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A CASE REPORT ON RHEUMATOID ARTHRITIS ASSOCIATED INTERSTITIAL LUNG DISEASE IN A PATIENT INCIDENTALLY DIAGNOSED WITH POLAND SYNDROME

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ABSTRACT About 1/3rd of rheumatoid arthritis patients are associated with pulmonary involvement. Most common being pleuritis with or without exudative effusion followed by interstitial lung disease. About 70-80% patients are positive for rheumatoid factor. Most common HRCT pattern include usual interstitial pneumonia (UIP) followed by Non specific Interstitial pneumonia (NSIP). Poland syndrome is a rare congenital Anomaly characterised by unilateral absence of sternocostal head of pectoralis major muscle and pectoralis minor muscle frequent association with or without ipsilateral upper limb brachysyndactyly. Hence, we present a rare occurrence of rheumatoid arthritis associated interstitial lung disease with Poland syndrome In the following case report.

KEYWORDS : Rheumatoid arthritis, interstitial lung disease, Poland syndrome, usual interstitial pneumonia.

INTRODUCTION

Poland syndrome is a rare congenital anomaly characterized by unilateral chest wall hypoplasia with or without brachysyndactyly[1]. Proposed mechanism is due to hypoplasia of the subclavian artery or its branches.Few case report shows Poland syndrome associated with leukemia, carcinoma of the hypoplastic breast[2]. Poland syndrome has been hypothesized to have a genetic association due to deletion mutations of genes. Rheumatoid arthritis (RA) is a systemic inflammatory disorder,involves class II major histocompatibility complex (MHC) region,human leukocyte antigen (HLA)-DRB1 allele. Other studies found that MUC5B variant has been associated with RA-ILD (particularly those with UIP) pattern, as well as fibrotic hypersensitivity pneumonitis [3,4].In addition, a study using whole exome sequencing found that patients with RA-ILD demonstrated an excess of mutations in TERT, RTEL1, PARN and SFTPC [5]

Case

A 60 year old male,who is working in provisional store was admitted in general surgery ward planned for hemorrhoidectomy. Patient was referred to repiratory medicine OPD for routine fitness for surgery, who had no respiratory complaints, vitals are stable. On examination Chest wall was asymmetrical, atrophy of right sided breast and pectoralis muscle(Figure 1). On auscultation-bilateral Velcro crackles present. Chest radiograph revealed reticular opacities(Figure 2). HRCT revealed UIP pattern with absent pectoralis major muscle(Figure 3A & 3B). Pulmonary function test revealed restrictive pattern. Rheumatoid factor and Anti CCP is positive, ANA is negative. Patient was diagnosed as Rheumatoid Arthritis associated Interstitial Lung Disease(RA ILD) and started on immunomodulators.

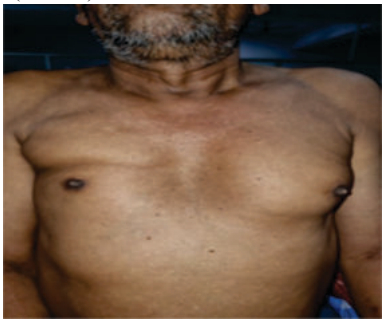

Fig 1- Atrophy Of Right Sided Breast And Pectoralis Muscle

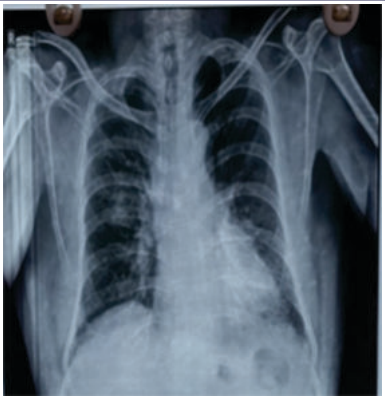

Fig 2-chest Radiography Bilateral Lower Zone Reticulations Present(left>right)

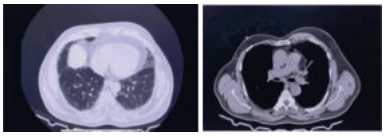

Fig 3a & 3b Ct Chest- Bilateral Lower Lobe Sub Pleural Fibrosis Absence Of Pectoralis Muscle Noted On Right Side

DISCUSSION

RA-ILD in a patient with Poland syndrome is a rare scenario. This intriguing convergence of congenital malformation and autoimmune disease presents a unique scenario. The monozygotic twin patients described by Vaccari et al. shared a heterozygous chromosome 11q12.3 deletion including genes involved in cellular growth, differentiation, and apoptosis through HRAS signaling pathway.Large deletion of chromosome 6q21-q22.1 was recently reported in a patient characterized by mental disability and Poland syndrome.Unilateral hyperlucency, which is supposed to be there in patients with Poland syndrome was masked in our case due to the reticulations and bronchiectatic changes[6,7]

CONCLUSION

The coexistence of Poland syndrome and RA-ILD raises questions about potential shared genetic or immunological factors contributing

to both conditions and prompts us to delve into the complexities of this intersection. In this modern era of imaging, the importance of clinical examination and comprehensive evaluation even in the absence of overt musculoskeletal complaints has been emphasized. Since most patients are asymptomatic and it is just a cosmetic problem. Poland syndrome is underreported, hence the exact incidence is not known.

REFERENCES

1. Ravitch, Mark M. "Congenital deformities of the chest wall and their operative correction." (1977).
2. Maria Victoria Romanini, Maria Grazia Calevo, Aldamaria Puliti, Carlotta Vaccari, Maura Valle, Filippo Senes, Michele Torre,Poland syndrome: A proposed classification system and perspectives on diagnosis and treatment,Seminars in Pediatric Surgery,Volume 27, Issue 3,2018,Pages 189-199,ISSN 1055-8586,https://doi.org/10.1053/j.sempedsurg.2018.05.007.
3. Dawson JK, Fewins HE, Desmond J, et al. Fibrosing alveolitis in patients with rheumatoid arthritis as assessed by high resolution computed tomography, chest radiography, and pulmonary function tests. Thorax. 2001;56:622–627.
4. Kadura S, Raghu G. Rheumatoid arthritis-interstitial lung disease: manifestations and current concepts in pathogenesis and management. Eur Respir Rev. 2021 Jun 23;30(160):210011. doi: 10.1183/16000617.0011-2021. PMID: 34168062; PMCID: PMC9489133.
5. Juge PA, Borie R, Kannengiesser C, et al. . Shared genetic predisposition in rheumatoid arthritis-interstitial lung disease and familial pulmonary fibrosis. Eur Respir J 2017; 49: 1602314.
6. Bodolay E, Szekanecz Z, Devenyi K, et al. Evaluation of interstitial lung disease in mixed connective tissue disease (MCTD) Rheumatology (Oxford) 2005;44:656–661.
7. Carmona L, Gonzalez-Alvaro I, Balsa A, et al. Rheumatoid arthritis in Spain: occurrence of extra-articular manifestations and estimates of disease severity. Ann Rheum Dis. 2003;62:897–900.