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Case Report

Osteopathia striata with cranial sclerosis and lumbar spinal stenosis

F. E. Deniz[1], **R. D. Köseoğlu**[2]

[1] Department of Neurosurgery, Gaziosmanpaşa University Faculty of Medicine, Tokat, Turkey
[2] Department of Pathology, Gaziosmanpaşa University Faculty of Medicine, Tokat, Turkey

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Summary

Osteopathia striata with cranial sclerosis is one of the dysplasias of endochondral bone formation. We report a patient with this condition combined with lumbar spinal stenosis and intervertebral disc herniation. This coexistence has not been previously reported in the medical literature. The possible mechanisms of this clinical association is discussed.

Keywords: Osteopathia striata; cranial sclerosis; lumbar spinal stenosis; disc herniation.

Introduction

Osteopathia striata with cranial sclerosis (OS-CS) is a sclerosing bone dysplasia, which is subgrouped within dysplasias of endochondral bone formation [15]. OS-CS is inherited as a dominant trait but it remains unclear whether it is an autosomal dominant or X-linked trait [9]. It is generally a benign condition and thus remains mainly a disorder of radiological and academic interest; some affected individuals have facial dysmorphism and symptoms of cranial nerve encroachment. The typical and name-giving roentgenographic finding of OS-CS is the fine, uniform, linear striation of the tubular bones. Fan-like striations of the iliac bones are present in more than 50% of patients [5]. OS-CS can occur in isolation, or may be part of a syndrome together with heart defects, malrotation of the abdominal organs, omphalocele, partial agenesis of the corpus callosum and Pierre-Robin sequence [14]. About one hundred patients with OS-CS have been reported previously.

Lumbar spinal stenosis (LSS) is defined as narrowing of the lumbar spinal canal, nerve root canal or intervertebral foramina. The lower limit of normal for the antero-posterior diameter of the lumbar spinal canal is 15 mm and less than 11 mm is considered as severe LS. The narrowing can be measured from antero-posterior diameters in axial images of computed tomography (CT) or magnetic resonance (MR) scans. However, the cross-sectional area of the dural sac is a more reliable diagnostic measurement [2].

It is important to distinguish between spinal stenosis found incidentally and symptomatic spinal stenosis. Severe stenosis may remain asymptomatic as a result of the adaptability potential of the thecal sac. The symptoms of LSS are back pain, leg pain, weakness and neurogenic claudication. These symptoms may be acutely aggravated by disc herniations.

According to the original aetiological classification, LSS can be divided into two subgroups with either congenital or acquired causation. LSS occurring in a congenitally small canal may present symptoms as early as the third decade of life. The operative treatment for uncomplicated LSS is decompressive laminectomy with or without lateral recess and foraminal decompression with preservation of facet joints and stability.

Correspondence: Fatih Ersay Deniz, Department of Neurosurgery, Gaziosmanpaşa Üniversitesi Hastanesi, Nöroşirürji ABD, 60100 Tokat, Turkey. e-mail: ersaymd@yahoo.com

Case report

A 33-year-old man had a four year history of pain in his lower extremities on walking. Over a four-day period, the pain in the left leg increased with weakness of dorsiflexion of the left foot. Neurological examination revealed weakness in the left tibialis anterior and extensor hallucis longus of grade 3/5, loss of sensation to light touch in the left leg and diminished left knee and bilateral ankle reflexes. Radiography demonstrated increased density of the cranial base, thickened calvarium, striation of femur, tibia, metacarpal and metatarsal bones, fan-like striation of the pelvis and sclerosing of the vertebrae suggesting OS-CS as the diagnosis (Figs. 1, 2). There were no complaints relating to the bones or joints. Laboratory test results were normal



Fig. 1. A-P radiograph of the pelvis demonstrating fan-like striations of iliac bones and typical linear fine striations of proximal femur



Fig. 2. Lateral radiograph of the skull demonstrating basal sclerosis and thickened calvarium

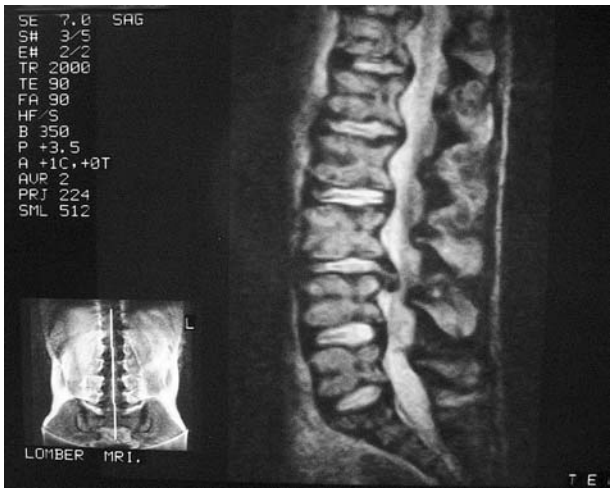


Fig. 3. Sagittal T2-weighted MR image demonstrating severe dural sac compression of the L3–4 level by extruded disc herniation and moderate canal stenosis with posterior compression at multiple levels

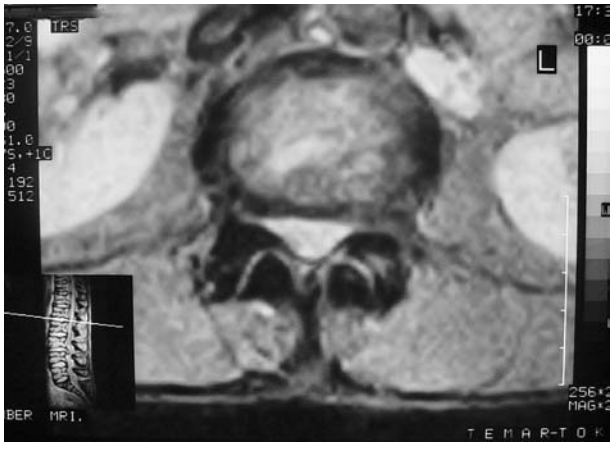


Fig. 4. Axial T2-weighted MR image at the level of L1–2 demonstrating moderate stenosis with A-P diameter of 13 mm

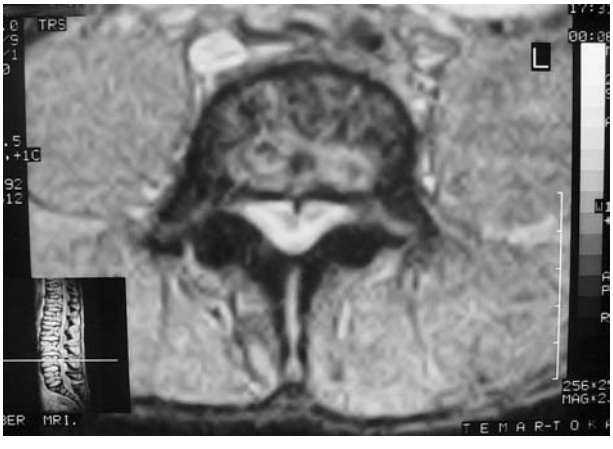


Fig. 5. Axial T2-weighted MR image at the level of L4 demonstrating moderate stenosis with A-P diameter of 14 mm

except serum alkaline phosphatase of 160 U/L (normal range 50–136) and total creatine kinase of 666 U/L (normal range 21–232).

MR imaging of the lumbar spine demonstrated a moderately stenotic canal at both vertebral and intervertebral levels except at the level of L5 where the canal size was 18 mm. The dural tube was compressed at the L3–4 intervertebral level by an extruded disc herniation (Figs. 3–5).

He underwent laminectomy of L3, discectomy of L3–4 and foraminatomies of bilateral L4 nerve roots. Facet-sparing laminectomy was performed in order to prevent the patient from developing symptomatic stenosis that may be aggravated after surgery and by ageing. After the operation, he made positive progress towards recovery and follow-up evaluation two years after the operation indicated resolution of the pain and weakness (Figs. 7, 8).

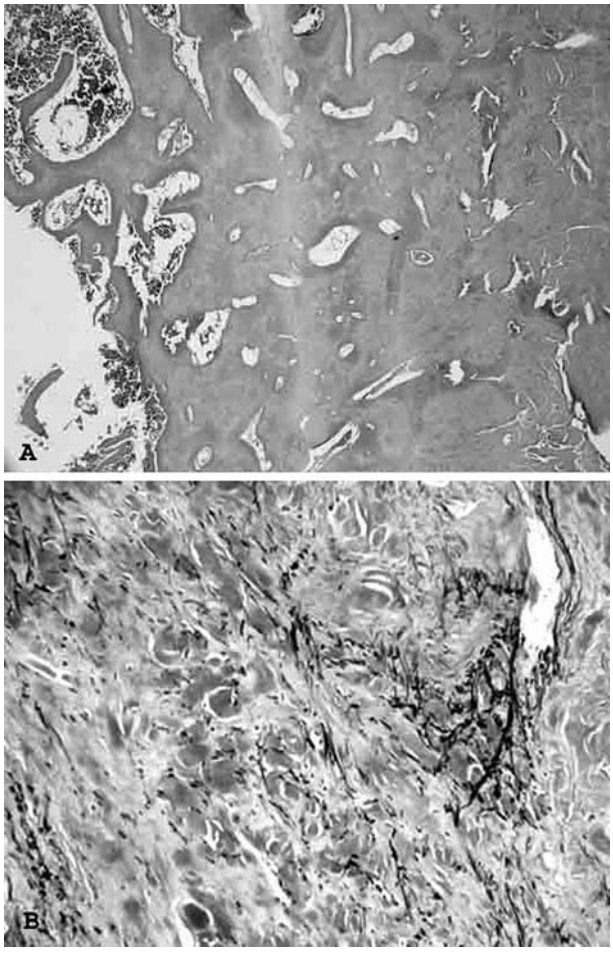


Fig. 6. (A) Biopsy of vertebra demonstrating increased trabecular thickness (H&E; × 10). (B) Area of ligamentum flavum demonstrating very few and thin elastic fibers scattered in a collagenous matrix (Elastic stain; × 20)



Fig. 7. A-P radiograph of the lumbar vertebrae 2 years after surgery



Fig. 8. Lateral radiogram of the lumbar vertebrae 2 years after surgery

The tissues removed at operation including bone and fragments of ligamentum flavum were sent for routine histopathological analysis. The prepared sections were stained with hematoxylin-eosin and with elastic stain in addition for the sections of ligamentum flavum. Increased trabecular thickness was noted in the sections of laminectomy material (Fig. 6A). The sections of ligamentum flavum showed coarse collagen bundles and focal degeneration areas of matrix. The decreased elastic fibers in the ligamentum flavum was revealed with the elastic stain (Fig. 6B). Essentially, the coarse pattern

of the collagen bundles was accompanied by decreased elastic fibers. In our patient, chondroid metaplasia and calcification were not observed.

Discussion

As OS-CS is usually an asymptomatic condition, it may not be discovered throughout life and only about one hundred patients have been reported in the literature. To the best of our knowledge, OS-CS with neurological symptoms due to LSS complicated by disc herniation has not been reported in the medical literature.

OS-CS is classified as a dysplasia of endochondral bone formation. The bones of the extremities and those parts of the axial skeleton that bear weight, like the vertebrae, develop by endochondral ossification. However, this does not mean that it persists as endochondral bone because by a remodeling process, new bone replaces the preexisting bone by appositional growth. As bones elongate, remodeling occurs with resorption of bone in some areas and the deposition of bone in other areas [12]. The thickening of the cortex is due to periosteal and endosteal bone apposition and a dysequilibrium between periosteal bone formation and endosteal bone resorption may result in an increase in bone thickness. In our patient, increased trabecular thickness was detected and we speculate that LSS was also due to a dysequilibrium between periosteal bone formation and endosteal bone resorption.

There is a lack of data concerning the extensive operative methods and their relationship to instability of the lumbar spine. Often, anecdotal data and dogmatic recommendations render the subject even more controversial [13]. The generally accepted concept is removal of the disc via an interlaminar fenestration and if a wider decompression is required to combine this with spondylodesis. In younger patients treated with massive extirpation of the disc material, there is an increased likelihood of intervertebral instability [7]. Instability following decompressive lumbar laminectomy is rare (1%), and stability is thought to be maintained if more than 50–66% of the facets are preserved and the disc space is not violated [3]. Rao *et al*. investigated nine fresh calf spines and concluded that laminectomy causes more destabilization of a spinal motion segment than laminotomy and significantly increases disc stress in the anterior annulus [10].

Lumbar facet joint anatomy can be preserved if the appropriate trajectory is used for lateral recess decompression. The surgical trajectory of a laminectomy provides lateral recess access and decompression without significant facet joint and spine destabilization. A laminotomy involves a more vertical approach but access to the lateral recesses is obstructed by the spinous processes with a greater degree of facet joint resection being required and this may needlessly destabilize the spine [1]. Schaller reported 2353 discectomy procedures of which nine percent were performed via hemilaminectomy and 12 patients developed single-level segmental instability. Of these twelve patients, 8% had interlaminar fenestration while 92% had laminectomy; 58% had a monosegmental approach while 42% had bisegmental resection; finally 67% had monosegmental discectomy while 33% had bisegmental discectomy. He concluded that the pars interarticularis is an important structure for lumbar spine stability and should be preserved intraoperatively, whenever possible [13]. Lee reported that the incidence of newly developed post decompression olisthesis after extensive decompression was 3.7% [6]. Herron reported on forty-six patients who had previously undergone laminectomy for lumbar disc herniation and treated for recurrent disc herniation by repeat laminectomy and discectomy and concluded that fusion is not routinely required in such patients [4]. In one study, Retwiler *et al*. compared differences in biomechanical stability between Christmas tree laminectomy (CTL) in which bilateral facetectomies and foraminotomies were performed and facet-sparing laminectomy (FSL) in which the facets are undercut but not resected. The sixteen motion segments obtained from five human cadaver lumbar specimens were examined for spinal instability immediately post-decompression and again after post-discectomy. The CTL specimens had significantly larger increases in motion, than the FSL treated counterparts compared with the intact condition, and subsequent discectomy caused greater increases in motion in the CTL group. The authors concluded that the treatment of LSS with FSL induces less biomechanical instability and these findings support the use of FSL in treating LSS [11].

LSS can be asymptomatic due to the adaptability potential of the thecal sac and symptoms may be acutely aggravated by disc herniations. It is important to correlate the physical findings with the imaging examinations in LSS. Imaging studies of the lumbar spine are not performed in asymptomatic patients and this is probably the reason for our patient being the first report of the association of OS-CS with LSS. Perhaps OS-CS should be added to the list of the possible aetiologies of spinal stenosis but more reports are mandatory to resolve this issue.

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Comment

This is a case presentation of a patient with a rare congenital bony abnormality (ostopathia striata) plus a common spinal problem (spinal stenosis). Symptomatic spinal stenosis is often the result of degenerative changes superimposed on congenital narrowing of the spine. The narrowing is congenital in the since that at skeletal maturity the patient has a small spinal canal and limited reserve space. As the spine ages naturally, the canal is further narrowed through bulging discs, thickened intraspinal ligaments and bony hypertrophy. Individuals with the same degree of degenerative change but the good fortune of a wide canal at skeletal maturity remain asymptomatic. This case is interesting because a congenital condition, ostopathia striata may have generated an acquired stenosis after the patient reached skeletal maturity. It also appears that the patient has had superimposed degenerative narrowing. The question remains if the degenerative changes are the result of normal ageing (spondylosis) or accelerated as a result of the ostopathia striata.

H. Louis Harkey and *Jackson*