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Research Paper

Ankylosing Spondylitis- An Osteological Study of Human Axial Skeleton.

Medical Science

KEYWORDS : Ankylosing spondylitis, Ossification of spinal ligaments, Sacroiliac joint, Ligamentum flavum, Pudendal nerve entrapment syndrome.

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ABSTRACT

Human Axial Skeleton has drawn much curiosity among researchers because of the evolution of erect posture. The vertebral column plays an imperative role in stability and weight transmission. At the same time the vertebral column is the site for many orthopedic disorders like Ossification of spinal ligaments, Ankylosing spondylitis, kyphosis, and scoliosis. The sacroiliac joint, the largest axial joint, has also gained its essence in various studies in relation to the revolutionary treating aspects in various degenerative diseases of the joint.

The present case describes a rare human axial skeleton, showing ossification of various spinal ligaments, sacrospinous, sacrotuberous, transverse acetabular ligaments and ankylosis of sacroiliac joint. These ossified structures may hamper the movements of the vertebral column, pelvic bone and may compress the neurovascular bundle passing in between these structures resulting in various painful conditions.

Anatomical and radiological knowledge of ossification of the ligaments and joints of axial skeleton as found in this case may be helpful for clinicians, radiologists and orthopedic surgeons for differential diagnosis and can be implicated in the development of pioneering treatments of back and perineal pains.

Introduction

Ossification of spinal ligaments (OSL) is a pathological entity that causes ectopic bone formation and consequently results in various degrees of neurological deficit. The etiology of OSL is still poorly understood[1]. It has been associated with various diseases like Ankylosing spondylitis (AS), diffuse idiopathic skeletal hyperostosis (DISH), fluorosis, Paget's disease, adenocarcinomatous metastasis, hypophosphatemic vitamin D resistant rickets and disorders of calcium metabolism. It was found to occur almost solely in Japanese population; but the disease is now being gradually more recognized in the Indian population [2,3,4,5]. It is characterized by ossification of various spinal ligaments, such as the anterior (ALL) and posterior longitudinal ligament (PLL), ligamentum flavum (LF), interspinous (ISL) and supraspinous ligaments (SSL). This leads to decreased spinal mobility and functional impairment of these patients.

Sacro-iliac joint (SIJ) is the largest axial joint designed primarily for stability, and its main function is to transmit the truncal weight to the lower extremities. Stability of the joint is maintained by the intrinsic and extrinsic ligaments associated with it[6]. Ankylosis or fusion of this joint and ossification of the adjacent ligaments can decrease the mobility of the joint. When muscles and ligaments act on the joint, they can be the cause of pain and inflammation if these joints are in dysfunction. It has also been associated with AS and chronic perineal pain syndromes[7,8].

Generally, these bony pathological features are reported in various radiological investigations but an actual skeletal experience would facilitate the orthopaedicians who performs corrective surgeries on these bones. Here, we are reporting the details of one unusual axial skeleton, with ossification of most of its associated ligaments which may be a severe case of AS.

Case report

We are reporting the details of an unusual axial skeleton comprising of vertebral column with articulated pelvis procured from our bone bank in the Department of Anatomy, Maulana Azad Medical College, New Delhi, India (Fig-1,2). The vertebral column was completely kyphotic. It had completely ossified ALL in the region of T2 to T6, SSL in the region of T3 to sacrum, ISL in the region of T5 to sacrum and LF from T2 to the sacrum. Partial ossification of ALL in the region of L1-L2, L3-L4 and L4-L5 was also seen. No ossification of PLL was noticed.

The bony pelvis showed bilateral ankylosis of sacroiliac joint (SIJ). The antero-inferior synovial part was partially fused whereas the postero-superior interosseous part was completely fused. The anterior and posterior sacroiliac ligaments of both the sacroiliac joints were ossified. The posterior superior iliac spine showed numerous osteophytic extensions of ossified ligaments attached to it. The sacrospinous ligament (SCSL) of both sides was completely ossified, extending between the ischial spine and the fourth piece of sacrum. The sacrotuberous ligaments (STL) on both sides were partially ossified and it was not fused with sacrum. Complete ossification was also seen of iliolumbar ligament (ILL) of both sides and pubic symphysis (PS). Partial ossification of both sides iliotibial tract (upper part) was also noticed.

The transverse acetabular ligament (TAL) on right side was completely ossified, which transformed the acetabular notch into a foramen for the passage of neurovascular bundle to the hip joint. On the left side, there was partially ossified TAL.

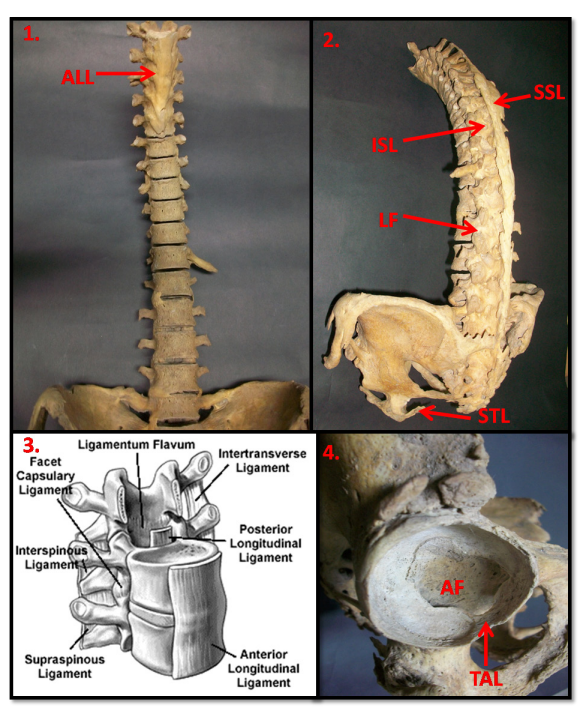

Fig-1: (1) Anterior view of axial skeleton, (2) Oblique lateral view of axial skeleton, (3) Schematic diagram of spinal ligaments, (4) Right side acetabular fossa showing ossified

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transverse acetabular ligament. ALL- Anterior longitudinal ligament, SSL- supraspinous ligament, ISL- Interspinous ligament, LF- Ligamentum flava, STL- sacrotuberous ligament, TAL- Transverse acetabular ligament, AF- acetabular fossa.

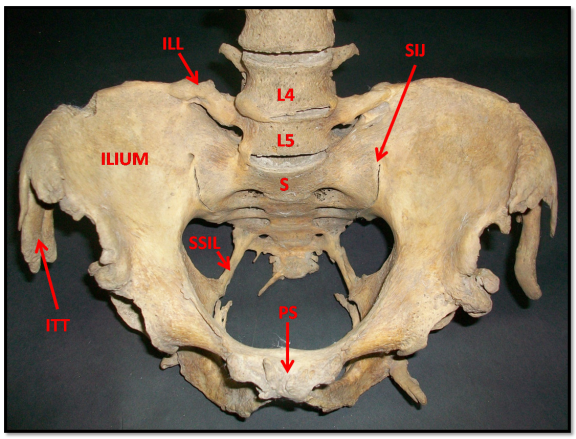

Fig-2: Superior view of articulated pelvis showing ossified pelvic ligaments. ILL- iliolumbar ligament, SIJ- sacroiliac joint, SSIL- sacrospinous ligament, ITT- iliotibial tract, PS- pubic symphsis, S- sacrum, L- lumbar vertebra, ILIUM- ilium.

Discussion

The ALL extends along the anterior surfaces of the vertebral bodies, from the basilar part of the occipital bone cranially to the front of the upper sacrum caudally. The PLL lies on the posterior surfaces of the vertebral bodies in the vertebral canal, attached between the body of second cervical vertebra and the sacrum, and continues with the membrana tectoria above. LF connects laminae of adjacent vertebrae in the vertebral canal. The SSL connects the apices of the spinous processes from 7th cervical to the sacrum. The ISL interconnects the spinous processes, from the root to the apex of adjacent processes and they run from the lower edge of one spinous process to upper edge of the next. These spinal ligaments provide stability to the vertebral column and help in maintenance of erect posture[6].

Ossification of these spinal ligaments can cause abnormal curvature of vertebral column like kyphosis as in the present case, compress neurological structures and lead to various related problems. Ossification of ISL and SSL associated with lumbar vertebrae can cause compression of cauda equina leading to loss of bladder and bowel control, numbness sensation in perineum and weakness in the thighs[9,10,11]. Ossified LF can contribute to spinal stenosis, most often in the lower thoracic or lumbar spine. Ossification of LF is classified into five types based on MRI findings: localized, continued, skip, combining with anterior pressure, and combining with cervical and/or lumbar stenosis[3]. In the present case, continuous type of LF was found. OSL can also lead to decreased spinal mobility and functional impairment of these patients. Ossification of the posterior longitudinal ligament (OPLL) is most commonly found in the cervical spine region. OPLL is classified into the following: local type, segmental type, continuous type, and mixed type based on plain radiographic findings[12]. In our present case, we did not find OPLL.

OSL is stated to be common in the Asiatic population and therefore, genetic factors are considered to be an important aspect for its prevalence. There have been many studies on collagen genes, including on the human collagen A2 gene (COL11A2) which showed their role in OSL. COL11A2 gene is located at chromosome 6p close to the human leukocyte antigen region[12]. Other factors which are also associated with OSL are retinoic X receptor b, transforming growth factor-b (TGF-b), and bone morphogenetic protein (BMP)[1,12].

Sacro-iliac joint (SIJ) consists of two types of articulation: (1) an antero-inferior synovial joint between the C-shaped auricular surfaces of the sacrum and ilium and (2) a postero-superior syndesmosis situated between the interosseous surfaces of the ilium and sacrum. The joint surfaces are irregular with ridges and depressions that are reciprocal and are greater in males. These irregular articular surfaces restrict the movements and contribute to the strength of the joint during the transmission of the weight from the vertebral column to the lower limbs. Stability of the joint is provided by the intrinsic ligaments i.e., anterior sacro-iliac ligament in the ventral region, posterior and interosseous ligaments in the dorsal region, and by the extrinsic STL and SCSL[13,6].

Ankylosis of the SIJ is a pathological condition due to formation of fibrous adhesions in SIJ and thus decrease in the size of the synovial cavity. In 15- 25% of cases of low back pain, the cause is ankylosis of SIJ. The major histopathological changes associated are the inflammation of the joint with erosion followed by gradual ossification of the joint resulting in ankylosis. It involves both the synovial joint and anterior and posterior sacroiliac ligaments. The ossification also extends to the neighboring ligaments, STL and SCSL. SIJ ankylosis is more frequent in men than in women. This is due to increased mobility of the joint after puberty and during pregnancy in women[13].

STL and SCSL are the key structural components of the pelvic cavity as they connect the pelvic bones to vertebral column. STL extend from the posterior superior iliac spine, lower sacral tubercles, lateral margins of the sacrum and coccyx to the medial margin of the ischial tuberosity. This attachment extends along the ischial ramus as the falciform process which blends with the fascial sheath of pudendal canal containing pudendal nerve and internal pudendal vessels. This ligament is pierced by coccygeal branches of inferior gluteal artery and perforating cutaneous branches of the coccygeal plexus. The SCSL extends from ischial spine to the sacrum. These ligaments jointly convert the greater and lesser sciatic notches into their corresponding foramina[6,13].

The pudendal nerve along with internal pudendal vessels traverses the greater sciatic foramen and then winds around the SCSL to enter into the lesser sciatic foramen, where it lies between the STL and SCSL. Sometimes, the pudendal nerve may travel through the SCSL[6,7,13].

Complete ossification of the SCSL and the partial ossification of the STL, as found in our case, may compress the structures passing through lesser or greater sciatic foramen, mainly the pudendal nerve, internal pudendal vessels, nerve to obturator internus and sciatic nerve. Compression of sciatic nerve leads to sciatica, pain radiating to the lower limb[13]. Other structures such as nerve to obturator internus and pudendal nerve compression may lead to perineal muscle weakeness, chronic perineal pain and pudendal nerve entrapment syndrome[7]. The pudendal nerve can get compressed at three possible sites, as it winds and passes through the fibers of SSL, between the STL and SCSL, and along the falciform process of the STL[14]. Pudendal nerve entrapment syndrome results in pain or loss of sensation in the perineal region[7,8]. The coccygeal branches of the inferior gluteal artery pierce the STL and supply the gluteus maximus. Ossification of STL may lead to the compression of the vessel and subsequently result in ischemia of the region supplied by it.

TAL alongside with the acetabular labrum deepens the acetabular fossa and so the cavity of hip joint. TAL helps to maintaining the stability of the hip joint. The space between TAL and the notch transmits medial circumflex femoral and acetabular branches of obturator vessels which supplies the acetabular fat and head of femur[6]. Thus, if this ligament gets ossified as in this case, may lead to the restricted movements of the hip joint and also leads to the compression of the vessels and consequently results in isch-

emia of the area supplied by it[13,15].

The iliotibial tract (ITT) is a thickened portion of the deep fascia (fascia lata) over the lateral aspect of thigh. Upper end of the tract splits into two layers. The superficial lamina is attached to the tubercle of iliac crest and deep lamina to the capsule of hip joint. Gluteus maximus and tensor fascia lata muscles get inserted into this upper part[6]. Thus ossification of ITT as observed in this case, may lead to pain during walking, running and hip abduction.

Fibroblasts cells are present in large number in ligamentous tissue and are reported to be linked with heterotopic ossification in ligaments. However, the proper mechanism by which fibroblasts mediate this remains uncertain. Scapinella, in a radiological study, reported ossification of supraspinous and infraspinous ligaments in patients. He concluded that it may be due to external stimulus which caused differentiation of fibroblasts into chondroblasts and finally into osteoblasts cells, leading to ossification[16]. Albert Oppenheimer conducted an X- ray study on ossification of vertebral ligaments. He inferred that ossification of ligaments is a method of healing of ligaments when there is a continuous increase in tension like in ligamental tear in trauma[17]. Ossified ligaments are formed mainly through endochondral ossification[2,18]. Histology of these ossified ligaments is composed of a lamellar bone with mature Haversian canals, in addition to fibrous cartilages and woven bones wrapped with calcified cartilage[12].

Such association of severe OSL together with SIJ ankylosis, is seen most commonly in Ankylosing Spondylitis. It is a chronic disease characterized by progressive stiffening of the joints. It has a predilection for the axial skeleton especially the joints of the sacroiliac and lumbar vertebrae. In severe cases it may involve hip[19], knee and manubrium- sternal joints and rarely temporomandibular joints[20]. Vertebral involvement is also characterized by ossification of various spinal ligaments. Genetic and environmental factors play a key role in pathogenesis of AS, as indicated by its strong association with histocompatibility antigen HLA-B27[9]. However, HLA-B27 contribution to AS genetic risk is approximately 16%[21]. Tumor necrosis factor-alpha (TNF-α) also has a pro-inflammatory function in AS[22].

Conclusion

In our specimen, such enormous degree of ossification of ligaments of axial skeleton is a rarity. Knowledge of this type of variations such as OSL, ossification of the SIJ, STL and SSL occurring as a single entity would be of great value clinically for precise diagnosis and treatment of the back pain, neurovascular compression syndromes and also during the reconstructive procedures of the pelvic floor.

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