International Journal of Current Advanced Research
ISSN: O: 2319-6475, ISSN: P: 2319-6505, Impact Factor: SJIF: 5.995
Available Online at www.journalijcar.org
Volume 6; Issue 10; October 2017; Page No. 6993-6995
DOI: http://dx.doi.org/10.24327/ijcar.2017.6995.1058

Research Article

A NEPHROTIC SYNDROME REVEALING KARTAGENER'S SYNDROME COMPLICATED BY RENAL AMYLOIDOSIS

Nadia Kabbali[1,2]., Rim Mejbar[1]., Taoufiq Harmouch[3] and Tarik Sqalli[1, 2]

[1]Nephrology Department, Hassan II University Hospital, Fez, Morocco
[2]R.E.I.N Teamwork, Faculty of Medicine, Sidi Mohamed Ben Abdellah University, Fez, Morocco
[3]Pathology Department, Hassan II University Hospital, Fez, Morocco

A R T I C L E I N F O

Article History:

Received 26th July, 2017
Received in revised form 19th August, 2017 Accepted 25th September, 2017
Published online 28th October, 2017

Key words:

Primary ciliary dyskinesia, situs inversus, renal amyloidosis

A B S T R A C T

Kartagener's syndrome is a group of primary ciliary dyskinesia associating *situs inversus totalis,* bronchiectasis and recurrent chronic sinusitis. It's secondary to structural and functional ciliary abnormalities leading to impaired mucociliary clearance. It's a rare genetic disorder transmitted in autosomal recessive mode (Prevalence 1/10000 to 1/20000). We report a case of a 54 year-old-woman who was admitted for a nephrotic syndrome. Investigations revealed a Kartagener's syndrome complicated by a renal amyloidosis. It's a rare complication of Kartagener's syndrome resulting probably from bronchiectasis and chronic respiratory infections.

INTRODUCTION

Case report

A 54 year-old-woman was admitted in nephrology for a nephrotic syndrome. Her parents were first degree relatives. She was treated for high blood pressure, diabetis and gastric ulcer from several years. Since childhood, she had chronic cough, abundant bronchorrhea and recurrent episodes of respiratory tract infections. She was followed for asthma and was treated by bronchodilatator therapy. Physical examination showed a normal blood pressure (110/70 mmHg), rhoncus and crackles at pulmonary auscultation and edema of lower extremities. Dipstick urinalysis showed proteinuria without hematuria. She had 6g/day in 24-hour urine analysis. Other Laboratory findings revealed a serum creatinin at 186 µmol/l, urea at 13.9 mmol/l, proteins at 42g/l, albumin at 17g/l, sodium at 136 mmol/l, potassium at 3,6mmol/l, corrected calcium at 101 mg/l. Hepatitis B and C and HIV serologies were negative. Complement levels C3 and C4 were normal. Serum protein electrophoresis was normal. Complete blood count revealed a microcytic anemia with hemoglobin rate at 10,9g/dl, normal red blood cells and platelets rate. Chest radiograph showed dextrocardia, right lung atelectasis with tracheal attraction (Fig.1). Computed tomography showed bronchiectasis, right lung destruction and complete transposition of mediastinal and abdominal organs which confirmed *situs inversus totalis* (Fig.2a, b, c). She also had mucosal thickening of maxillary sinuses (Fig.2d). Renal biopsy was performed revealing renal amyloidosis (Fig.3).

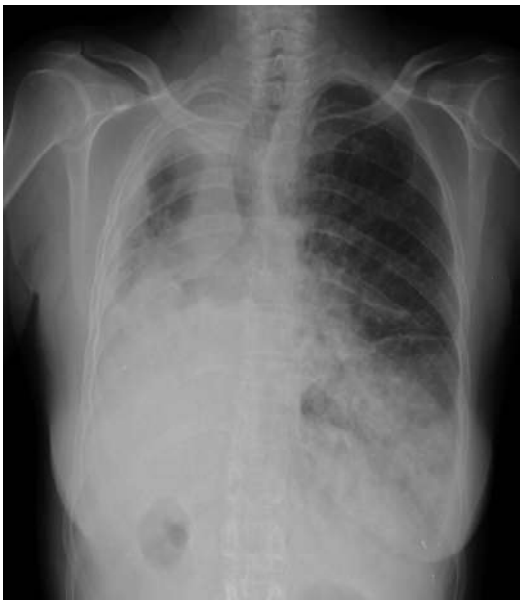

Fig 1 Anteroposterior chest X-ray showing intercostal right space narrowing, tracheal attraction and dextrocardia.

Given these data, the diagnosis of Kartagener's syndrome was concluded. It was not possible to study ciliary function or carry out genetic tests confirming the diagnosis because these laboratory tests are not available in our laboratories.

Corresponding author:* **Nadia Kabbali
Nephrology department, Hassan II university hospital, Fez, Morocco

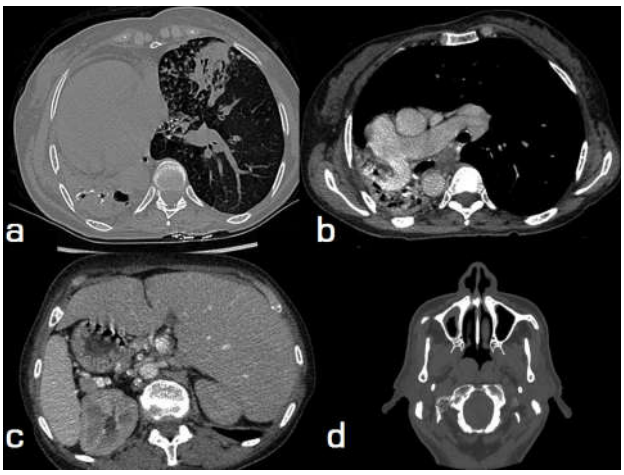

Fig.2 (a,b,c) High resolution computed tomography (HRCT) of the thorax showing *situs inversus totalis*, bronchiectasis with 'tree-in-bud' appearance in the upper left lobe suggestive of bronchiolitis, right lung atelectasis and bronchiectasis.
Fig.2 (d) HRCT showing a slight mucosal thickening of maxillary sinuses

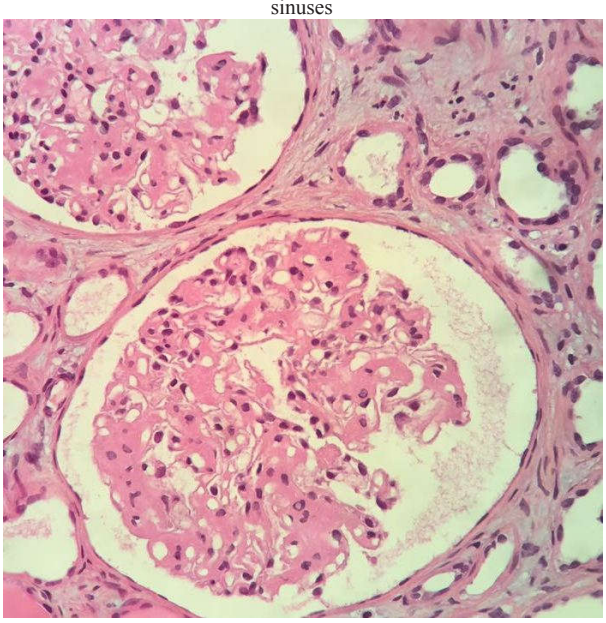

Fig 3 Pathological study of our patient renal biopsy showing congo red stained glomerular amyloid fibrils

Two months later, our patient was readmitted for digestive hemorrhage (hematemesis and melena). Oesophago-gastro-duodenoscopy was normal. Renal function has profoundly impaired and eventually, she progressed to end stage renal failure requiring hemodialysis.

DISCUSSION

Primary ciliary dyskinesia (PCD) is a rare recessive autosomal disease (Prevalence 1/10000 to 1/20000), characterized by ultrastructural and functional abnormalities of respiratory cilia, leading to impaired mucociliary motility and then abnormal ciliary clearance. Kartagener's syndrome is a form of PCD which associates *situs inversus totalis* (50% of PCD) [1,2].

PCD is primarily responsible for respiratory symptoms including upper and lower respiratory tract infections and reccurent rhinosinusitis progressing to bronchiectasis and sinonasal polyposis. However, other extrarespiratory symptoms and abnormalities can be encountered. Indeed, most affected male individuals have sterility due to immobility of spermatozoa, while women have a high risk of extrauterine pregnancies. In addition, cardiac malformations, ocular retinitis pigmentosa, chronic otitis media, conductive hearing loss, some cases of hydrocephalus and polycystic kidney disease have been described in association with PCD [1,3]. Diagnosis can be confirmed by performing genetic tests or by screening ciliary structure or function, while treatment is primarily supportive, mainly represented by respiratory kinesitherapy, to slow down the disease evolution [4].

AA amyloidosis is a rare complication of Kartagener's syndrome and is probably related to bronchiectasis. Four cases have been described in the literature. Three of them had a normal renal function [5-7] while the fourth has progressed to end stage renal failure such our patient [8]. Another case of chronic renal failure associated with Kartagener's syndrome have been reported, but its etiology is unkown because renal biopsy was not performed due to advanced renal failure [9]. Not only humans are concerned by this complication of Kartagener's syndrome. Bartolo reported a renal amyloidosis as a result of this syndrome in a dog [10].

Type AA of amyloidosis is certainly the form usually described in this pathology but it is not the only form presented in the literature. In fact, one case of transthyretin amyloidosis associated to Kartagener's syndrome has been reported [11]. Nephrotic syndrome in this pathology seems therefore to be essentially related to renal amyloidosis, however other renal disorders have been described: focal segmental glomerulosclerosis and mesangioproliferative glomerulonephritis [12,13].

CONCLUSION

Renal amyloidosis is a rare but serious complication of Kartagener's syndrome. It can rapidly progress to end stage renal failure. Early management is therefore necessary, hence the importance of earlier diagnosis. Even if treatment is essentially supportive, it can considerably halt the evolution to end stage renal failure. Therefore, we should be more vigilant about this diagnosis.

References

1. Escudier E, Tamalet A, Prulière-Escabasse V, Roger G, Coste M. Dyskinésie ciliaire primitive. Revue française d'allergologie et d'immunologie clinique 46 (2006) 530-537
2. Braun JJ, Boehm N, Metz-Favre C, Koscinski I, Teletin M, Debry C. Diagnosis of primary ciliary dyskinesia: When and how?. *Eur Ann Otorhinolaryngol Head Neck Dis*. 2017 May 10. pii: S1879-7296(17)30093-5.
3. Leigh MW, Pittman JE, Carson JL, Ferkol TW, Dell SD, Davis SD, Knowles MR, Zariwala MA. Clinical and genetic aspects of primary ciliary dyskinesia/Kartagener syndrome. *Genet Med*. 2009 Jul;11(7):473-87.
4. Skeik N, Jabr FI. Kartagener syndrome. Int J Gen Med. 2011; 4: 41–43.
5. El Houssni S, Laine M, Bziz A, Alhamany Z, Bourkadi JE, Bayahia R. Amylose rénale révélant un syndrome de Kartagener. *Néphrol ther* (2014).
6. Ozkok A, Keskinler MV, Eken E. et al. A rare complication of a rare disease: Amyloidosis in situs inversus. *Open Journal of Internal Medicine*, 2013, 3, 55-5.

7. Demir M, Kutlucan A, Sezer MT. Renal amyloidosis in a patient with Kartagener syndrome. *Ren Fail.* 2007; 29(1):117.
8. Osman EM, Abboud OI, Sulaiman SM, Musa AR, Beleil OM, Sharfi AA. End-Stage Renal Failure in Kartagener's Syndrome. *Nephrology Dialysis Transplantation*, Volume 6, Issue 10, 1 January 1991, Pages 747.
9. Imafuku H, Ogihara T, Kudo H, Hayashi K, Ohtake KI, Iyori S. Kartagener's syndrome associated with infundibular pulmonic stenosis, chronic renal failure and azoospermia: A Report of a Case. *JapJ Med* Vol 25,No 2 (May 1986).
10. Celona B, Crinò C, Bruno C, Di Pietro S, Giudice E. Renal amyloidosis associated with Kartagener syndrome in a dog. *Top Companion Anim Med*. 2017 Jun; 32(2):61-65.
11. Fritschka M, Schlegl M, Borges A, Werner M, Gebker R, Pieske B, Kelle S. Unusual case of ATTR amyloidosis with cardiac manifestation and situs inversus totalis. *Clin Res Cardiol*. 2017; 106(4): 311-316.
12. Vikrant S, Kumar S, Raina R, Sharma A. Nephrotic syndrome in a patient with situs inversus totali. *Clin Exp Nephrol* (2008) 12:215-218.
13. Krishnamurthy S, Choudhary B, Rajesh NG, Ramesh A, Srinivasan S. Kartagener syndrome associated with mesangioproliferative glomerulonephritis. *Paediatr Int Child Health* 2012;32:116-8

How to cite this article:

Nadia Kabbali *et al* (2017) 'A Nephrotic Syndrome Revealing Kartagener's Syndrome Complicated By Renal Amyloidosis', *International Journal of Current Advanced Research,* 06(10), pp. 6993-6995.
DOI: http://dx.doi.org/10.24327/ijcar.2017.6995.1058

****