CASE REPORT

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UDC: 616-036.886 DOI: https://doi.org/10.2298/VSP190522022V



The analysis of a sudden death caused by the unusual shape of the *foramen magnum* stenosis

Analiza iznenadne smrti izazvane neobičnim oblikom stenoze velikog potiljačnog otvora

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Abstract

Introduction. The anomalies of the size and shape of the foramen magnum (FM), followed by its stenosis, take a special place in clinical and forensic practice. The clinical picture of FM stenosis is variable, but it is not specific, which is why these anomalies are sometimes not recognized in time and are only detected by autopsy. Case report. This paper analyzes the case of a sudden death caused by the unusual shape of the FM stenosis. On autopsy, we found exostoses on the front and rear edges of the FM with their prominence into the FM. The exostoses reduced the sagittal diameter of the FM and practically divided it into two asymmetric semicircles, which is why we called this stenosis the "keyhole" stenosis of the FM. This form of the FM stenosis pushes the medulla oblongata aside, resulting in its incarceration into one of the FM semicircles. Conclusion. A stenosis of the FM can represent a threat to life, especially when combined with its unusual shape, as in our case. In this way, the critical stenosis of the FM becomes clinically clearly manifested, and there is a loss of consciousness that can also end with a sudden death, as in our case.

Key words:

constriction, pathologic; death, sudden; foramen magnum; forensic pathology.

Apstrakt

Uvod. Anomalije veličine i oblika velikog potiljačnog otvora (foramen magnum - FM) praćene njegovom stenozom zauzimaju važno mesto u kliničkoj i forenzičkoj praksi. Klinička slika stenoze FM je raznovrsna, ali nije specifična, zbog čega ovakve anomalije ponekad nisu na vreme prepoznate i otkrivaju se tek autopsijom. Prikaz bolesnika. U ovom radu analiziran je slučaj iznenadne smrti izazvane neobičnim oblikom stenoze FM. Autopsijom su nađene egzostoze na prednjoj i zadnjoj ivici FM sa prominencijom tuberkula egzostoza u sam otvor. Egzostoze su znatno redukovale sagitalni dijametar FM i praktično ga podelile u dva nejednaka polukruga, zbog čega smo ovakvu stenozu označili kao "ključaonica" stenozu. Ovakav oblik stenoze FM potiskuje produženu moždinu u stranu, sa njenim posledičnim uklještenjem u jedan od polukrugova FM. Zaključak. Stenoza FM može predstavljati pretnju po život, posebno kada je kombinovana sa njegovim neobičnim oblikom. Na ovaj način kritična stenoza FM postaje klinički jasno manifestna, nastaje gubitak svesti koji može da završi iznenadnom smrću kao u našem slučaju.

Ključne reči: suženje, patološko; smrt, iznenadna; foramen magnum; patologija, sudska.

Introduction

The anomalies of the size and shape of the *foramen* magnum (FM), followed by its stenosis, take a special place in clinical and forensic practice. The clinical picture of the FM stenosis is variable, but it is not specific, which is why these anomalies are sometimes not recognized in time and

are only detected by autopsy. The FM is characterized by the reduction of the diameter and the surface of the FM opening. The outcome of the stenosis is not only determined by the values of the diameter and the surface of the FM, but also by the shape of the FM. We found only a few cases in the literature in which a sudden death was described in association with the unusual shape of the FM stenosis ^{1, 2}.

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We presented and analyzed the occurrence of a sudden death caused by the unusual form ("keyhole") of the FM stenosis.

Case report

A 15-year-old boy suddenly passed away during the game of rope jumping. In the medical chart, it was noted that he had had occipital headaches, dizziness, languor and occasional loss of consciousness. An autopsy confirmed the FM stenosis, which was induced by exostoses on the front and rear edges of the FM. The antero-

posterior diameter of the FM was 13 mm. In addition to stenosis, we also found pronounced enlarged and asymmetric jugular tubercles of the occipital bone (Figure 1). The diameter of the *medulla oblongata* (MO) at the level of the FM was 12 mm.

The signs of compression of exostoses were visible as impressions and deformations of the shape and crosssection of the MO in the stenosis zone (Figure 2).

On microscopic examination, we found extensive edema, the collapse of the central canal in the compression zone, dilatation of the central canal above the compression level and partial gliosis (Figure 3). On the peripheral parts,

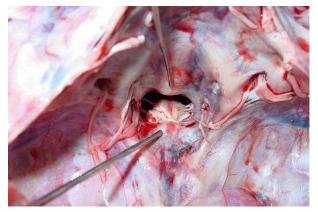


Fig. 1 – "Keyhole" stenosis of the *foramen magnum*.

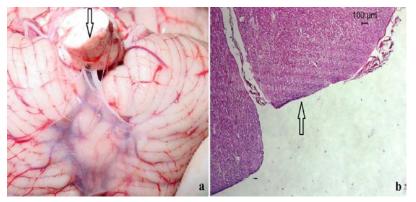


Fig. 2 – Deformations of the shape and cross-section of the *medulla oblongata* in the stenosis zone: a) macroscopic examination (arrow); b) microscopic examination (arrow), hematoxylin-eosin staining, ×100.

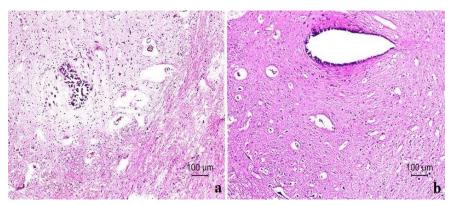


Fig. 3 – Microscopic examination of the brainstem (hematoxylin-eosin staining, ×100): a) collapse of the central canal at the level of compression; b) dilatation of the central canal above the level of compression.

we also found rare microtraces of iron in the tissue as a part of the decaying blood products (Figure 4). degree of the FM stenosis, the *medulla* made direct contact with them. The contact surfaces between them were semicir-

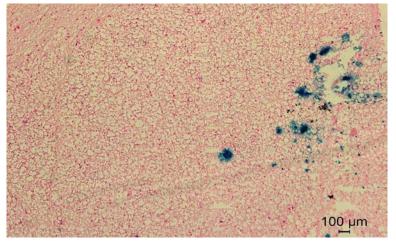


Fig. 4 – Perls' Prussian blue staining (×100) shows rare microtraces of iron in the tissue as a part of the decaying blood products.

Discussion

In the reviewed literature, most of the sudden deaths caused by the FM stenosis have been associated with achondroplasia ³, but only a few cases with bony formations at the rim of the FM and subsequent lethal stenosis have been described ^{1, 2}.

The average length of the sagittal diameter of the FM determined by computerized tomography is 33.1 ± 3.5 mm, while the average length of the transversal diameter is 27.6 ± 3.1 mm. The FM surface area is 729.15 ± 124.87 mm^{2 4}. Vasović ⁵ states that the values of the FM diameters depend on the life age (Table 1).

Table 1

| Mean values of sagittal and transversal diameter of |
|---|
| the <i>foramen magnum</i> according to age ⁵ |

| the jor amen magnum according to age | | |
|--------------------------------------|---------------|---------------|
| Age | Sagittal | Transversal |
| | diameter (mm) | diameter (mm) |
| 1st day | 20.8 | 15.3 |
| 6 months | 28.7 | 22.7 |
| 2 years | 29.5 | 25.8 |
| 3–6 years | 31.5-32.5 | 25.8-27.7 |
| 8–10 years | 33 | 28 |
| 15 years | 34.4 | 28.4 |
| > ww15 | 35 | 28.4 |
| years | | |

In our case, the diameter of the MO at the level of the FM was 12 mm, while the antero-posterior diameter of the FM in its narrowest part was 13 mm. In addition, the clinical data on the presence of occipital headaches, languor, dizziness, and intermittent loss of consciousness may have indicated a chronic compressive effect on that portion of the MO. Consequently, due to such narrowing and asymmetry of the jugular tubercles, we present a mechanism that explains the sudden death in this case.

The MO crosses over the most protruding points of the semicircular exostoses of the FM. Due to their size and the

cular and smooth, which is why the slipping of the MO into the left or right semicircle of the FM was facilitated. Head movements in the atlantooccipital joint in the form of flexion and/or rotation, as well as rotational brain acceleration in head trauma, contribute to the movement of the MO into one of the FM semicircles. Within the movements of the cervical spinal column, there are movements of the cervical spinal cord that is in direct anatomical relation to the MO. The movement of the MO aside pulls its rostral thicker part toward the FM. Its displacement to the side is confirmed by the deformation of the cross-sectional shape with the exostoses impressions on the posterior contour. In this way, in conjunction with the compressive effect on the ipsilateral vertebral artery, whose V4 segment passes through the FM, in our opinion, it had led to a sudden deterioration that resulted in the incarceration of the MO into one of the FM semicircles with the consequent reflex shutdown of vital centers in the brainstem, which eventually led to the fatal outcome. Because of this stenosis, the movement of the MO to the side had existed before, but not to the degree that would be termed as lethal incarceration.

All other possible causes of the sudden death, both by macroscopic and microscopic examination, were excluded. A toxicological analysis was not performed in this case because there were no indications that it was needed.

The vertebral arteries originate from the subclavian arteries and, as a result, there is a competition between the vertebrobasilar circulation and the circulation of the upper limbs. Some functional conditions of the organism that affect the circulatory flow rate through the vertebral arteries can contribute to final ischemic effects on the brainstem in the critical FM stenosis. Elevated body temperature redirects a portion of the circulatory volume to the upper extremities at the expense of the circulatory volume of vertebral arteries ⁶. Pamphlett et al. ⁷ demonstrated the possibility of the compression of vertebral arteries in the stretching of the neck or rotation of the head in the experimental model of postmortem angiography in neonates. Saternus and Adam ⁸ state that the compression of the vertebral arteries is possible in other positions of the neck and head. Excessive stretching of the neck and head rotation is especially possible in infants since they have instability of atlantooccipital joint ⁹. The compression of the vertebral arteries is thought to be associated with sudden infant death syndrome ^{10, 11}.

When it comes to critical FM stenosis, the question that arises is when the FM stenosis can be assessed as critical, or when it becomes a potential threat to the life of a patient. Vakili et al. ¹ described the lethal ischemia of a brainstem in atlantooccipital fusion in a 24-year-old man, where the antero-posterior FM diameter was 16 mm, i.e. 45.7% of the normal value. In our case, the antero-posterior diameter of the FM in the boy aged 15 years was 13 mm, which is 37.8% of the normal diameter value

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for the age of 15 years. The values of the FM diameter and its shape, which can be obtained by radiological methods, can be parameters for assessing a patient's life-threatening condition.

Conclusion

A stenosis of FM can represent a threat to life, but when the stenosis is combined with an unusual shape of the FM, as a "keyhole" stenosis in our case, it can cause consequent incarceration of the *medulla oblongata*. In this way, the critical stenosis of FM becomes clinically clearly manifested, and there is a loss of consciousness that can also end with a sudden death, as in our case.

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Received on May 22, 2019 Revised on February 3, 2020 Accepted on February 28, 2020 Online First March, 2020