A Diffuse Idiopathic Skeletal Hyperostosis (DISH) Case with Systemic Involvements

SİSTEMİK TUTULUMLU DİFFÜZ İDİYOPATİK İSKELETSEL HİPEROSTOSİS OLGUSU

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Summary

We present a case of diffuse idiopathic skeletal hyperostosis (DISH), in which the spine and peripheral joints were profoundly involved, most notable at the cervical spine. Besides musculoskeletal involvement there were calcifications in his wrist arteries. So we discussed the metabolic factors and systemic involvement in DISH.

In spine radyograms, the calcification was seen in the anterior longitudinal ligament along the cervical spine and was better seen in the inferior parts of dorsal spine. Posterior longitudinal ligament calcification was seen at the level of cervical 5-6 and ligamentum nuchea and also atherosclerotic calcifications was detected in wrist.

In these patients the risk of aterosklerozis might be related to the tendency of ossification in DISH. The calcification of nuchael ligament may be significant in the prognosis of DISH.

Key Words: Diffuse idiopathic skeletal hyperostosis, Vascular involvement

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Özet

Servikal omurgada daha belirgin olmak üzere, omurga ve periferik eklemleri ileri derecede tutulan, diffüz idiyopatik iskeletsel hiperostosis (DİSH) olgusu ve bu olguda kas-iskelet sistemindeki tutulumlar dışında, el bileği arterlerinde saptadığımız kalsifikasyonlar nedeniyle sistemik tutulumlu DİSH ve DİSH'de metabolik etkenler tartışıldı.

Omurga radyografilerinde, servikal omurgada boylu boyunca, dorsalde alt dorsal bölgede daha belirgin olan anterior longitüdinal ligamentte kalsifikasyon izleniyordu. Servikalde nükheal ligamentte ve servikal 5-6 arasında posterior longitüdinal ligamentte de kalsifikasyon mevcuttu. El bileğinde aterosklerotik kalsifikasyonlar mevcuttu.

Bu kişilerde artmış ateroskleroz riski, DİSH'teki kemikleşmeye olan yatkınlıkla direkt sorumlu olabilir. Nükheal ligamentte kalsifikasyonun varlığı DİSH'in prognozunda belirleyici değer taşıyabilir.

Anahtar Kelimeler: Diffüz idiyopatik iskeletsel hiperostosis, Damarsal tutulum

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Diffuse idiopathic skeletal hyperostosis (DISH) is seen especially in middle old aged men with an unknown etiology (1). According to autopsy specimens, the incidence of axial and extra-axial skeleton DISH is reported 6-12% (2). It is characterized with the increase in the amount of normal bone, heterotopic bone formation, and more important new bone growth into the entheseal regions where tendon, ligament, joint capsule or annulus fibrosus fibers insert into bone. Criteria for diagnosis of DISH are the need for the new bone formation to bridge four contiguous vertebral bodies in the absence of degenerative disc disease and the absence of inflammatory sacroiliac or facet changes (3).

In this case, not only high-grade involvement was found in musculo-skeletal system but there were calcifications in his wrist arteries. So, we discussed DISH and the metabolic factors and systemic involvement in DISH.

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Case Report

While 65 years old man was hospitilizing in endocrinology clinic for type II diabetes mellitus

(DM), he was examined for his left shoulder pain by physical therapy and rehabilitation clinic. According to his cervical radiographs and physical examination DISH was thought. After regulation of the glucose level of the blood, he was accepted to physical therapy and rehabilitation clinic.

Medical history: 6 months ago, the patient fell down on his left shoulder and after this, he suffers from pain and restriction of motion which increased day by day. He didn't apply to practitioner for shoulder pain. As far as today the patient did not suffer any complaint of his musculo-skeletal system and he didn't have any inflammatory rheumatic disease, either.

He had DM type II for 24 years. Until 2 months he was taking oral antidiabetics and he was taking insulin. He had hypertension for 6 years. He had a coronary by- pass operation in 1993.

Family history: His brothers had hypertension and DM.

Musculoskeletal System Examination: Height: 1.62 cm, weight 63 kg, body mass index: 24 kg/m2, arterial blood pressure was under control with antihypertensive pills.

Passive and active range of motion (ROM) in cervical spine was evaluated. Restriction of flexion was %50, restriction of extension and rotation was %75. Palpations of cervical spinous process and neck muscles were painful. The distance between mandible-sternum was 6 cm., the distance between occiput-back wall was 5 cm, and the distance between tragus-acromion was 12 cm on the right and 14 cm. on the left. There was an increase in thoracic kyphosis.

ROM in shoulders was as,

	Right shoulder	Left shoulder
Active/ Passive flexion	120/130	75/90
Active/ Passive extension	60/80	40/45
Active/ Passive abduction	80/95	65/80
Active/ Passive adduction	30/40	20/20
Active/ Passive internal rotation	90/90	90/90
Active/ Passive external rotation	45/55	20/30

ROM in shoulders was painful. Yergason test in the left shoulder was positive, in the right negative.

ROM in the elbows was as follows,

	Right elbow	Left elbow
Active/ Passive extension	15/20	5/15
Active/ Passive flexion	130/140	120/155

ROM in the wrists was as follows,

	Right wrist	Left wrist
Active/ Passive extension	55/55	40/50
Active/ Passive flexion	55/65	70/75

ROM was painless in the wrists and elbows. Prayer's sign were bilateral positive in hands.

Expansion of thorax was measured 1 cm. Lordosis of lumbar spine was decreased. ROM in the lumbar region were restricted approximately 50% to every direction. The palpation of lumbar muscles was painless. The examination of sacroiliac joints was normal. FABER tests were bilateral positive. ROM in the hips was as follows.

	Right hip	Left hip
Active/ Passive flexion	60/70	70/75
Active/ Passive extension	0/10	5/5
Active/ Passive external rotation	7/10	5/5
Active/ Passive internal rotation	10/10	3/3

There was no pain and restriction at the other peripheral joints. Schober's test was 1 cm. at the dorsal, 3 cm. at the lumbar, the finger floor distance was 32 cm. In the neurologic examination, deep tendon reflexes for the lower extremities were hypoactive and superficial sensation got lost as sock. Babinsky was bilateral indifferent. Neurologic examination in the upper extremities was normal. Pulses around the wrists were weak. When he puts down his leg, edema and cyanosis occur. There was no other circulation problem except this.

His laboratory findings were as in the following: ESR: 35 mm/h, Hb: 11 g/dl, Hct: 32.9%, WBC: 4.9 mm3, PLT: 278.000 mm3, fasting blood glucose: 153 mg/dl, insulin: 55 mIU/ml other routine biochemical studies were in normal ranges. Urinalysis: Not any significant pathology. ASO: (-), CRP: 1.87, RF (latex titer): (-).

Radiographic Findings:

Cervical spine radiographs: The configuration of vertebral bodies and the intervertebral discs and the intervertebral foramens remains normal. Calcification all over anterior longitudinal ligament

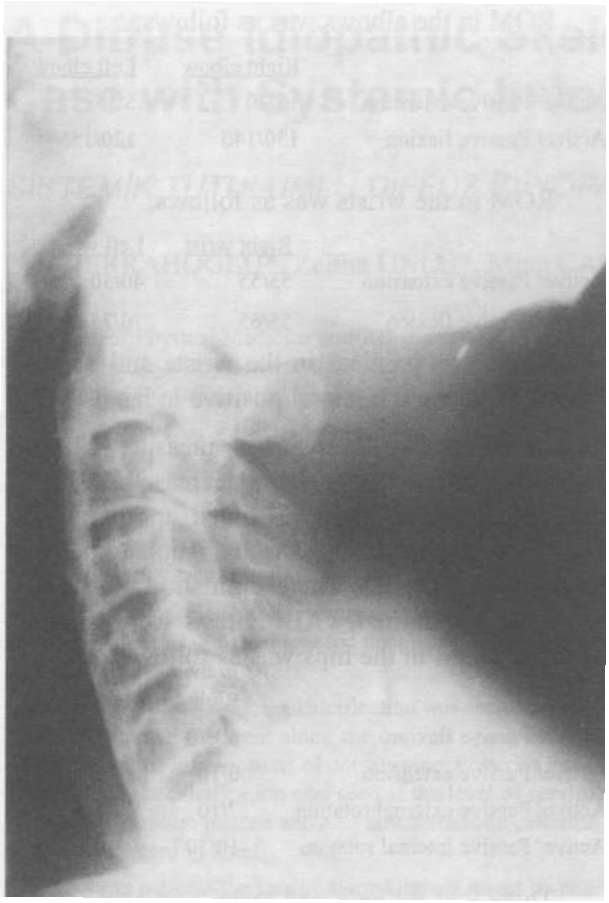

Figure 1. Calcification in the nucheal ligament was shown.

(ALL) was found. Posterior longitudinal ligament (PLL) was partially calcified in between the fifth and sixth cervical vertebrae. Calcification was seen in the nucheal ligament (Figure 1).

Thoracic spine radiographs: Thoracic kyphosis was increased. The radiolucent appearance of bones were favor of the osteoporosis. The calcification of the ALL begins at the level of the eight thoracic vertebra and particularly seen in the lower thoracic spine. There was disc calcification in between the tenth and eleventh thoracic vertebrae. Paravertebral ligaments were calcified especially in the lower thoracic spine.

Lumbar spine radiographs: Calcification was rather less than seen in the thoracic region. There were ligament calcifications between the first to third lumbar vertebrae. Vertebral peduncles, apophyseal joints and intervertebral disc spaces were normal.

Figure 2. Atherosclerosis calcifications can be seen along the right ulnar artery trace.

Sacroiliac joints radiographs: Sacroiliac joints were normal. There was calcification in iliolumbar and coxofemoral ligaments.

Left shoulder radiographs: Articular space was normal. There was paraarticular soft tissue calcification at superior of glenoid fossa. This calcification was accepted pericapsular or peritendonal calcification. The articular space was narrow in the left acromioclavicular joint. Right shoulder radiographs was normal.

Hand radiographs: At the right wrist, there were atherosclerosis calcifications all along the right ulnar artery trace. There were minimal degenerative changes in the distal interphalangeal joints. There was calcified hypertrophied changes in the interosseous tendons and attachment of collateral ligaments to phalanges (Figure 2).

In the knee, elbow, ankle radiographs: there was absolute calcified hypertrophied changes at the attachment of the tendons.

Computerized tomography (CT) of sacroiliac joints: subchondral faces of joints were normal and joint spaces were clear. There was calcification and bridging at the ligaments of sacroiliac joints.

Cervical spine where the most prominent involvement detected was evaluated by magnetic resonance imaging (MRI). A MRI of the cervical spine revealed that the third through seventh cervical intervertebral discs were degenerated. There was no compression to make myelopathy. CT scan of the atlanto axial joints was found normal.

Doppler Ultrasonography: diffuse atheroma plaques in the extracranial segments of bilateral carotis arteries were found. Approximately 40% stenosis was seen in common carotis artery and internal carotid artery. According to doppler examination of bilateral lower extremities, calcifications and degeneration all over femoral artery especially of the left leg was found. These changes were seen at the level of popliteal and distal parts too. And lymphedema findings were bilaterally positive too.

In cranial CT: Atrophic changes were found.

Thorax CT: There was no pathology about vena cava superior syndrome. Cardiac and lung changes of atherosclerosis and hypertension were seen.

Electroneuromyography examination showed diabetic polyneuropathy in the lower extremities.

Otorhinolaryngology (ORL) clinic consultation was normal. According to neurology clinic consultation findings were thought to be diabetic polyneuropathy.

All according to these findings, the patient's diagnosis was discussed as DISH.

Electrotherapy and kinesiotherapy was managed for 3 weeks. The patient's complaints diminished after these physical therapies and restriction of joint movements passed away.

Discussion

DISH is a diffuse systemic condition and local factors may modify its expression. In DISH cases, 17%-60% of them has glucose tolerance impaired. The prevalence of DISH in patients with type II DM ranges from 13-50% and no patients have been found for type I DM. In type I DM, spine is not attacked, as it is in type II DM. Except type I diabetics almost all diabetics are hyperinsulinemic. Higher than normal values are required for glucose control because of the endogenous insulin resistance at the cells. New bone deposition in DISH may be related to the insulin's growth factor like activity (3). Patients with acromegaly have new bone deposition like DISH by growth hormone. Obesity, hypertension, adult onset diabetes and associated hyperinsulinemia in-groups have high prevalence of DISH support this hypothesis (3). As it is in our case DM and obesity are separate factors in appearance of DISH. All these information might tell us the effect of DM in DISH.

At DISH, in entheseal areas which might be inactive ossification centres are involved and its etiology is unknown. Miedany et al (4) revealed that hypervascularity of the ossified ligaments and vertebra involved, in addition to a significant increase in the size of the affected vertebrae, pointing to the possible role of a vascular disorder in the DISH pathogenesis. So increased cortical thickness of tubular bones of the hand and increase in size of the sesamoid bones are reported in patients with DISH as in our case (3). In our case, new bone formation in the entheseal areas were most prominent and besides severe ossification arterial walls were interesting. As known, lipid and glucose metabolism abnormalities may cause hypertension and DM have an increased risk of atherosclerosis (3). Atherosclerosis was responsible of this case's bypass operation. In addition, DISH may effect ossification of arterial walls directly. Increased ossification diathesis due to metabolic abnormalities in DISH may effect on the arterial involvements. Thus, in this case, the arterial calcifications may be associated with the ossification diathesis in DISH. Similarly, new heterotopic bone formations were found in DISH cases with spine attacked after the operations such as hip arthroplasty (3).

At first description of DISH, it was told that spinal ossification did not originate from ALL; it was probably originated from perivertebral fibrous sheath (5). Later, Resnick et al. (2) found out that the early findings of DISH were ALL calcifications in thoracic spine with chondroid metaplasia and endochondral ossification. According to them, new bone formation in cervical region arises in the particularly ligamentum nuchae. In our case, calcification in the nucheal ligament was observed and this might be important for the severity of involvements. Ossification of the PLL is considered to be part of the generalized hyperostotic tendency. The current results confirmed the parallel association between ALL and PLL ossification (6).

In DISH, spinal and extraspinal involvements are usually asymptomatic. Although stiffness at the

spine is the significant complaint conversely, the ROM is saved interestingly (7). However in our case, spinal and extraspinal joint involvement of DISH was severely restricted and painful.

We detected os nuchae and ALL calcifications in cervical radiographs associated with DM, so we made the diagnosis DISH. In conclusion, if the calcification in nuchal ligament in radiologic evaluation was detected in a diabetic patient with extraspinal complaints DISH might be considered. In these patients the risk of atherosclerosis might be directly related to the tendency of ossification in DISH. The calcification of nucheal ligament may be an important factor with respect to the severity and prognosis of DISH. The systemic and extraspinal involvement in DISH has not been studied at all. Further clinic studies are needed to investigate the relationship of nuchal ligament calcification and atherosclerosis in DISH cases.

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