

A Case with Situs Inversus, Polysplenia Syndrome and Juvenile Rheumatoid Arthritis: Evaluation of the viscerotrial situs abnormalities by magnetic resonance

SİTUS İNVERSUS, POLİSPLENİ VE JUVENİL ROMATOİD ARTRİTLİ BİR OLGU: VİSSEROATRİAL SİTUS ANORMALLİĞİNİN MR İLE DEĞERLENDİRİLMESİ

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Summary

Some kinds of viscerotrial situs abnormalities like situs inversus, situs ambiguus, isomerism, polysplenia and asplenia are rarely found in population. Juvenile rheumatoid arthritis (JRA) is the common major connective tissue disease in children. We decided to present this case, who can be accepted as a first case with viscerotrial situs abnormalities and also JRA in the literature, and give informations about MR findings of patient's viscerotrial situs abnormalities.

Key Words: Situs inversus, Polysplenia syndrome, Juvenile rheumatoid arthritis, Viscerotrial situs abnormalities, Case report

T Klin Tıp Bilimleri 1999, 19:165-168

Özet

Situs inversus, situs ambiguus, izomerizm, polispleni, ve aspleni gibi visserotrial situs anormallikleri toplumda seyrek görülür. Juvenil romatoid artrit (JRA) çocukluk çağında sık görülen konnektif doku hastalığıdır. Belki literatürde hem JRA'li ve hem de visserotrial situs anormalliği bulunan ilk olguyu sunmayı ve aynı zamanda hastanın visserotrial situs anormalliğinin MR bulguları hakkında bilgi vermeyi amaçladık.

Anahtar Kelimeler: Situs inversus, Polispleni sendromu, Juvenil romatoid artrit, Visserotrial situs anormalliği, Olgu sunumu

T Klin J Med Sci 1999, 19: 165-168

Some kinds of viscerotrial situs abnormalities like situs inversus, situs ambiguus, isomerism, polysplenia and asplenia are disorders which are rarely seen in the population (1). On the other hand, juvenile rheumatoid arthritis (JRA) is the most common major connective tissue disease in children and affects frequently the large joints like knees, wrists and ankles (2,3).

Situs solitus is defined as a normal position of the atria and viscera; or it can be defined as a normal differentiation and placement of the assymet-

ric body parts like lungs, atria and viscera (1,4,5). Situs inversus indicates the mirror image location. Situs ambiguus is an abnormal type of situs which the relationship of the atria and viscera is inconsistent; and occasionally is termed partial situs inversus (1,5). In this type of situs abnormality, asymmetric structures can be symmetric (1,4). Isomerism is a term used to describe symmetric morphology. Asplenia is a syndrome characterized by situs ambiguus with bilateral right sidedness (right isomerism). Polysplenia syndrome is a second situs ambiguus characterized by bilateral left sidedness (left isomerism) (1).

JRA has three major types of presentation which involves oligoarthritis, polyarthritis and systemic disease (2,3). Renal failure due to amyloido-

Geliş Tarihi: 04.11.1998

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sis secondary to JRA can be developed, then death can occur in the patients (2).

We decided to report this case which could be accepted as the first case with JRA in addition to situs inversus and polysplenia syndrome together in literature. Likewise, we generally meet case reports or studies about sonographic and computed tomography (CT) evaluations of situs inversus and polysplenia in literature. That's why, we have also approved the presentation of magnetic resonance (MR) findings of situs inversus and polysplenia syndrome.

Case Report

A 22-year old male with musculoskeletal complaints admitted to Erciyes University, Faculty of Medicine, Department of Physical Medicine and Rehabilitation 6 months ago. In 1986, he had been diagnosed as acute rheumatic fever and rheumatic carditis and treated with corticosteroid and penicilline. In 1990, Sulfasalazine treatment had been applied to the patient because of swelling in one of his knee. Because of ineffectivity, this treatment had been stopped three months later. When we accepted the patient to our service, he had been complaining of articular and muscular pain and inability

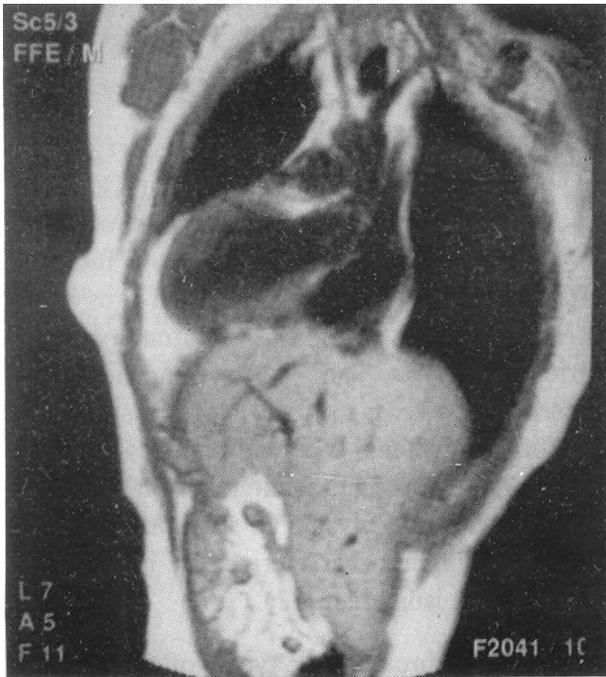


Figure 1. T1-weighted coronal MR image; heart in the right side and liver in the left side (situs inversus totalis).

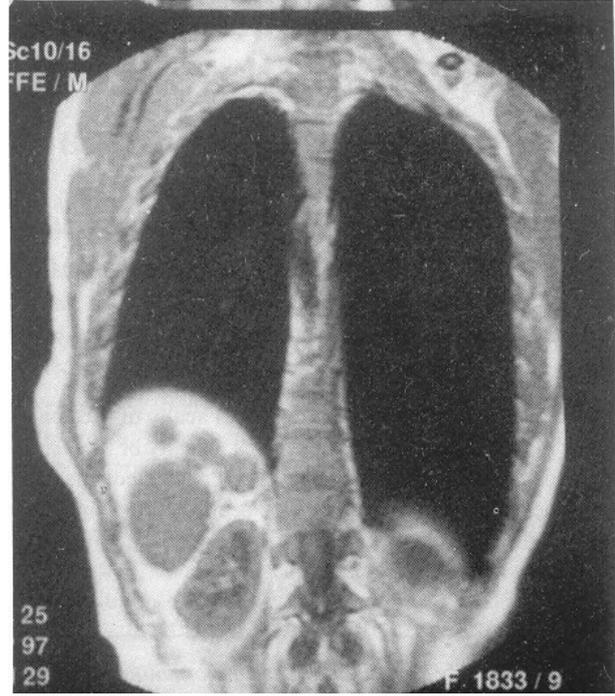


Figure 2. T1-weighted posteriorly coronal MR image; polysplenia in the the right side.

ty of walking and standing for 6 months. In physical examination, there were swelling, heat and tenderness in his ankles and knees, contractures in hip and knee joints. In abnormal laboratory findings, erythrocyte sedimentation rate as 100 mm/h, hemoglobin as 10.6 g/dl, rheumatoid factor as 184 IU/ml, C-reactive protein as 97, proteinuria as +++, serum Fe level as 14 mg were detected. The patient was diagnosed as JRA according to American College of Rheumatology diagnostic criteria for the classification of juvenil rheumatoid arthritis (6). Rectal biopsy was applied due to chronic diarrhea and proteinuria and found amyloidosis secondary to JRA. The patient were evaluated by a cardiologist as well. Dextrocardia was detected in echocardiography. In addition, we have learned that aortic murmur was found in his physical examination and aortic insufficiency and dextrocardia were detected in echocardiographic evaluation in 1992. That's why, the patient's dextrocardia was evaluated by abdominal sonography, CT and MR. In these examination, situs inversus and polysplenia were detected in the patient. The MR imaging of the visceratrial situs abnormalities are shown on Figures 1-3.

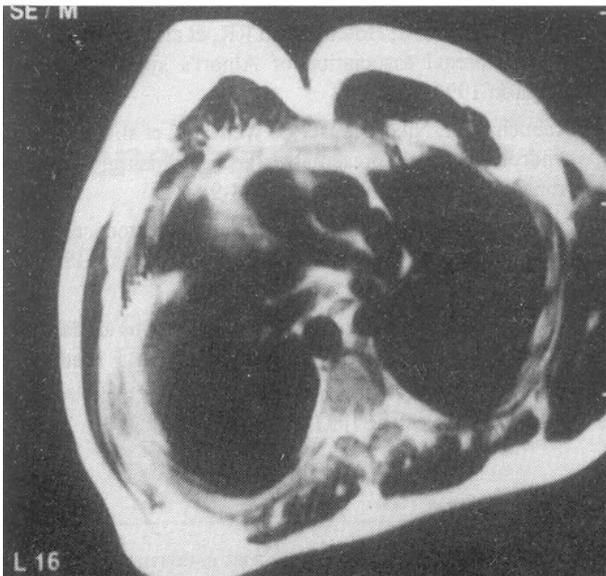


Figure 3. T1-weighted axial MR image: pulmonary artery and descending aorta in the right side, ascending aorta in the left side and hemiazygos in the left side of descending aorta.

Discussion

Although polysplenia syndrome and situs inversus are congenital abnormalities, it may be first seen in adulthood, especially when congenital heart disease is not present (1).

The cases with viscerotrial situs abnormalities and also other disorders like Alport's syndrome, Kartagener's syndrome, group A streptococcal infection, and others were reported in literature (7-9). But we did not meet the cases with viscerotrial situs abnormalities and also JRA together in literature. Probably this is the first case report of the case with situs inversus, polysplenia syndrome and also JRA together.

The concept of situs or laterality of body organ is an important one in understanding congenital visceral and vascular malformations. Situs inversus is diagnosed when body asymmetry is entirely the mirror image of situs solitus. The diagnosis of polysplenia syndrome is made by identifying two or more splenules (5).

Because each patients will have a unique constellation of vascular and visceral derangements, patients suspected of having viscerotrial situs abnormalities should be evaluated individually to determine abdominal visceral and vascular arrangement and to investigate associated problems (5).

This investigation should begin with plain film to see cardiac and gastric positions and pulmonary vascularity. But plain films are known to be often inaccurate in identifying real viscerotrial situs abnormalities (10). In a study, it was seen that plain films were misleading in 30% of cases (5). Bronchial anatomy is a much more reliable predictor of true viscerotrial situs abnormalities (10). Assessment of the bronchial branching patterns should be done in all patients. Then the other methods like sonography, CT and MR can be applied (5).

Splenic tissue originates in the dorsal mesentery of the stomach, that's why, splenuli are located in the retrogastric region. In order to determine the splenic tissue, the retrogastric region scanning is necessary (5). Identification of vena cava and its location with respect to the aorta and midline can easily be made noninvasively. The findings of these two vessels on the same side of the spine has been reported for diagnosis of asplenia (1).

Sonography will delineate the presence or absence of splenic tissue and the anatomic status of the cava and the portal vein. When no spleen is found and there is conjestive view of pulmonary vascularity, we should suspect and verify a subdiaphragmatic total anomalous pulmonary venous connection. If sonography is unable to answer the pertinent questions, it would be needed of MR which has high cost and require sedation (5).

As a result, we concluded that each patient with situs abnormalities should be evaluated not only by plain graphies and sonography, but also by MR, if needed. In addition, when dextrocardia was suspected or detected accidentally, the patients should be investigated about the other situs abnormalities as well.

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