

**ATOLL SIGN IN CRYPTOGENIC ORGANIZING PNEUMONIA**Dharani M<sup>1</sup>, Ketaki Utpat<sup>2</sup>, Unnati Desai<sup>3</sup>, Jyotsna Joshi<sup>4</sup><sup>1</sup>Resident Doctor, Department of Pulmonary Medicine, TNMC BYL Nair Charitable Hospital, Mumbai, Maharashtra.<sup>2</sup>Assistant Professor, Department of Pulmonary Medicine, TNMC BYL Nair Charitable Hospital, Mumbai, Maharashtra.<sup>3</sup>Associate Professor, Department of Pulmonary Medicine, TNMC BYL Nair Charitable Hospital, Mumbai, Maharashtra.<sup>4</sup>Professor and HOD, Department of Pulmonary Medicine, TNMC BYL Nair Charitable Hospital, Mumbai, Maharashtra.**ABSTRACT****BACKGROUND**

Cryptogenic Organizing Pneumonia (COP) is one of the major idiopathic interstitial pneumonia (IIP) according to revised American Thoracic Society/ European Respiratory Society (ATS/ERS) classification of IIP.<sup>1</sup> Clinically, these patients present mainly with cough and dyspnoea on exertion. However, patients presenting with haemoptysis is very rare. COP has a wide range of radiological manifestations ranging from peripheral consolidation to very rarely nodular opacities. Hereby, we are presenting a prototypical case of COP presenting with a classical Atoll sign.

**KEYWORDS**

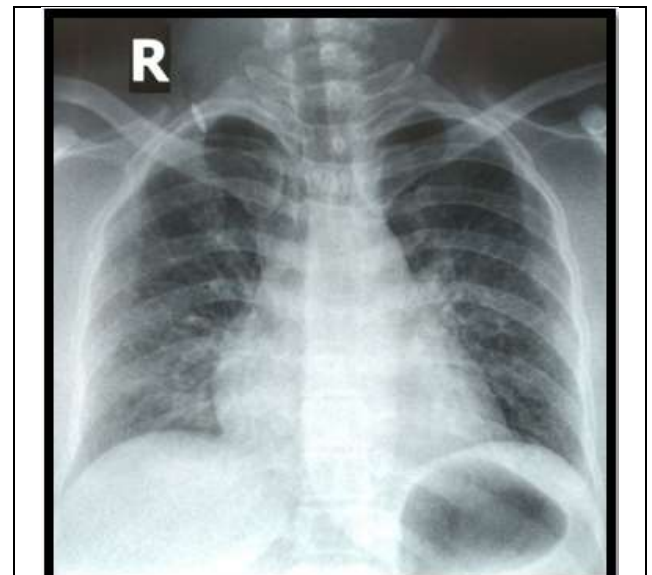
Cryptogenic Organizing Pneumonia (COP), Idiopathic Interstitial Pneumonia (IIP).

**HOW TO CITE THIS ARTICLE:** Dharani M, Utpat K, Desai U, et al. Atoll sign in cryptogenic organizing pneumonia. J. Evid. Based Med. Healthc. 2019; 6(1), 68-70. DOI: 10.18410/jebmh/2019/13

**BACKGROUND**

A 35-year-old lady was symptomatic since 1 year with complaints of streaky haemoptysis, managed symptomatically in private hospital. However, in view of non-relief, patient followed up with us. There were no other respiratory complaints in the form of dyspnoea on exertion, chest pain or fever. Her past, personal or family history was not contributory. Physical examination revealed no abnormality except for grade 1 clubbing. Haematological and biochemical investigations were within normal limits. Chest radiograph (CXR) showed bilateral reticulonodular opacities. (Figure 1). Spirometry was suggestive of restrictive abnormality with forced expiratory capacity (FVC) of 2.08 L (73% predicted), forced expiratory volume in one second (FEV1) of 1.98L (81% predicted) and FEV1/FVC ratio of 100%. High Resolution Computed tomography (HRCT) of chest (Figure 2) revealed presence of characteristic "Atoll sign" in the form of ill-defined bronchocentric areas of ground glass attenuation with surrounding halo of consolidation, suggestive of non-fibrosing interstitial lung disease (ILD), organizing pneumonia (OP) pattern. Sputum Cartridge Based Nucleic Acid Amplification Test (CBNAAT) examination result was mycobacterium tuberculosis (MTB) not detected. Test for anti-nuclear antibodies (ANA) and anti-neutrophil cytoplasmic antibodies (ANCA) were negative.

A Transbronchial Guided Lung Biopsy (TBLB) done via a flexible Fibre Optic Bronchoscope (FOB) tissue histopathology showed intra-alveolar Masson bodies with hook like configuration in peribronchiolar alveoli, fibroblasts and granulation tissue suggestive of organizing pneumonia (OP) (Figure 3). Special stains were negative for infection and malignancy. Hence a diagnosis of COP was made based on clinical, radiological and histopathological correlation. Patient was started on oral steroids tablet prednisolone 1 mg/kg body weight per day followed by tapering doses. She improved clinically, radiologically and spirometry showed an improvement in FVC of 300 ml.



**Figure 1. Chest X-Ray Showing Bilateral Reticulonodular Opacities**

*Financial or Other, Competing Interest: None.*

*Submission 28-11-2018, Peer Review 17-12-2018,*

*Acceptance 31-12-2018, Published 07-01-2019.*

*Corresponding Author:*

*Dr. Ketaki Utpat,*

*Assistant Professor,*

*Department of Pulmonary Medicine,*

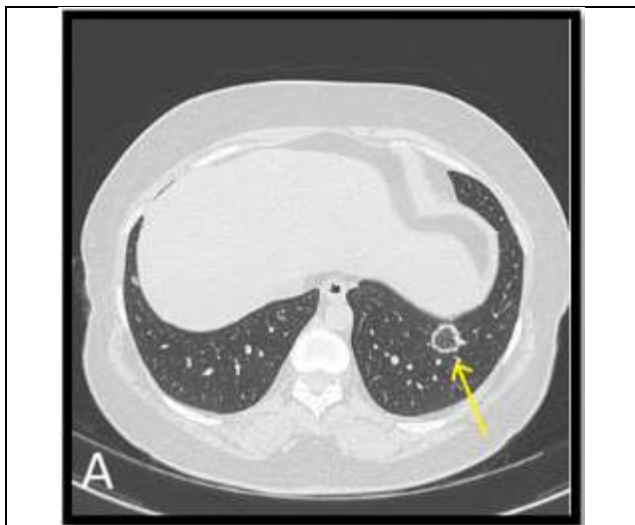
*TNMC & BYL Nair Hospital,*

*Mumbai, Maharashtra.*

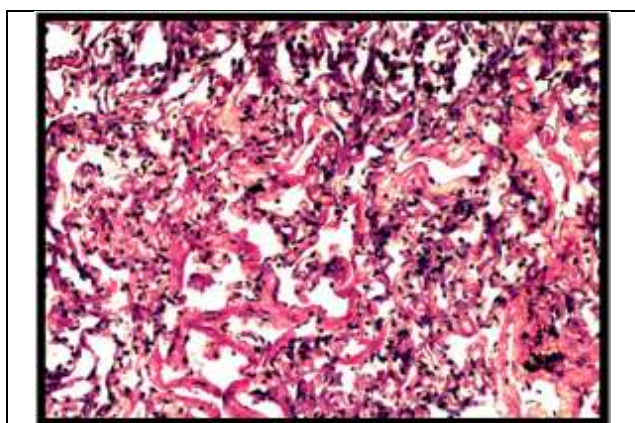
*E-mail: drketakibarve@gmail.com*

*DOI: 10.18410/jebmh/2019/13*





**Figure 2. High Resolution Computed Tomography- Axial view Showing Classical Atoll Sign in Left Lower Lobe**



**Figure 3. TBLB Histopathology showing Intra-Alveolar Masson Bodies with Hook Like Configuration in Peribronchiolar Alveoli, Fibroblasts and Granulation Tissue Suggestive of Organizing Pneumonia (OP)**

## DISCUSSION

The term 'Atoll sign' was coined by Zompatori.<sup>1</sup> However, ground-glass opacification surrounded by ring shaped or crescentic opacities were described first by Voloudaki in the setting of OP.<sup>2</sup> The word Atoll means an island consisting of a lagoon surrounded by circular coral reef. It is derived from the Maldivian word 'atholhu'. On histopathological examination, the central ground-glass opacification is a tell-tale of inflammation of septa with associated cellular debris within the airspaces whereas peripheral ring-shaped or crescentic opacity signifies OP in the alveolar ducts.<sup>3</sup> COP is one of the major IIP according to revised ATS/ERS classification of IIP.<sup>4</sup> Although the pulmonary lesions in COP are mainly intra-alveolar, it is included in the classification of the IIP, mainly due to: 1) its idiopathic nature; 2) confusion with other forms of IIPs, especially when there is progression to fibrosis; and 3) interstitial inflammation in the involved areas on histopathological examination.<sup>5</sup> These patients usually present with subacute onset of symptoms in the form of cough, dyspnoea, fever and pleuritic chest pain.

Patients presenting only with haemoptysis is very rare which was seen in our case. The symptomatology of COP and secondary OP is similar.

HRCT Chest helps in better delineation of lesions. COP can present as focal subpleural or peribronchovascular consolidations which are usually bilateral and asymmetrical. The reverse halo sign is an area of ground glass opacity surrounded by a crescent or ring of consolidation with smooth or spiculated borders. It can rarely present as pure nodular form which have solid, mixed density or rarely ground glass nodules with scattered or peribronchovascular distribution. Other features include peribubular abnormalities presenting as curved or arcade-like bands of parenchymal consolidation with blurred borders or thick radial bands of consolidation (>8 mm) extending towards the pleura or crazy paving sign or progressive fibrotic pattern.<sup>6</sup> Features pointing more towards invasive fungal infections rather than organizing pneumonia include outer rim thickness of more than 1 cm, presence of reticulation inside the reverse halo sign and associated pleural effusion.<sup>7</sup> To diagnose cryptogenic form, other secondary causes of OP including drugs and substance abuse including amiodarone, b-blockers, cocaine abuse, systemic inflammatory diseases like rheumatoid arthritis, polymyositis/dermatomyositis, polymyalgia rheumatica, solid tumours like carcinoma colon or breast, hematologic malignancies like non-Hodgkin's lymphoma, renal transplantation and infections have to be ruled out.<sup>8</sup> Mild to moderate restrictive ventilatory defect is the most common finding on lung function test in these patients. There could be associated mild hypoxemia at rest or on exercise.<sup>5</sup> Histopathological examination of the tissue shows intra alveolar fibroblasts, myofibroblasts and granulation tissue as was seen in our case too.<sup>9</sup> The modus operandi for obtaining a lung biopsy tissue can be a TBLB, CT guided biopsy, thoracoscopic lung biopsy or an open lung biopsy.

## CONCLUSION

In our case, we were successful in attaining the diagnosis based on a TBLB which is the least invasive of these. The diagnosis of COP mainly depends on typical clinical, radiological and histopathological features and also by excluding other secondary causes. Once diagnosis is made by this multidisciplinary approach, steroids are the prime modality of treatment. However spontaneous resolution is also reported.<sup>10</sup> Our patient was thus a rare case of COP with an unusual clinical presentation demonstrating the classical Atoll sign on imaging.

## REFERENCES

- [1] Zompatori M, Poletti V, Battista G, et al. Bronchiolitis obliterans with organizing pneumonia (BOOP), presenting as a ring-shaped opacity at HRCT (the atoll sign). A case report. *Radiol Med* 1999;97(4):308-310.
- [2] Voloudaki AE, Bouros DE, Froudarakis ME, et al. Crescentic and ring-shaped opacities. CT features in

- two cases of bronchiolitis obliterans organizing pneumonia (BOOP). *Acta Radiol* 1996;37(6):889-892.
- [3] Walsh SLF, Robertson BJ. The atoll sign. *Thorax* 2010;65(11):1029-1030.
- [4] Travis WD, Costabel U, Hansell DM, et al. An official American Thoracic Society/European Respiratory Society statement: update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. *Am J Respir Crit Care Med* 2013;188(6):733-748.
- [5] Cordier JF. Cryptogenic organising pneumonia. *Eur Respir J* 2006;28(2):422-446.
- [6] Baque-Juston M, Pellegrin A, Leroy S, et al. Organizing pneumonia: what is it? A conceptual approach and pictorial review. *Diagn Interv Imaging* 2014;95(9):771-777.
- [7] Marchiori E, Marom EM, Zanetti G, et al. Reversed halo sign in invasive fungal infections: criteria for differentiation from organizing pneumonia. *Chest* 2012;142(6):1469-1473.
- [8] Drakopanagiotakis F, Paschalaki K, Abu-Hijleh M, et al. Cryptogenic and secondary organizing pneumonia: clinical presentation, radiographic findings, treatment response, and prognosis. *Chest* 2011;139(4):893-900.
- [9] Colby TV. Pathologic aspects of bronchiolitis obliterans organizing pneumonia. *Chest* 1992;102(1 Suppl):38S-43S.
- [10] Cagnina RE, Conces MR, Stoler MH, et al. Reversed halo sign. A case of cryptogenic organizing pneumonia with spontaneous resolution. *Am J Respir Crit Care Med* 2015;192(1):109-110.