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A RARE PRESENTATION OF POLYOSTOTIC FIBROUS DYSPLASIA IN CERVICAL, THORACIC AND LUMBAR SPINE

SERVİKAL, TORAKAL VE LOMBER OMURGAYI BİRLİKTE TUTAN NADİR BİR POLİOSTOTİK FİBRÖZ DİSPLAZİ OLGUSU

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SUMMARY:

A 30 years old male applied to our institution with the complaint of back pain due to polyostotic fibrous dysplasia (PFD) involving the posterior elements of C5 vertebra and the bodies of T5, T10, T12, L1 and L5 vertebrae, left iliac bone and right proximal femur. He had a previous history of open nonspecific T12 biopsy and three level posterior spinal instrumentation. Re-biopsy was performed and PFD diagnosis was confirmed. In the same session with open biopsy, spinal instruments were extracted due to local pain caused by their superficial position under the skin. Following the procedure, pain relieved. The patient remains asymptomatic at 26 months postoperatively

There are few studies demonstrating PFD in the spinal column. Cervical, thoracic and lumbar spine, pelvis and proximal femur involvement altogether is rare. This is the first report of polyostotic fibrous dysplasia involving cervical, thoracic and lumbar spine together with pelvis and proximal femur.

Key words: Fibrous dysplasia, polyostotic variant, cervical, thoracic, lumbar, spine.

Level of evidence: Case report, Level IV.

ÖZET:

Otuz yaşında bir erkek hasta hastanemize C5 vertebranın arka elemanlarını ve T5, T10, T12, L1 ve L5 vertebraların cisimlerini, sol iliak kemik ve sağ proksimal femuru tutan polyostotik fibröz displaziye (PFD) bağlı sırt ve bel ağrısıyla hastanemize başvurdu. Hastanın anamnezinde daha önce dış merkezde yapılmış olan T12'den açık nonspesifik bir biyopsi ve bu seviyenin bir üst ve altı dahil olmak üzere 3 seviye spinal enstrümentasyon hikayesi mevcuttu. Hastaya cerrahi müdahalede bulunuldu. Açık re-biyopsi yapıldı ve aynı seansta tüm spinal implantlar ciltaltında yüzeyel yerleşim sebebiyle ağrıya sebep olduklarından dolayı çıkartıldı. İşlem sonrası hastanın ağrıları azaldı. Yirmi altı aylık takip sonunda son kontrolde hasta ağrısızdı.

Literatürde omurgada PFD'ye ilişkin az sayıda çalışma bulunmaktadır. Servikal, torasik ve lomber omurgayla birlikte aynı zamanda iliak kemik ve proksimal femur tutulumu olan bu olgu literatürde bildirilen ilk vakadır

Anahtar kelimeler: Fibröz displazi, polyostotik varyant, torasik, lomber, omurga.

Kanıt düzeyi: Olgu sunumu, Düzey IV.

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INTRODUCTION:

Fibrous dysplasia (FD) is a benign bone lesion characterized with irregularly shaped, fibroblastic tissue that contains metaplastic woven bone tissue[8,9]. Disease is subcategorized on the basis of number of bone involvement. Monostotic fibrous dysplasia (MFD) is the one bone involved form and seen more frequently, whereas polyostotic fibrous dysplasia (PFD) is the multiple bone involved form. FD occurs commonly in the extremities. Axial skeleton is rarely affected. PFD commonly effects the skull bones[13,27]. Both MFD and PFD limited to the spine have been reported in the literature[1-2,14,16-18,20,22,25-26,30-32]. The spine is reported to be involved in 1,4 % to 5,5 % of fibrous dysplasia cases[3,12,23]. Vertebral bone involvement is rarely (~4%) seen in PFD[17]. Spinal cases with PFD are generally asymptomatic and diagnosed co-incidentally[10,26].

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CASE REPORT:

A 30 year old male presented to the clinic with left groin and back pain of 1 year duration. He had a history of a nonspecific T12 biopsy performed in another institution 8 years before. Following biopsy, T11-L1 short segment posterior spinal instrumentation was performed for fusion in the same institution.

On physical examination, a thoracolumbar 6 cm long previous operation scar was noted. The pain flared up with bending, flexion and extension motion of the trunk and palpation on the left iliac crest and the incision site. There was no neurological deficit.

Direct radiographies followed by magnetic resonance imaging (MRI) of the whole spinal column and pelvis were performed. A mild sagittal imbalance was observed on lateral whole spinal column radiography. Osseous lesions involving the posterior elements of C5 vertebra, the bodies of T5, T10, T12, L1 and L5 vertebrae were detected on MRI. MRI also revealed non-destructing lesions in the iliac bone and femur cortices. A bone scan demonstrated increased uptake in the body of L5 vertebra, bilateral sacroiliac joints and the right iliac crest but not in the other lesions revealed by MRI (Figure-1).

All laboratory test results were within normal ranges.

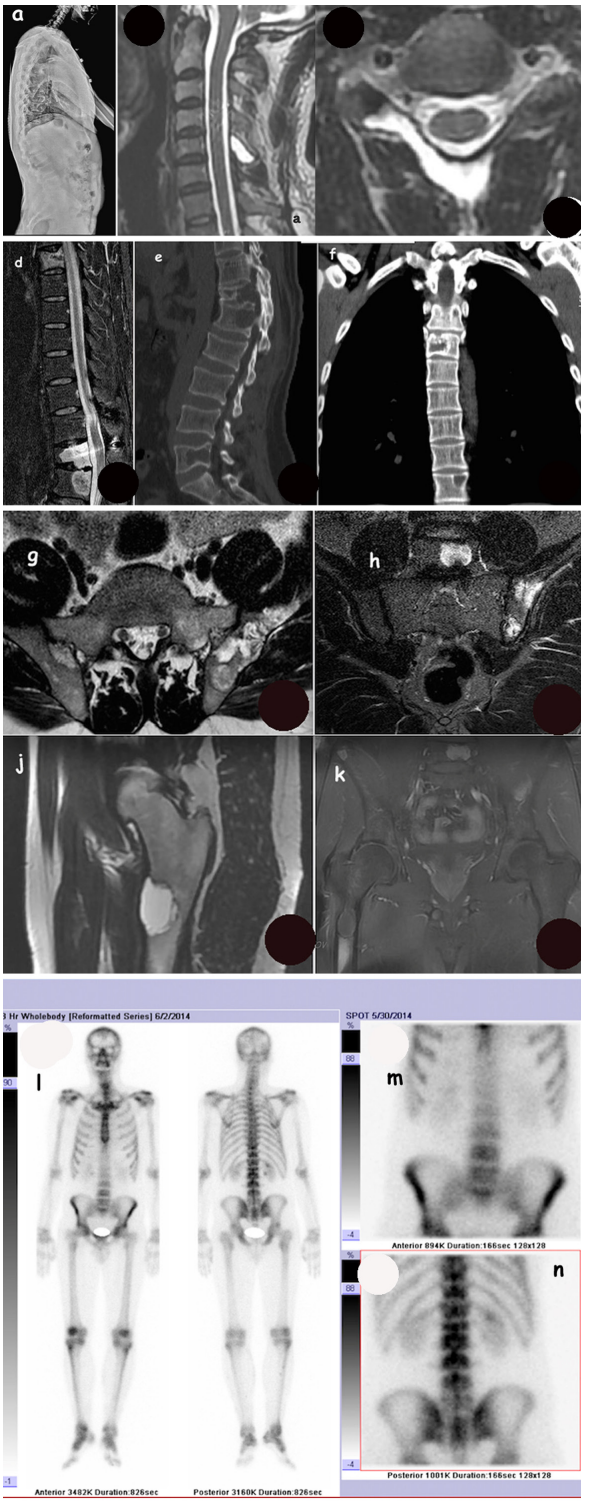

Figure 1. a) Preoperative lateral direct radiograph of the patient. Note the mild sagittal imbalance. **b-c)** Sagittal and axial MRI revealing the lesion in C5 vertebra. **d-e-f)** Sagittal MRI and CT reveal the lesions in T5 and T10, **g-h-j-k)** T12 and L1 and L5 vertebrae. Pelvic lesions demonstrated in axial and coronal MRI and right proximal femoral lesions demonstrated in sagittal and coronal MRI. **l-m-n)** The bone scan performed in the patient.

Open re-biopsy was performed from the bodies of T12 and L1 vertebrae and previous posterior instrumentation was removed in the same session because of the superficial position of the implants under the skin, leading local pain. Posterior fusion was observed during surgery and as the spinal column was found stable a new instrumentation was not performed.

The histopathological evaluation confirmed PFD with multiple bone involvement after positive pathology with cellular fibrous matrix and poorly oriented bony trabeculae.

Following the surgery, although there was mild sagittal imbalance, back pain relieved and groin pain was controlled with medications. No post operative complication was observed. The patient remains asymptomatic at 26 months postoperatively (Figure-2).

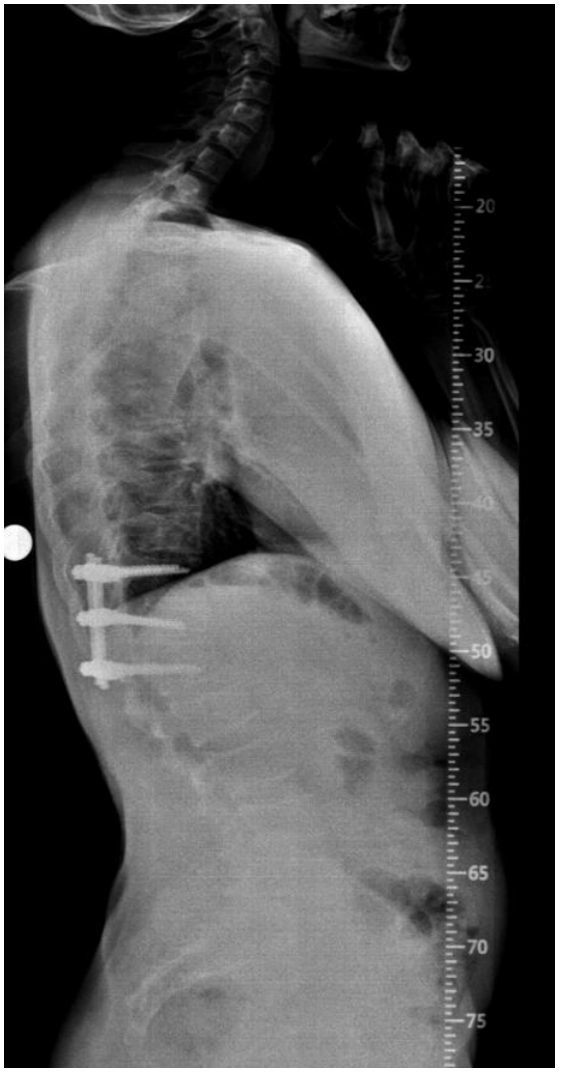

Figure-2. Lateral radiograph obtained at the end of the 26th postoperative month.

DISCUSSION:

Fibrous dysplasia is a benign neoplastic lesion characterized with the replacement of medullary normal trabecular bone with immature fibro-osseous tissue. Generally the lesions progress slowly. Genetical mutation in the GNAS1 gene which encodes alpha subunit of stimulatory G protein is reported[24]. The disease has two forms: monostotic and polyostotic form. MFD is more common than polyostotic form[25]. PFD is associated with McCune-Albright syndrome in which, cutaneous and endocrine manifestations are accompanied with fibrous dysplasia[4].

FD is generally an asymptomatic disease. It is usually diagnosed co-incidentally on radiographies[22]. In few symptomatic patients, pain, swelling and deformity are the main manifestations. In a small percentage of patients pathologic fractures may be seen[2,12,19]. Malignant formation occurs with a incidence of 0.5 % in MFD but in McCune Albright syndrome this ratio rises up to 4 %[12]. In our case, back and groin pain were the main symptoms.

Radiographic findings of fibrous dysplasia include a matrix "ground glass" apperance of marginal sclerosis near the lytic zones[7]. Only radiographic findings are not sufficient for a definite diagnosis of fibrous dysplasia. A needle biopsy is used to rule out hemangioma, giant cell tumor and aneurysmal bone cyst diagnosis. In the elderly, multiple myeloma and metastatic carcinoma should also be considered[7].

Computerized tomography (CT) scan can be the best imaging modality to demonstrate the radiographic characteristics of FD[13]. A CT scan confirms decreased cancellous bone and cortical thinning, ballooning, or collapse of vertebrae[13]. In our case CT scan revealed lithic lesions in the cervical, thoracic and lumbar segments both in the posterior elements and the vertebral bodies (Figure-3).

MRI is useful as it reveals the extent of tumor involvement, extent of neural compression, and contributes for the differential diagnosis. FD appears as a homogeneous hypo-intense lesion on T1-weighted imaging unless there is a pathologic fracture. On T2-weighted imaging, the lesion is heterogeneous depending on the amount of fibro-osseous tissue, cellularity, cystic alterations, hemorrhage, and thus, FD appears hypo-intense on both T1 and T2-weighted imaging, and uniformly enhances after intravenous gadolinium. MRI revealed similar findings also in our patient. A bone scan may also help for diagnosing PFD, as in our case[29].

Due to the rarity of PFD in spinal column, the optimal treatment options are unclear. Treatment options may include nonoperative and operative treatments. Nonoperative treatment include calcitonin, biphosphonates, mitramycin and pamidronate[28].

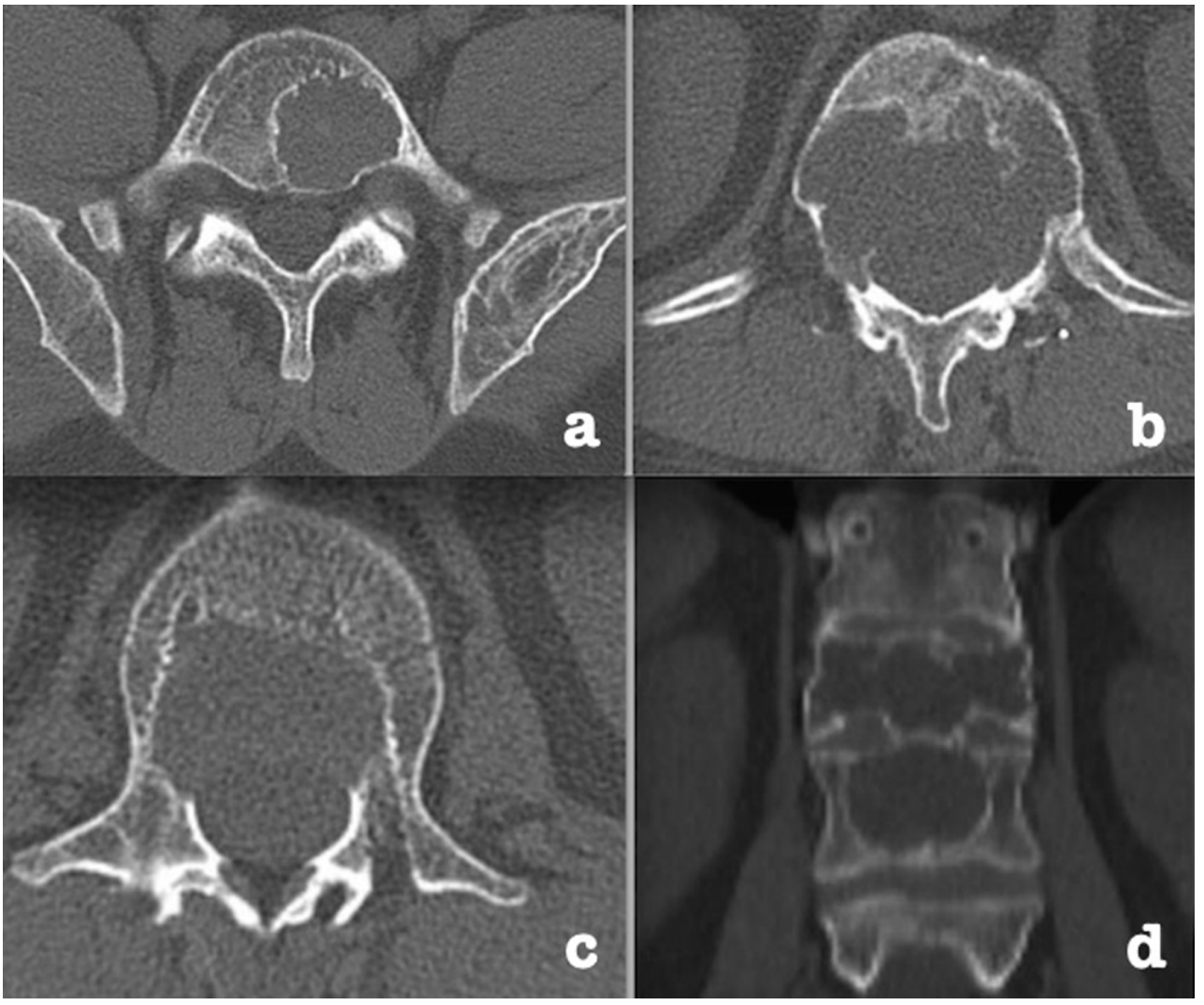

Figure-3. Axial and coronal CT images revealing **a)** the lesions in L5, **b-c-d)** T12 - and L1 vertebrae.

Curettage, internal fixation and bone grafting are the operative treatment modalities[28]. Agressive resection and rigid fixation with fusion are suggested by some authors[18,21,30]. However FD becomes silent or rarely regress after the end of the bone growth. Thus, this approach is controversial[1,22,28]. In the more common case of FD involving the non-axial skeleton, many cases have been managed conservatively, even following pathological fracture[10-11].

Indeed, some have also advocated conservative management for vertebral FD[28,31], in particular with PFD, where such lesions are frequently asymptomatic. For example, Smith et al. conservatively managed a patient with multilevel cervical spine PFD for over 30 years, during which time several vertebrae had spontaneously fused[26].

Surgery, however, is usually indicated in cases with fracture[6,17], neurological deficit[32], progressive deformity[14,16] or persistent pain[5].

The cases with cervical, thoracal and lumbar involvement are reported in polyostotic fibrous dysplasia before[14,16-17]. In this case, polyostotic fibrous dysplasia involves theposterior elements of C5 vertebra, the bodies of T5, T10, T12, L1 and L5 vertebrae, left iliac bone and the right proximal femur. The main complaint of the patient was back and left groin pain. After removal of previous posterior spinal instrumentation, back pain mostly relieved and left groin pain was controlled with medications.

Fibrous dysplasia of the spine is a rare condition. Multiple spinal segments can be involved. Polyostotic fibrous dysplasia may be a part of systemic disease. This patient had multisegment spinal, pelvic and right femur involvement. After implant removal, patient's pain relieved and did not need any radical surgery. At the 26 months follow up, patient did not have any complaints about pain or sagittal imbalance.

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