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Ankylosing Spinal Hyperostosis or Forestier Disease: Difficulty in Diagnosing or Lack of Knowledge?

Abstract

The article demonstrates a clinical case of a disease that occurs mainly in older age groups — ankylosing spinal hyperostosis (ASH), or Forestier disease. The rarity and lack of knowledge of ASH and the associated lack of alertness in relation to this disease makes its diagnosis a challenge. The disease is more common in older people, which requires differential diagnosis with degenerative changes of the vertebrae. ASH is an oligosymptomatic disease. Clinical symptoms are determined not by hyperostosis of the ligaments and tendons by themselves, but by the development of reactive inflammation of these structures during the process of ossification, overloading of still moving segments of the spine adjacent to ankylosed vertebrae. The damage of the anterior longitudinal ligament, the continuity of its ossification for at least four vertebrae (in contrast to trauma, tumors) is of particular importance for this disease. The absence or low severity of ankylosis of the facet joints, sacroiliitis, paravertebral ossification symmetry, characteristic of ankylosing spondylitis, is also important. The absence or mild severity of degenerative changes in the intervertebral discs detected in osteochondrosis matters. The presence of laboratory and clinical signs of inflammation, damage to other organs and systems is not typical to this pathology. At present, we have no treatment standards for ASH, which also makes the choice of patient management approach complicated.

The insufficient amount of information about this disease makes wider coverage of the pathology necessary in order to improve diagnostic skills, timely and complete treatment.

Key words: *ankylosing spinal hyperostosis, Forestier disease, ossification of the anterior longitudinal ligament*

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ASH — ankylosing spinal hyperostosis

Ankylosing spinal hyperostosis (ASH), or Forestier disease, is a rare non-inflammatory disease, with ossification of the anterior longitudinal ligament of the spine. It is part of diffuse idiopathic skeletal hyperostosis, which is characterized by multiple ossification of the tendons, ligaments, aponeuroses,

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capsules of joints. ASH is more common in the elderly (94.5% of patients older than 60 years). The reference value specified is not true (source of information checked), and has been corrected to the true value [1]. The disease occurs without any external cause, and is therefore referred to as idiopathic. Rare occurrence makes it difficult to study the causal factors of ASH. Late onset is a reason to believe that ASH is caused by the aging processes occurring in the connective tissue. Much less often, similar changes occur in young or middle age, and in this case, this is a manifestation of some endocrine or metabolic disease.

The corresponding pattern, named "ankylosing senile hyperostosis" of the spine, was first described by J. Forestier and J. Rots-Querol in 1950. Subsequently, this pathological process was designated as "Forestier disease".

In most cases, the disease is manifested by a lesion in the thoracic spine, often capturing its central part, then the lumbar and cervical sections become involved in the process [2–4]. Ossification starts mainly on the right side, which is presumably due to the absence of aortic pulsation, which prevents calcification of the spine tissues on the left.

Diagnosis of ASH is based mainly on the analysis of X-ray changes [5]. A lateral view is crucial to get an image of the spine and anterior longitudinal ligament. Distinctive features of the disease are: ossification with a sharp thickening of the anterior longitudinal ligament along the entire anterior surface of the vertebrae from the occipital bone to the sacrum, the continuity and extent of ossification at the level of several vertebral segments, the absence of significant changes in the vertebral bodies and intervertebral spaces, and the symmetry of paravertebral ossifications. Less typical for ASH is the formation of osteophytes, ossifications of the posterior longitudinal ligament, and capsules of the facet joints.

ASH is an oligosymptomatic disease. However, debilitating daily pain in many parts of the spine that do not respond to standard therapy may be encountered.

Clinical symptoms: pain, discomfort, stiffness in different parts of the spine are determined not so much by hyperostosis of the ligaments and tendons themselves, but by the development of reactive inflammation of these structures in the process of ossification, overloading of still-mobile segments of the spine adjacent to ankylosed vertebrae. If the thoracic spine is affected, there may be a restriction of the respiratory excursion of the chest; if the cervical spine is involved in the process, dysphagia, dysarthria occur due to compression of the esophagus, larynx, and trachea. In the case of hypertrophy of the posterior longitudinal ligament, there is a threat of compression of the spinal cord with the development of myelopathy, paresis and paralysis [6]. Markers of inflammation do not exceed the norm.

Since there is no clear understanding of the factors that cause ASH, its specific treatment is the subject of exploration and research. To alleviate the condition of patients, symptomatic therapy is used, including the prescription of anti-inflammatory drugs (meloxicam, celecoxib, ibuprofen), magnetic therapy, laser therapy, hydrotherapy (hydrogen sulfide and radon general health baths), massage, exercise therapy (remedial gymnastics). Radiating pain is relieved by local administration of glucocorticosteroids and anesthetics, application of anti-inflammatory medicines, and the prescription of phonophoresis of hydrocortisone ointment. In case of an adverse course of the disease, especially accompanied by compression of internal organs, surgical treatment is possible: using the Cloward technique, the enlarged ossified anterior longitudinal ligament is removed and a fixing plate is implanted on the anterior surface of the vertebral bodies.

The rarity of this pathology makes it preferable to cover individual clinical cases of the disease in order to improve its diagnosis and find the optimal treatment approach.

Here we present a clinical case that demonstrates the peculiarities of the symptoms and diagnosis of ASH.

Patient P., 58, salesperson, was admitted to Rheumatology Department of SHCI "Regional clinical hospital" with complaints of constant pain at the front of the neck, not connected with movements and not increasing at rest, "girdle sensation" around the neck, difficulty in swallowing, especially solid food, dysfunction of phonation of the voice, hoarseness, difficulty of movement in the cervical spine when bending forward and to the side.

From the history of the disease, it is known that in 2016 (at the age of 56 years), she was observed by

a neurologist with complaints of neck pain and related difficulties in spine movements. The diagnosis of osteochondrosis of the cervical spine was established, and drug and physical therapy was carried out with a positive effect. The changes detected during magnetic resonance imaging (MRI) in the cervical spine — ossification of the anterior longitudinal ligament with a length of 35 mm — were not properly evaluated (Figure 1).

In the summer of 2018 (at the age of 58), she suffered acute respiratory infection complicated with purulent otitis. The patient was successfully treated by an ENT specialist, but soon began to notice the above complaints. Outpatient fibrolaryngoscopy was performed, which revealed the mobility of the vocal and scapular ligaments, their symmetry during phonation, a wide glottis, free sinuses, and the absence of changes in the epiglottis and subglottic space. Along the posterior wall of the oropharynx (at the level of the upper edge of the epiglottis), a submucosal formation up to 2 cm in diameter with mucosa that was unchanged and displaced above it was visualized.

To clarify the diagnosis, standard computed tomography (CT) of the larynx, pharynx and cervical spine was performed. There were pronounced structural changes in the cervical spine with the straightening of the cervical lordosis, fragmentary ossification of the posterior longitudinal ligament that protrudes into the vertebral canal. On the ventral surface of the vertebral bodies, massive bony growths of the ossified anterior longitudinal ligament were determined with thickness of up to 9 mm on the right and 11 mm on the left, with the axial size up to 14 26 mm, merging with each other in the vertical direction, forming a bony array up to 77 mm long (increased more than 2-fold in comparison with the year 2016). The dorsal sections of the oropharynx and esophagus wall were pushed back by bone growths ventrally, and had an asymmetric localization. The left sections of the upper and middle constrictors of the pharynx, the palatopharyngeal tonsil, the aryepiglottic fold, the vestibular fold and the posterior wall of the pharynx are thickened relative to the right sections by 0.3–2.6 mm. The oropharyngeal lumen is asymmetric due to the reduction of the size on the left. The epiglottis is located slightly obliquely with ventral displacement of the left sections. Its caudal section on the left was twice as wide as the right half. As a conclusion of the study it was noted that the deformation of the oropharynx and asymmetry of the dorsal structures forming the rear wall with thickening and an increase in the volume on the left are due to the effect of disfiguring ossifying ligamentosis of the anterior longitudinal ligament of the cervical spine. Incomplete closure of the vocal cords during phonation with diastasis between false folds of 0.2 mm and between true folds of 1.3 mm was revealed (Figures 2, 3).

When examined in hospital, the patient's appearance is unremarkable. There is a slight smoothing of the physiological curves of the spine. Movement in the thoracic and lumbar spine without restriction: Thomayer test is 3 cm, Schober and Ott test are 5 cm, chest excursion is 5 cm. Certain deviations were detected from the side of cervical region: distance between the chin and the sternum is 3 cm, the angle of lateral inclination of the cervical spine in both directions is less than 45 degrees. There is no pain in the spinous processes throughout the spine. The joints are not changed, movements there are not restricted. There was no abnormalities on the part of internal organs.

In order to exclude the disease from the group of seronegative spondyloarthritis, laboratory parameters were studied: complete blood count (CBC) and urinalysis (UA) — no pathological changes; C-reactive protein, glucose, cholesterol, alkaline phosphatase, rheumatoid factor within normal limits, HLA-B27 antigen was not detected.

No abnormalities were observed in the MRI examination of the sacroiliac joints. Repeated CT scans further revealed ossification of the anterior longitudinal ligament along the anterior border of the vertebral bodies throughout the thoracic and lumbar spine, most pronounced at the level of Th5–Th10, Th12–L3 vertebrae, no changes in the shape of the vertebral bodies were observed.

The examination of the patient made it possible to establish the following diagnosis: *Ankylosing hyperostosis of the spine (Forestier disease).*

The patient was treated with etoricoxib 60–90 mg/day, magnetic therapy, which led to the complete disappearance of neck pain and difficulties in swallowing. The patient was further advised follow up by a rheumatologist and, if necessary, drug or surgical treatment.

Figure 1. *Magnetic resonance imaging of the cervical spine. Ossification of the anterior longitudinal ligament over 35 mm*

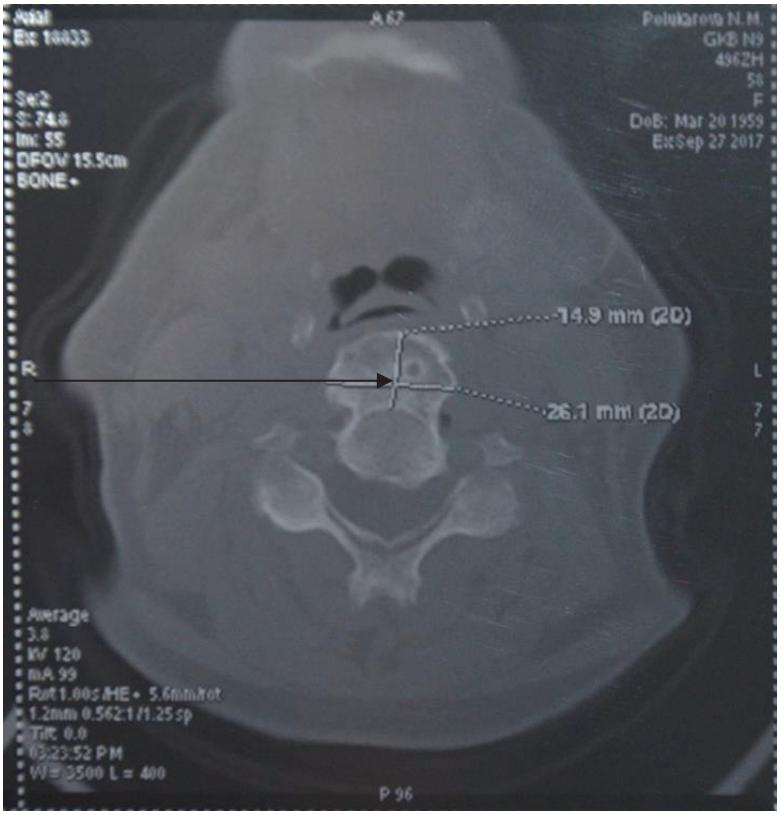

Figure 2. *Computed tomography of the cervical spine, axial view. The arrow indicates the ossification of the anterior longitudinal ligament on the anterior surface of the vertebral body measuring 14.9 × 26.1 mm*

Figure 3. *Computed tomography of the cervical spine, lateral view. Arrows indicate osteophytes of the anterior longitudinal ligament, merging with each other in the vertical direction, forming a mass 77.7 mm long, up to 16.0 mm wide*

Discussion

Nowadays, more and more attention is paid to the changes occurring in the body of elderly people, including the presence of chronic inflammation, pain syndrome, selection of adequate, safe therapy. Back pain occurs in almost every person during life, but most often in older people. The pain syndrome can be associated with different pathological processes. However, physicians, especially at the primary level, when dealing with back pain in older patients, do not always pay sufficient attention to diagnostics, linking the symptoms to the most common degenerative-dystrophic diseases. Awareness and a broad-minded approach of doctors will not only contribute to early diagnosis, but will also increase patient adherence to treatment, reduce polypragmasia, which is especially important in elderly patients. In our work, we demonstrated a clinical case of a disease that occurs mainly in elder patients.

The rarity and insufficient knowledge of ASH, and the associated lack of alertness in relation to this disease, make the diagnostic search extremely difficult for the doctor and, sometimes, severe for the patient [7]. The resemblance of ASH in many aspects to some diseases accompanied by spinal cord affection (tumors, injuries, osteochondrosis, ankylosing spondylitis, etc.) complicates diagnosis. In this regard, it is necessary to use the most informative methods of research, CT and MRI, taking into account diagnostic signs (D. Resnica and Y. Niwayama, 1988), which helps to better understand features of symptoms and differential diagnosis of ASH [6]. Noting the predominant lesion of the anterior longitudinal ligament in ASH, the authors consider the first sign of the disease to be the continuity of its ossification for at least four vertebrae. This condition makes it possible to distinguish ASH from such pathological conditions as infections, tumors, and injuries characterized by local processes in the spine. The second feature, according to the authors, is the absence of ankylosis of the facet joints and sacroiliitis, which are characteristic of ankylosing spondylitis. The lack of symmetry of paravertebral ossification, clinical and laboratory signs of inflammation in ASH can be added to that. The third sign of ASH from the point of view of said authors is the absence (or mildness) of degenerative changes in the intervertebral discs (decrease in their height, marginal sclerosis of the vertebral bodies), which are detected in osteochondrosis.

In addition to these signs, it is also possible to note the absence of damage to other organs and systems characteristic of ASH, with the exception of ectopic ossification of the enthesis, ligaments and tendons.

All specific features of ASH, including age, correspond to the signs that were noted in our patient, which suggests the validity of the diagnosis of ASH.

Conclusions

The rarity and insufficient knowledge of ASH makes it necessary to discuss this disease more widely in order to develop diagnostic skills and provide timely and proper care to patients.

Distinctive features of ASH are: the elderly age of patients, the continuity of ossification of the anterior longitudinal ligament of at least four vertebrae, the absence or weak severity of degenerative changes in intervertebral discs, X-ray signs of ankylosis of facet joints, and the symmetry of paravertebral ossifications of other organs and systems.

In our case, the diagnosis was made one year after the MRI scan, where the ossification of the anterior longitudinal ligament was not properly assessed. According to the literature, the path to establishing the diagnosis of ASH is complicated and long: osteochondrosis, expansive processes of the esophagus and larynx are detected during repeated CT scans, in some cases tracheostomy is performed and biopsy of the formation is recommended before the diagnosis of ASH is verified [1, 5–6].

Contribution of Authors

E. N. Skryabina: the concept of the article, analysis, interpretation of data, writing of the manuscript, verification of intellectual content, approval of the manuscript for publication.

N. A. Magdeeva: analysis, interpretation of data, writing of the manuscript, verification of intellectual content.

Yu. M. Korneva: data collection, analysis, and interpretation of results.

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