CASE REPORT

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Renal Cell Carcinoma in A Patient with Kartagener Syndrome: First Case Report in English Language

Kartagener Sendromu Olan Bir Hastada Renal Hücreli Karsinom: İngilizce Yazılmış İlk Olgu Sunumu

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ABSTRACT

Cardiac and pulmonary anomalies are common among patients with situs inversus totalis. Renal anomalies, including renal agenesis, dysplasia, hypoplasia, ectopia, polycystic kidney, and horseshoe kidney have been reported. We report a case of renal cell carcinoma in a patient with situs inversus totalis (SIT). Our case represents the fourth case report of renal cell carcinoma in a patient with situs inversus totalis and to the best of our knowledge this is the first report in English language. Due to the higher frequency of cardiac, pulmonary, renal, and vascular anomalies the management of patients with situs inversus and urologic disease requires careful preoperative evaluation.

Key Words

Kidney neoplasms, situs inversus totalis, Kartagener syndrome, infertility, primary ciliary dyskinesia

ÖZET

Situs inversusu totalis hastalarında kardiyak ve pulmoner anomaliler sık görülmektedir. Renal agenezi, displazi, hipoplazi, ektopi, polikistik böbrek ve atnalı böbrek bildirilen renal anomaliler arasında sayılabilir. Bu olgu sunumuzda bir situs inversus totalis olgusunda saptanan renal hücreli karsinomu sunmayı amaçladık. Olgu sunumumuz, bu tip bir olguda bildirilmiş dördüncü, İngilizce dilinde ilk olgu sunumu olma özelliğini taşımaktadır. Situs inversus ve ürolojik hastalıkların beraber gözlendiği bu tip olgularda, yüksek oranda kardiyak, pulmoner, renal ve vasküler anomalilerin eşlik etmesi nedeniyle dikkatli bir preoperatif hazırlık yapılması önemlidir.

Anahtar Kelimeler

Böbrek neoplazmları, situs inversus totalis, Kartagener sendromu, infertilite, primer siliyer diskinezi

Introduction

Stewart was the first to describe a patient with situs inversus, chronic sinusitis and brochiectasis in 1904. In 1933 Maner Kartagener reported this condition as a congenital syndrome and since then it has been named after him. Kartagener syndrome is one of the most known types of the primary ciliary dyskinesia (PCD) syndromes and it shows an autosomal recessive inheritance. Camner et al. described ciliary dysfunction of spermatozoa in an infertile Kartagener Syndrome patient in 1975. A similiar defect effecting nasal bronchial cilia was later reported by Afzelius in the same group of patients (1,2,3).

In this case report, we discuss the clinical presentation and radiological work-up of a patient with situs inversus totalis (SIT) diagnosed with renal cell carcinoma (RCC) together with a review of the literature.

Case Report

A 52 year old male infertile patient admitted to our hospital for a routine clinical examination and underwent an abdominal USG after positive result in a direct coombs test. Abdominal USG revealed SIT and a right renal mass. The patient was consulted to our urology clinic after thorax X-ray, paranasal sinus and thorax CT were done with the suspicion of Kartagener syndrome.

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The patient had a history of infertility not solved by any treatment modality. Spermiogram revealed asthenoteratospermia (rapidly progressive+non progressive motility: 13.7%, normal morphology according to WHO: 16.7%). There were no other pathological findings in thorax X-ray and CT scan beside dextrocardia (Figure 1). Nasal polyps were detected in ethmoidal sinuses mostly at left side (Figure 2). Abdominal CT scan was performed to evaluate the renal anatomy and mass that was described in abdominal USG. Liver was found left sided (Figure 3) and 3 cm mass lesion was detected at lower pole of right kidney (Figure 4). Right partial neprectomy was planned. Right flank incision was made to reach the kidney and partial neprectomy was performed to a 3 cm lower pole mass. No complication occured perioperatively and the patient was discharged at postoperative 3. day.

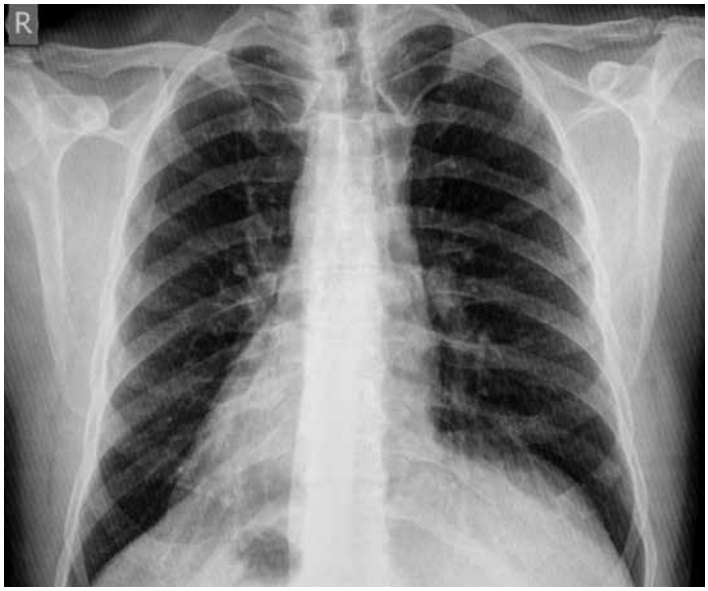

Figure 1. Thorax X-ray scan showing dextrocardia

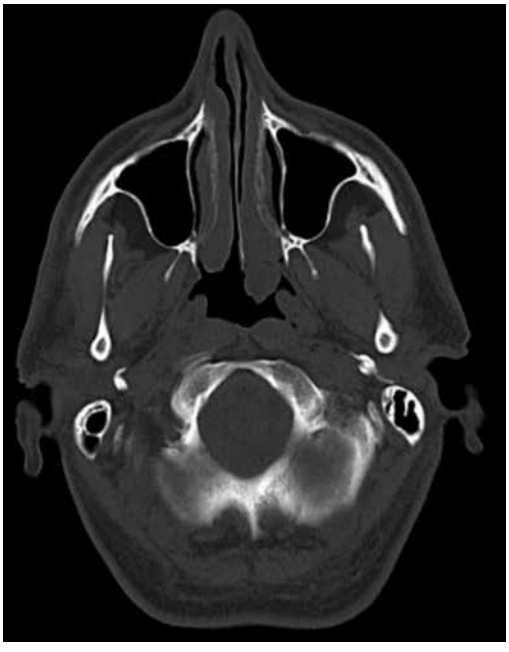

Figure 2. Nasal polyps are detected mostly lef sided on paranasal CT scan

Histopatological examination revealed Fuhrman grade 3 conventional type of clear cell renal cell carcinoma. Surgical margins were negative so the patient has been taken under routine control programme of renal cell carcinoma. Follow-up time reached 24 months postoperatively and no recurrence of kidney disease has been detected.

Discussion

There is an increased risk of cardiac and pulmonary diseases in patients with SIT. There are a few reports that renal anomalies can also be seen in these patients but there is no increased risk and anomaly detection rates are similar to normal population (4,5).

The first gene in which mutations were found to be associated with PCD was DNAI1 gene (6). The dynein intermediate gene DNAI1 is

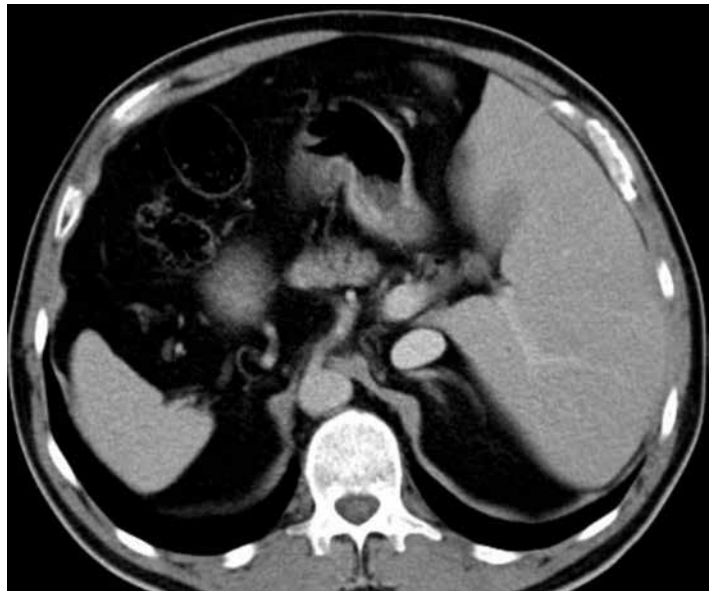

Figure 3. Abdominal CT showing left sided liver

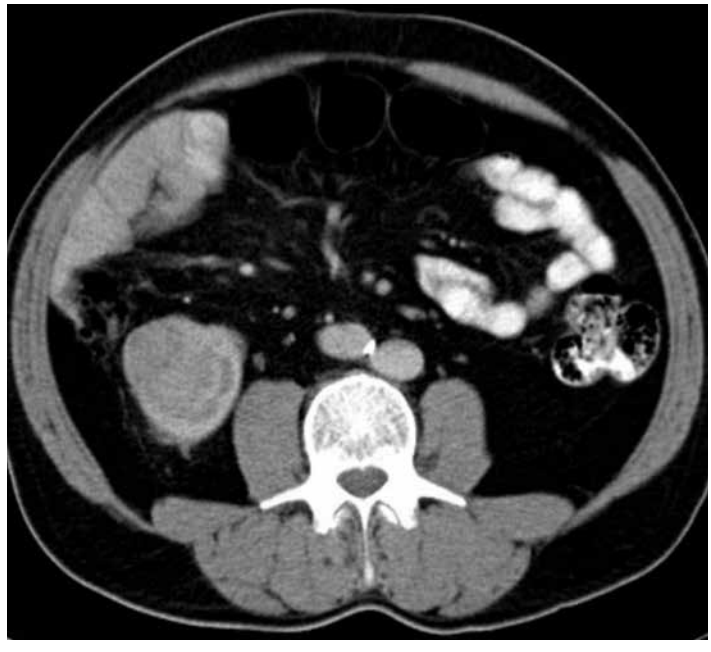

Figure 4. Abdominal CT revealed a right renal mass at lower pole

localized on 9p13-p21 and is composed of 20 exons (7). An additional dynein heavy chain gene (DNAH10) and a pseudogene (DNAH7p) were recently reported, along with a classification and chromosomal mapping of known human dynein heavy chain genes encoding a 699–amino acid protein (8).

Lots of genes related to sporadic and hereditary forms of RCC were reported but there is no evidence found that these genes may play a role in genetic development of Kartagener syndrome.

Our patient was diagnosed with SIT, nasal polyps, and finally infertility related to Kartagener syndrome.

There was SIT, nasal polyps and infertilty of Kartagener syndrome in our case. A right renal mass was incidentally found upon imaging. There are only three case reports of renal cell carcinoma concominant with situs inversus in literatüre (4,9,10). Our case is the first one in English language.

SIT does not have a surgical importance alone but concomitant congenital anomalies and abdominal pathologies can make it a surgical problem. Such a patient's preoperative evaluation with CT scan and magnetic resonance imaging can be helpful to see anatomical variations and determine surgical approach (11). In our patient we used CT scan to assess abdominal and renal anatomy and partial neprectomy was done without complication with an open surgical approach.

All three cases in literature were also treated with open surgery. We think that type of surgical approach is related to the time of publication because these reports are publicated in a period when laparoscopic urologic procedures were not commonly preferred although there were enough reports of laparoscopic procedures in general surgery literature (4,9,10,12,13).

Conclusion

It is very rare to find SIT and renal cell carcinoma together but when it is detected imaging modalities are important to assess abdominal and renal anatomy for determination of treatment choices. In the absence of renal and abdominal anomalies, laparoscopic or robot assisted renal surgeries can be treatment choices in suitable experienced centres.

Conflict of interest

There are no conflicts of interest.

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