Case Report:

Situs inversus totalis with azoospermia in a patient presenting with liver abscess

P. Mohan Rao,[1] A. Sridhar,[3] I.V. Renuka,[2] M. Venugopal,[2] C. Aparna[2]

Departments of [1]General Medicine, [2]Pathology, [3]Neurology, Guntur Medical College, Guntur

ABSTRACT

Situs inversus with dextrocardia is a rare congenital anomaly. Azoospermia and situs inversus may be encountered in ciliary dyskinesia syndromes. We report the case of a 30-year-old male who manifested *situs inversus totalis,* dextrocardia and azoospermia with maturation arrest at primary spermatogenesis who presented with liver abscess. The patient responded well to treatment with i.v. metronidazole and oral chloroquine.

Key words: *Situs inversus, Azoospermia, Immotile cilia syndrome, Infertility*

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INTRODUCTION

Situs inversus with dextrocardia has a prevalence of 1 in 10000 population.[1-3] It has been associated with Kartagener's syndrome which is a disorder of ciliary dyskinesia. Azoospermia is a disorder associated with Young's syndrome. Clinical presentation with a combination of these two features is rare with few such reports[1-3] being published till date. We report a case of situs inversus totalis with dextrocardia and azoospermia.

CASE REPORT

A 30 year-old-male farmer presented with fever and pain in the left hypochondrium of ten days duration. On examination apex beat was palpated in the 5th right intercostal space and a tender mass was felt in the left hypochondrium. He had a normal onset puberty and had normal libido and potency. He was married for the last 7 years and the couple were childless. Haematological, biochemical investigations were within normal limits. Nasal mucociliary clearance measured by the saccharin test was less than sixty minutes. Computed tomography (CT) of the chest and abdomen (Figure 1) showed a right lung with two lobes, a left lung with three lobes and a left-sided pleural effusion. Liver was found to be on the left side and an abscess measuring 6 × 5 cm was noted. There was dextro-rotation of all the organs. Ultrasonography of scrotum was normal. Echocardiography showed dextrocardia with normal atrio-ventricular, ventriculo-atrial

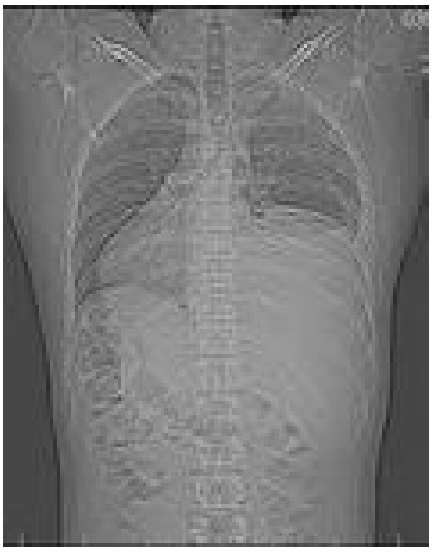

Figure 1: CT topogram showing *situs inversus totalis*

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Corresponding author: Dr P. Mohan Rao, Professor and Head, Department of General Medicine, Guntur Medical College, Guntur, India. **e-mail:** iamimenotu@gmail.com

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concordance; the superior and inferior cava opened into the right atrium, pulmonary veins opened into the left atrium, with a normal biventricular function and a right-sided aortic arch. Infertility work up for the patient's wife was within normal limits. Semen analysis of the patient revealed a volume of less than 3 mL and azoospermia. Serum levels of Luteinizing hormone (LH), follicle stimulating hormone (FSH) 8 mIU/mL (normal 1-18 mIU/mL), total testosterone 6 mIU/mL (normal 2-18 mIU/mL) and prolactin 12 ng/mL (normal <20 ng/mL) were within normal limits. Testicular biopsy (Figure 2) revealed minimally dilated seminiferous tubules with maturation arrest at the stage of spermatogonia and only 5% of the tubules showed spermatogenesis. Sertoli and Leydig cells were normal in appearance and distribution. CT of the paranasal sinuses was normal (Figure 3). Ultrasonography guided aspiration of the liver abscess revealed anchovy sauce pus. A presumptive diagnosis of amoebic liver abscess was made and the patient was treated with i.v. metronidazole and oral chloroquine and he responded well to treatment.

Figure 2: CT brain and paranasal sinuses (plain) (scout film) shows no abnormality

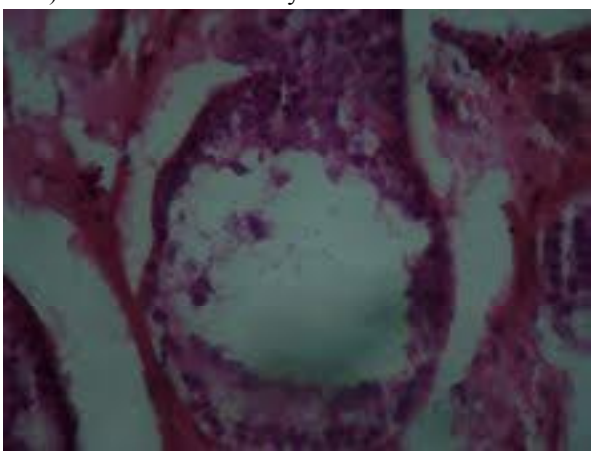

Figure 3: Photomicrography of testicular biopsy showing maturation arrest at stage of spermatogonia (Haematoxylin and eosin, × 400)

DISCUSSION

The term primary ciliary dyskinesia refers to a group of disorders caused by microtubular defects in cilia and sperm. These defects render the cilia in the respiratory tract ineffective in clearing secretions, resulting in sinusitis, repeated pulmonary infections, bronchiectasis, obstruction, and air trapping. Dysfunctional cilia in the middle ear leave patients vulnerable to repeated otitis media, and most suffer some degree of hearing loss. In disorders of primary ciliary dyskinesia, sperms, which contain an identical microtubular arrangement to cilia in their tails, have been shown to be capable of fertilizing an egg but usually lack sufficient motility to reach one. The microtubular defects found in primary ciliary dyskinesia have also been shown to cause motility problems in phagocytic cells where microtubules are involved in directed movement to capture pathogens.

There have been cases reported previously with a variety of clinical manifestations, suchs as, bronchitis, sinusitis, *situs inversus* and azoospermia,[1] immotile cilia syndrome with retinitis pigmentosa,[4] immotile cilia syndrome with azoospermia.[2]

Our patient had *situs inversus totalis* with azoospermia with primary testicular failure. The testicular failure can be explained on the basis of immotile ciliary apparatus from the testis to the epididiymis which can cause collection of the sperms in the reproductive tract. This in turn could have caused back

pressure or feed back inhibition of sperm production.[5]

It is possible that the back pressure could also have caused leak of the sperm, causing autoimmune orchitis and azoospermia.[5] However, in our case biopsy did not show any evidence of mononuclear cell infiltration typical of autoimmune azoospermia. Electron microscopic studies could not be done at our centre due to lack of infrastructure. With available clinical data it is likely that our patient manifested a variant of primary ciliary dyskinesia syndrome.

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