multiple joint pain

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and fine crepts on left side of the chest. He was then started treatment with short course of antibiotic with Sulfasalazine 1 gm, hydroxyxchloroquine sulfate (HCQS) along with thyroxine and prednisolone in tapering dose. He had the past history of trauma 10 years before the presenting illness for that he had multiple sutures over right leg and was hospitalised for 20 days. He was currently on drugs including sulfasalazine, HCQS, prednisolone and thyroxin. Examination of joints revealed polyarticular involvement including shoulder, knee, elbow, metacorpophalyngeal joints, proximal interphalyngeal joints and wrist joint with boutonniere deformity of first finger and Z deformity of left fifth finger were present [Figure 1a]. On the back, there was diffuse erythematous skin lesion. Oral ulceration was present on buccal mucosa and over the tongue. There was multiple irregular swelling over his scalp which was further examined by shaving his head and it was bony in consistency. Examination of eye revealed posterior subcapsular cataract. X-ray of the hand revealed erosive arthritis [Figure 1b]. For evaluation of headache and scalp swelling, radioimaging was done, which showed large arachnoid cyst involving posterior fossa [Figure 2a], hypoplastic right transverse sinus [Figure 2b] and focal calcified projection arising from outer table [Figure 3] of skull causing indentation over the scalp tissue. Two differential diagnoses were considered for this patient. First RA is a chronic inflammatory disease of unknown etiology marked by a symmetric, peripheral polyarthritis. It is the most common form of chronic inflammatory arthritis and often result in joint damage and physical disability. This patient fulfilled the new 2010 ACR-EULAR criteria[1] for RA by score of 7 by presence of >10 joints involvement (+5), raised ESR, CRP (+1) and duration >6 weeks (+1). Second SLE is an autoimmune disease in which organ and cells undergo damage initially mediated by tissue bonding autoantibodies and immune complex. Most people with SLE have intermittent polyarthritis, most commonly in hands, wrists, and knees. Joint deformity develops in only 10%. This patient fulfils the systemic lupus international collaboration clinic criteria[1] (SLICC) for SLE by score of 5 by presence of oral ulcers, anemia, sinuvitis, ANA positive by immunoassay, dsDNA positive. As this patient fulfilled the criteria for both SLE and RA, so he was coded as having both diseases that is 'RHUPUS'. The diminution of vision was explained by chronic use of steroid leading to posterior subcapsular cataract. Recently, developed rashes on the back of the skin were considered as drug reaction which was subsided after withdrawing sulfasalazine. The patient also complains of difficulty in vision with the use of drug HCQS, although macular oedema was not detected due to hazy media and considering patient's vision HCQS was also withdrawn and azathioprine was added. So he was finally diagnosed as a case of Rhupus with bilateral posterior subcapsular cataract, hashimoto's thyroiditis, adverse drug reaction and incidental findings of posterior fossa arachnoid cyst, hypoplastic right transverse sinus and calcified projections arising from outer table of the skull projecting into the scalp tissue. The arachnoid cyst found in the posterior fossa of the brain was not causing any sign and symptoms of compression, so it was considered as the incidental finding. Focal calcified projection arising from outer table of the skull kept under observation for further

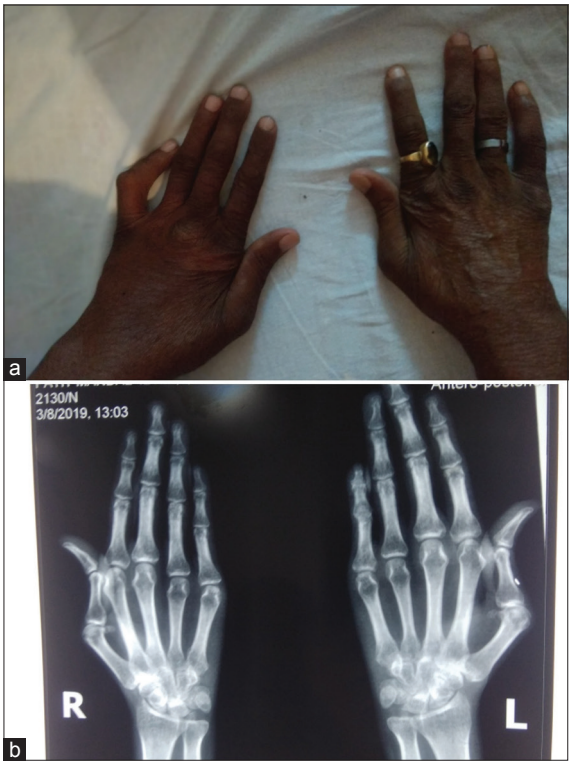

Figure 1: (a) Boutonniere deformity of first finger and Z deformity of left fifth finger were present. (b) X-ray of the hand revealed erosive arthritis

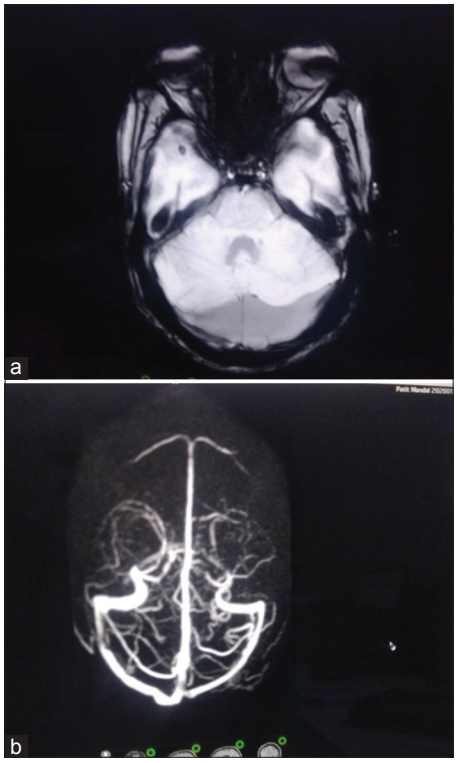

Figure 2: (a) Large arachnoid cyst involving posterior fossa. (b) Hypoplastic right transverse sinus

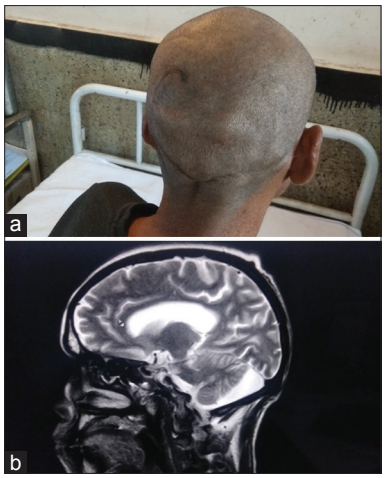

Figure 3: (a) Palpable bony swelling on the back of the head. (b) Radio imaging showing focal calcified projection arising from outer table of skull

progression and evaluation. This patient improved gradually, his skin rashes subsided and was able to walk without pain and support before discharge from this hospital and on follow-up of 6 months his focal calcified projection got subsided on its own.

Discussion

This patient on evaluation had features of both RA and SLE. As this patient had severe trauma that was considered as a precipitating factor for ongoing localized chronic inflammatory disorder.[4] Only one case report of huge arachanoid cyst in patient of RA was found.[5] Very few data are available concerning Rhupus syndrome treatment, and all the data that do exist are based on a few case series report.[6] The prognosis of Rhupus syndrome often depends on the severity of vital organ involvement, but is typically better than SLE and worse than RA.[7]

Conclusion

This case was unique in its presentation and it was undiagnosed for years. So, we would suggest early diagnosis and management of inflammatory polyarthritis which prevents joint deformities, once happened is irreversible. Care must be taken to rule out other disorders with symptoms that overlap, associated conditions like thyroid disorders and adverse effect of drugs.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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