LUMBAR SPINAL CANAL STENOSIS IN OSTEOPOIKILOSIS (CASE REPORT)

Cüneyt ŞAR* Azmi HAMZAOĞLU* Nadir ŞENER*
Önder KILIÇOGLU* Mişel KOKINO*

Osteopoikilosis is a rare form of bone displasia which is usually an incidental radiological finding. The disease has typical radiological findings and generally does not require treatment; however, in the recent years, co-existing pathologies have been reported. One of these pathologies is lumbar spinal stenosis, requiring surgical intervention.

In this article, an osteopoikilosis case with lumbar spinal stenosis is presented and diagnostic and therapeutic approaches are discussed.

Key Words: *Osteopoikiloses, canal stenosis, lumbar spine.*

Osteopoikilosis is an osteoschlerotic bony displasia which is an incidental finding on routine radiographs. It may be noted at birth or may appear during skeletaly growth and affects both sexes equally. Described first by Stieda in 1905, radiological findings consisting of bilateral, periarticular, well-defined, uniform in density, multiple round, oval or lanceolate dense spots of 2-3 mm diameter, are pathognomonic. Usually they tend to cluster around the epiphyseal and metaphyseal regions of long bones and around joints, in the carpus and tarsus, ant in the pelvis. The lesions do not affect the cortex or the contour of the bone. The skull, face, ribs, sternum and vertebrae are rarely involved (1, 5, 6, 8, 10, 21, 23).

Reports concerning its incidence are controversial. Some report an incidence of 1/10.000.000, while Jonasch diagnosed 12 cases after screening 211.000 people (13, 15, 21). Despite the fact that only 160 cases have been reported in the literature, in 1992, Benli et al. have reported 49 additional cases following documentation of four family trees (4). Thus, since it is usually asymptomatic and the diagnosis is incidental, the prevalance might be much higher than expected. It is inherited as an otosomal dominant trait.

Lately, many developmental disorders and pathological conditions have been associated with osteopoikilosis. These associated findings may concern all three germ ayers. Aortic coarctation, ureteral duplication and endocrine dysfunctions are related to the endoderm, exostoses, osteitis condensans ilii, melorheostosis, lumbar spinal stenosis, dwarfism, hyperostosis frontalis interna to the mesoderm and cleft lip and palate, dental anomalies, predisposition to keloid formation, dermatofibrosis, lenticularis disseminata to the ectoderm (10, 15, 21, 23). Dermatofibrosis lenticularis disseminata accompanies osteopoikilosis in 10% of cases and together they are referred to as the Buschke-Ollendorff syndrome (4, 21, 23). Also low back pain and various articular involvement have been reported (23). Biochemical evaluations and bone scintigraphy do not display pathology.

Etiology and pathogenesis are still obscure. The current theory emphasizes a genetic defect in normal development of trabeculae on lines of stress and consequent formation of bony islands of various dimensions, that is, an inappropriate respons to stress stimulus. Histology of these dense nodules reveales laminated bone merging with the surrounding spongiosa. The proportion of haversian spaces is the same as that seen in dense cortical bone. Osteopoikilotic nodules behave the same way like normal growing bone tissue. Within years, they may diminish in size or totally disappear. Malingnant transformation to osteosarcoma and chondrosarcoma have been reported in two cases with osteopoikilosis (11, 17).

The lesions must be differentiated from osteoblastic metastases, mastocytosis, tuberous sclerosis, osteomesopyknosis and multiple myeloma.

Osteopoikilosis generally does not require therapy. However, any accompanying symptomatic disorder should be treated. Lumbar spinal stenosis is one of these conditions and the only case in the literature has been reported by Weisz in 1982 (23).

This article reports lumbar spinal stenosis in a patient with osteopoikilosis and reviews the diagnosis and therapy of this condition.

CASE REPORT

The 32 year-old patient (height 170 cm, weight 70

* University of İstanbul, İstanbul Medical Faculty, Department of Orthopaedics and Traumatology, Çapa, İstanbul-Turkey

kg) suffering from low back pain for two years was referred to our clinic after the incidental finding of pelvic sclerotic lesions on a radiograph taken during an intravenous pyleogram. His complaint was pain in the lumbar region and both extremities, which increased during walking and standing and affected his daily activities.

Physical examination revealed normal range of motion in both hips and knee joints without any pain related to motion. Straight leg raising test was negative and neurological deficits like loss of reflexes, hypoesthesia, motor weakness and incontinance. However, his pain in the lumbar region and thighs increased when walking and standing, but diminished when resting. These symptoms were evaluated as neurogenic claudication. No associated skin lesions were present.

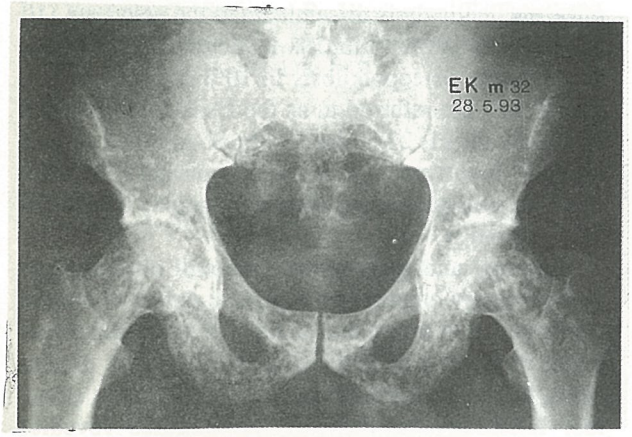

Figure 1a.

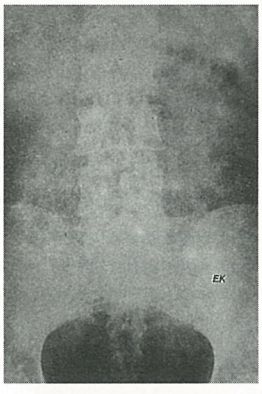

Figure 1b.

Figure 1. Radiographs of the pelvis, and vertebra of the patient.

Radiolojical evaluation revealed dense round clusters typical for osteopoikilosis in the pelvis, distal and proximal ends of both femora, tibiae and humeri, tarsus and carpus, lumbar vertebrae and the ankles and wrists (figure 1). No lesions were seen in the cranium, ribs, claviculae, dorsal and cervical vertebrae and the diaphyses of long bones. Joint spaces in both hips and knee joints were normal.

Biochemical analyses of the serum and urine samples did not display any pathology.

CT scanning of the thoracic, lumbar and lumbosacral regions exhibited severe stenosis of the spinal canal in both midsagittal and transvers diameters and enlargement of the pedicles, involving segments L1 through L5. In all CT sections the midsagittal diameter of the spinal canal was 10 mm and had a triangular configuration (figure 2).

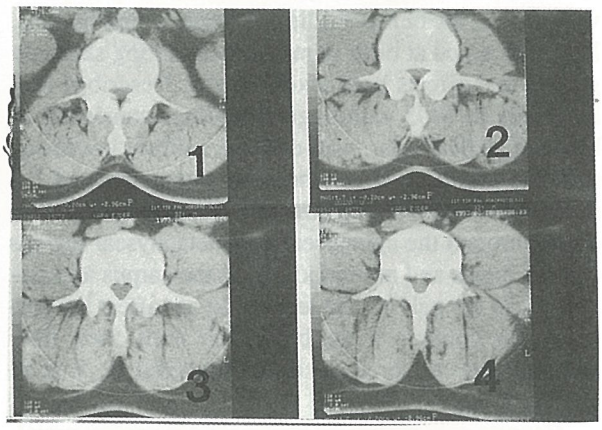

Figure 2. Lumbar CT section.

Similar lesions could not be demonstrated in none of his first degree relatives.

Decompressive laminectomy and foraminotomy involving 5 segments between L1 and L5 since his complaints interfered with his daily life. Compact-Cotrel Dubousset instrumentation and posterolateral fusion was required due to spinal instability (figure 3).

In the most recent follow-up examination in the 15th month postoperatively, all his symptoms had disappeared and complete fusion was achieved.

DISCUSSION

Diagnosis of osteopoikilosis is usually incidental since the external configuration of the bone remains intact and so the disorder is asymptomatic despite the existing radiological and histological pathologies. Moreover, the importance of the diagnosis decreases,

because the condition does not require any treatment. However, recent discovery of various accompanying pathologies and clinical symptoms emphasize the fact that further investigations concerning this seemingly benign disorder are necessary.

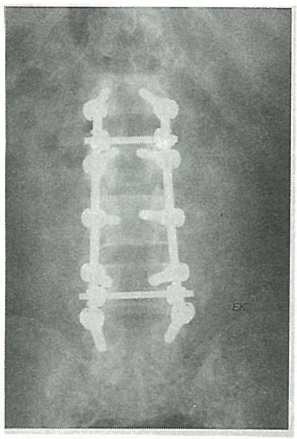

Figure 3a.

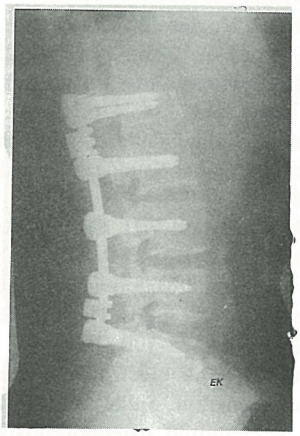

Figure 3b.

Figure 3. Postoperative radiographs of the patient.

Weisz, in his report of the first osteopoikilosis case with spinal stenosis, published in 1982, called the non-characteristic symptoms and signs also seen in the early stages of Paget's disease, as premyelopathic syndrome (23). In contrast to congenital and acquired form of spinal stenosis, he described a clinical picture with lumbar and lower limb pain unrelated to position and without neurologic deficits (23, 24).

In 1975, Arnoldi et al. proceeded to classify lumbar spinal stenosis based on the ways in which lumbar nerve roots can become compressed (2). Their scheme has subsequently been modified in classifying spinal stenoses as congenital, developmental and acquired (7, 16). Congenital spinal stenosis, which occurs as a result of a congenital malformation, constitutes only a small subset of patients with spinal stenosis. Sarpyener was one of the first to recognize and describe this entity in the thoracolumbar spine (19). Developmental stenosis refers to genetic disturbance of both fetal and postnatal development that usually continues until maturity. Developmental stenosis may be idiopathic or part of a generalized growth disturbance, which is most frequently found in achondroplasia (16, 22). Acquired spinal stenosis implies a process that occurs following birth that is caused by degenerative changes, trauma or bone dysplasia or is iatrogenic in nature (16, 22). Our case can be classified as developmental stenosis.

Verbiest, defined "absolute", "relative" and "mixed" stenosis through measurement of the midsagittal diameter (22). Absolute stenosis was defined as a midsagittal diameter of 10 mm or less. Midsagittal diameters between 10-13 mm were termed relative stenosis. Mixed stenosis represented patients with midsagittal diameters less than or equal to 10 mm in some locations and between 10 and 13 mm in others. In our case, the measurements revealed absolute stenosis.

Low back pain is a symptom frequently accountered in spinal stenosis, and it is mechanical type of pain developing gradually, increasing with activity and subsiding with rest. Pain typically distributes to the coccygeal and gluteal regions and increases spesifially in extension. Another spesific symptom is pain in the lower limb called neurogenic claudication or pseudoclaudication and is seen in 72% of spinal stenoses (16). Complaints like pain in both limbs starting from the lumbar region until the knee joints, burning, weakness and **cramps** appear typically during extension movements like standing, walking or climbing stairs, but diminishes on sitting or bending forward. Motor weakness, loss of reflexes and sensorial deficits are generally not a part of this group of symptoms. Straight leg raising test is negative. Our patient carried this typical symptomatolgy observed in spinal stenosis. Neurogenic claudication affected his normal daily activities.

Today, imaging techniques utilized in the diagnosis of spinal stenosis include mylography, CT scanning, computed myelography and MRI scanning. Plain radiography is not useful in identifying lumbar development stenosis. Myelography is one of the most effec-

tive means of detecting spinal stenosis. In a comparative study using GT, myelography proved to be sensitivite in 93% of cases while CT was sensitive in 89% (3). Myelography, however, is an invasive method and requires the use of contrast material, increasing the morbidity. CT has great advantages over myelography in that first, it is an objective indicator of the true shape and size of the spinal canal; second, it makes possible direct imaging of the lateral recesses and neural foraminae which can only be indirectly identified through myelography; third, imaging of pathologies below the involved segments producing blockage of contrast material are also possible; and last, it is noninvazive method. A combination of these two imaging techniques has been shown to increase sensitivity (3, 9). MRI has also been demonstrated to be as effective as CT and myelography. Modic et al. found comparable findings with MRI and surgery in 82.6% of cases, between CT and surgery in 83% and between myelography and surgery in 71.8% (18). When MRI and CT are combined, then this rate increases to 92.5%, and when myelography and CT together the rate is 89.4%. We have evaluated the degree of spinal stenosis with CT scanning and documented matching surgical findings.

EMG shows bilateral multiradicular findings in the late stages of spinal stenosis (12). False-negative results may be obtained since muscle groups may have multiple innervations and it should be taken into account that EMG usually measures motor nerve impulses and cannot evaluate sensorial disfunctions. However, in spinal stenosis, sensorial deficits are a major part of the clinical symptomatology. Therefore, EMG usually gives normal findings in spinal stenosis.

Indications for surgery in patients with spinal stenosis include pain not responding to medical therapy, neurogenic claudication and neurologic deficits. Objectives of treatment is to decompress all neural elements under pressure. This can be carried out by decompressive laminectomy and preservation of the facet articulations and also by adding foraminotomy in cases where stenosis of neural foraminae exist. Instability created by factors like the disruption of faset articulations, inclusion of the disk spaces during decompression and the high number of decompressed segments, necessitate fusion and instrumentation (9, 14). Multisegmental decompression and foraminotomy followed by stabilization with CCD and posterolateral fusion has been performed in our case.

Our case report has aimed to emphasize the fact that spinal stenosis can accompany osteopoikilosis and where indicated, decompression with instrumentation and fusion should be performed.

REFERENCES

1. Archer MC: Osteopoikilosis: Report of to cases. Radiology. 47, 279-283, 1946.
2. Arnoldi CC, Brodsky AE, Cauchoix J, et al.: Lumbar spinal stenosis and nerve root entrapment syndromes. Clin. Orthop. 115: 4-5, 1976.
3. Bell GR et al: A study of computer assisted tomography: comparison of metrizamid myelography and computed tomography in the diagnosis of herniated lumbar disc and spinal stenosis. Spine 9: 552-556, 1984.
4. Benli İT, Akalın S, Boysan E, Mumcu EF, Kı[a] M, Türkoğlu D: Epidemiological, clinical and radiological aspects of osteopoikilosis. J. Bone Joint Surg. 74 B: 504-506, 1992.
5. Berlin R: Osteopoikilosis; A clinical and genetic study. Acta Med. Scant. 181; 3, 305-314, 1967.
6. Bloom AR: Osteopoecilia. Am. J. Surg. 22; 239-243, 1993.
7. Dorwart RH, Vogler JB, Helms CA: Spinal Stenosis. Radiol. Clin. North Am. 21: 301-325, 1983.
8. Editorial: Osteopoikilosis. Familial documentation. Am. J. Dis. Child. 134, 416, 1980.
9. Garfin et al: Spinal stenosis. In Rothman RH, Simeone FA: The Spine. Saunders, Philadelphia, pp: 791-857, 1992.
10. Green AE, Ellsvood WH, Collins JR: Melorheostosis and osteopoikilosis. A.J.R. 87, 1096-1111, 1962.
11. Grimer RJ: Chondrosarcome chez un patient porteur d'osteopoikilie. A propos d'un cas. Revue de Chirurgie Orthop. 75: 188-190, 1989.
12. Haldeman S: The electrodiagnostic evaluation of nerve root function. Spine 12: 282-286, 1984.
13. Hinson A: Familial osteopoikilosis. Am. J. Surg. 45, 566-573, 1939.
14. Johnson KE, Willner S, Johnson K: Postoperative instability after decompression for lum bar spinal stenosis. Spine 11: 107, 1986.
15. Jonascs E: Faelle von Osteopoikilie. Fortschr. a.d. Geb. d. Rπntgenstrahlen, 82, 344-353, 1955.
16. Kabins MB: Congenital and Developmental Spinal Stenosis. In Weinstein SL. The Pediatric Spine. Principles and Practice. New York. Raven Press, pp. 275-305, 1994.
17. Mindell ER, Northup CS, Douglass H: Osteosarcoma associated with osteopoikilosis. J. Bone Joint Surg. 60A: 406-408, 1978.
18. Modic MT et al.: Lumbar herniated disc disease and canal stenosis: prospective evaluation by surface coil MR, CT and myelography. AJ.R. 147: 757-765,

1986.
19. Sarpyener MA: Congenital Stricture of the Spinal Canal. J. Bone Joint Surg. 27: 70-79, 1945.
20. Szabo AD: Osteopoikilosis in a Twin. Clin Orthop. 79: 156, 1971.
21. Tachdjian MO: Pediatric Orthopaedics. Saunders, Vol II pp: 815-816, 1990.
22. Verbiest H: Fallacies of the present definition, nomenclature and classification of the stenoses of the lumbar vertebral canal. Spine 1: 217-225, 1976.
23. Weisz GM: Lumbar Spinal Canal Stenosis in Osteopoikilosis. Clin Orthop. 166: 89-92, 1982.
24. Weisz GM: Lumbar canal stenosis in Paget's disease. Spine. 8: 192-198, 1983.