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Effect of Disodium Etidronate in Patient with Pulmonary Alveolar Microlithiasis

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Introduction: Pulmonary alveolar microlithiasis (PAM) is a rare autosomal recessive disease.. It is characterized by the widespread intraalveolar accumulation of calculi called microliths. There is no definitive treatment available to prevent the progression of PAM. Lung transplantation has been used to treat patients with end-stage. Disodium etidronate has been administered due to its alleged calcium phosphate precipitation-reducing effect, however, its effectiveness is controversial.

Case Presentation: A 13-year-old male with no remarkable medical history presented with a sore throat. Chest X-ray performed because of prolonged symptoms. He referred to pediatric pulmonology department because of the reticulonodular opacities in chest X-ray. Patient's physical examination was normal and he had no clinical complaints. Chest X-ray revealed reticulonodular opacities that had a greater distribution in the middle and lower lung zones. Pulmonary function test results showed a mild restrictive pattern characterized by decreased forced vital capacity, decreased forced expiratory volume in 1 second. Findings on computed tomographic (CT) images were consistent with interstitial lung disease. The patient underwent open lung biopsy, and histopathologic analysis results confirmed a diagnosis of pulmonary alveolar microlithiasis.. The patient started disodium etidronate treatment. Sandstorm-like appearance on chest x-ray decrease and restrictive pattern in pulmonary function test improved after three months of treatment. There was no significant change in parenchymal findings on CT. In follow up concomitant thoracic CT and MRG performed. However, the micronodular pattern observed in CT was not present in MRG. Only peribronchial thickening was also detected in MRG.

Conclusion: There is no known effective treatment for PAM, with the exception of lung transplantation. As PAM progresses patients commonly develop dyspnea on exertion, other symptoms, including a cough, chest pain, hemoptysis, asthenia, and pneumothorax, have also been reported. Cyanosis and clubbing of the fingers may be identified in serious cases Number of the report describes the beneficial effects of long-term treatment with disodium etidronate but in a certain case, it seems ineffective. Our patient's chest X-ray and pulmonary function improved after three months of disodium etidronate treatment. However, there was no major change in CT. It is possible to observe parenchymal improvement in long term treatment. CT is a major tool for the detection and follows up the patients. When the harmful effects of radiation in childhood are considered, the necessity of alternative diagnosis and follow-up methods arises. We performed thoracic MRG in follow up however the parenchymal patterns of PAM could not be detected in MRG.

Keywords: Pulmonary alveolar microlithiasis, etidronet, pediatric