Case Report

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Situs Inversus Totalis with Hydrocephalus in an Adult: a Rare Case Report

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Situs inversus is a rare anomaly where there is reversal of the visceral organ location and usually associated with Kartagener syndrome and digestive, circulatory and respiratory diseases abnormalities. The lateralization of visceral organs to a side is a feature of vertebrates and humans are externally symmetrical but internally asymmetrical. Normal anatomy imparts the heart to left and the liver to the right which is found not only in humans but also other animals. This case showed aqueduct stenosis with hydrocephalus with normal corpus callosum. The association of situs inversus with ciliary abnormalities and therefore is rarely associated with hydrocephalus, polycystic disease or retinal degeneration. PubMed search for situs inversus totalis with hydrocephalus showed only 7 results with 3 of them in animal studies, one was not relevant and the rest in human. Hydrocephalus was present in only two studies, one of them cadaveric finding. The association with hydrocephalus is very rare and we present such a case report.

Key words: Acqueductal stenosis, Hydrocephalus, Kartagener syndrome, Situs inversus

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Case report

A 25-year old male patient presented to the emergency department with headache, severe pain abdomen and chest pain for 3 days. He had history of physical assault with trauma to the head, chest and abdomen by wooden object 1 day before the onset of symptoms and headache for 1 year. On examination he was conscious and orientated with a Glasgow coma Score of 15. The pupils were bilaterally equal and reactive to light and there was no motor or sensory deficit. The head or abdomen did not have any signs of external injury. His biochemistry and hematological profile were normal and there was no other significant past medical illness. In view of his chronic headache Magnetic Resonance Imaging (MRI) was done which showed communicating hydrocephalus. Ophthalmological examination showed normal vision in both eyes. Fundus examination of both eyes showed no papilledema.The chest Skiagram showed dextrocardia. For the pain abdomen and tenderness in epigastric region a Computed Tomography scan (CT) abdomen was done which did not reveal any traumatic injuries but showed total reversal of the visceral organs with the heart spleen on

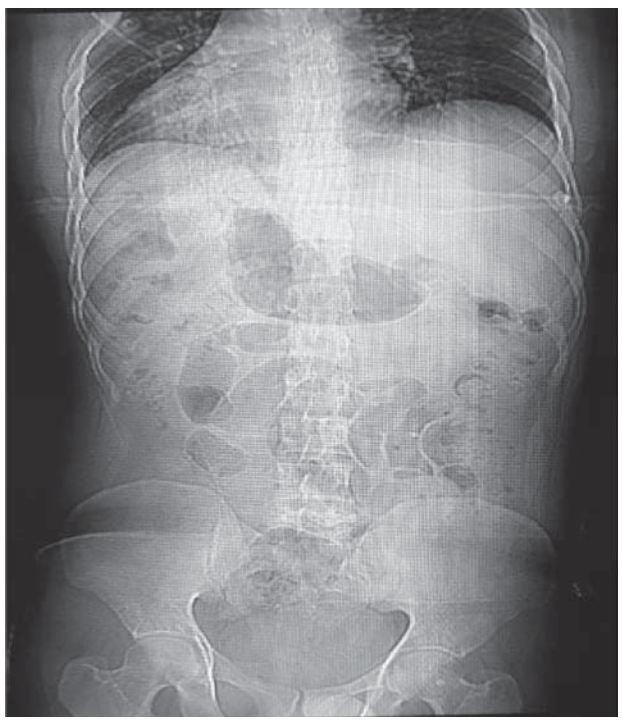

Figure 1: Chest Skiagram showing dextrocardia, stomach on right with liver on left.

right side and the liver on left side . The diagnosis of Situs inversus totalis was confirmed. Cardiac echocardiography did not reveal any congenital vascular anomalies. On further questioning it was found that he had been right hand dominance since birth and examination of his sibling did not reveal any situs inversus.He was managed with analgesics for 48 hours and discharged with advice for regular follow-up.

Discussion

The lateralization of visceral organs to a side is a feature of vertebrates and humans are externally symmetrical but internally asymmetrical.Normal anatomy imparts the heart to left and the liver to the right which is found not only in humans but also other animals.[10]Abnormal ciliary during embryogenesis can lead to an array of conditions for example, situs inversus totalis (complete reversal of visceral organs), situs ambigus (partial reversal of visceral organs).[5] Situs inversus was first referred to by Aristotle and was known as viscerum transposes, reversed asymmetry or lateral transposition and can occur sporadically or familial.[2,3]The process starts as early as late gastrulation or early neurulation where there is ciliary movement leading to lateralization of the organs.It may also manifest as just dextrocardia. Due to the abnormalities of cilia movement there can be associated digestive, circulatory and respiratory diseases which is similar to primary ciliary

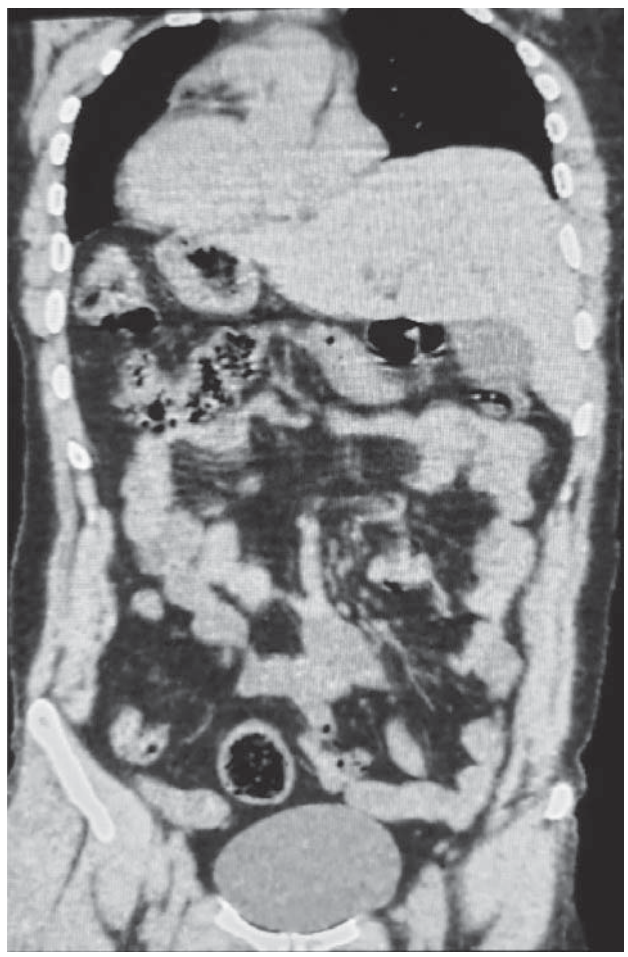

Figure 2: CT abdomen showing the spleen on the right, liver on left, stomach on right and the reversal of small and large intestine.

dyskinesia due to defects of imperfect synthetization of the motor protein Dynein and resulting in hypomotility (Kartagener syndrome). Situs inversus is uncommon with an incidence of 1 in 10000.[9,12] Most are diagnosed incidentally as the physiological function of the organs are normal. Mutation in numerous human chromosomes, ionizing radiation, retinoic acid, atracurium, hyperthermia and colchicine are some of the known factors that can cause situs inversus.[3,5,6,13]

Our case was diagnosed incidentally as he had presented with history of trauma. There was no record of him having visited any hospital and hence diagnosed of situs inversus totalis. He was right handed since childhood. Although there are many studies regarding the association of situs inversus with other congenital syndromes, diseases of the chest, asplenia, sinuses, intestine and even with myasthenia gravis, studies of intracranial anatomy and its physiological effect is limited by its paucity in literature.[1] Cerebellar hypoplasia, holoprosencephaly, larger transverse sinus and vein of Labbe was larger on

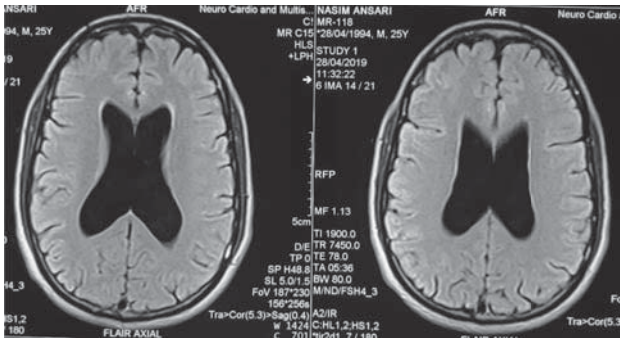

Figure 3: MRI brain with T1 axial cuts showing hydrocephalus.

right, larger internal carotid artery on right, frontal and occipital petalia reversal from their normal orientation and larger artery of Adamkiewicz on right are some of the anatomical findings in situs inversus.[13] In an study with three cases of situs inversus using structural and functional MRI and another study with 15 cases, to study the brain asymmetry it was found that the occipital (right) and frontal petalia (left) were larger in comparison to normal cases and that there was left language dominance of found as in normal cases.[4,14] This case showed aqueduct stenosis with hydrocephalus with normal corpus callosum. The association of situs inversus with ciliary abnormalities and therefore is rarely associated with hydrocephalus, polycystic disease or retinal degeneration. PubMed search for situs inversus totalis with hydrocephalus showed only 7 results with 3 of them in animal studies, one was not relevant and the rest in human. Hydrocephalus was present in only two studies, one of them cadaveric finding.[7,8,11] These studies suggest that the anatomy and lateralization of the human brain may depend on some other factors other than those controlling visceral organs.

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