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THE CASE OF HEMIFACIAL MICROSOMY IN BLOOD BROTHERS

Abstract: *The aim of this work was to present a clinical observation of a case of hemifacial microsomia in blood brothers who applied to maxillofacial surgery. Patient S. 31 years old. He entered the department of maxillofacial surgery of the city medical association of Samarkand with the aim of obtaining a medical opinion about VTEK. Goldenhar syndrome (oculoauriculo-vertebral dysplasia, hemifacial microsomia, disease of the oculoauriculo-vertebral spectrum) is a rare congenital disease associated with damage to structures emanating from the first and second branchial arches.*

A clinical example indicates the need for a comprehensive examination and a thorough collection of anamnesis of the disease of patients with Goldenhar syndrome for the timely implementation of appropriate treatment and rehabilitation measures and improving the quality of life of children.

Key words: *Goldenhar syndrome, atresia of the external auditory canal, hypoplasia of facial muscles.*

Language: *English*

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Introduction

The urgency of the problem

Goldenhar syndrome (oculoauriculo-vertebral dysplasia, hemifacial microsomia, disease of the oculoauriculo-vertebral spectrum) is a rare congenital disease associated with damage to structures emanating from the first and second branchial arches [2; eight]. Goldenhar syndrome got its name from the name of the American doctor who first described it in the middle of the last century. Since then, little information has been added about this pathology, due to its rarity and complexity of study, however, thanks to modern technologies, it is not only possible to diagnose it in utero, but also to be effectively treated. There are no domestic epidemiological studies on the incidence of Goldenhar syndrome, but, according to

foreign authors, it ranges from 1/3500 to 1/7000 live births and occurs in 1 case per 1000 children with congenital deafness [4]. The ratio of the disease among boys and girls is approximately 3: 2. The probability of the birth of a subsequent child with this disease is less than 1%, the probability of transmission of the disease to their children is less than 3% [3]. The etiology and type of inheritance are not well understood. Typical signs of Goldenhar syndrome are facial asymmetry and hypoplasia of the lower jaw, microtia and / or ear protrusions, which occur in 100% of cases [7]. In 85% of cases, anomalies are observed on one side, and bilateral lesions are also noted from 10 to 33% of cases. According to the literature, the right side is affected more often [4]. Combined conductive and sensorineural hearing loss occurs in 50% of cases [3].

Purpose of the research

Presentation of a clinical case report of hemifacial microsomia in blood brothers by reference.

Materials and Methods

A case from practice. Patient S. 31 years old. He entered the department of maxillofacial surgery of the city medical association of Samarkand with the aim of obtaining a medical opinion about VTEK. During the collection of the anamnesis, it turned out that in addition to the patient in the family, the younger brother also has the same pathology, for which the parents have not consulted the doctors until now. According to the mother, the children were born on time, by natural birth. On external examination, the elder brother shows a pronounced asymmetry of the face due to hypoplasia of the facial muscles and underdevelopment of the body, the branches of the lower jaw, as well as the temporomandibular joint on the left, aplasia of the auricle and atresia of the ear canal on the left (Fig. 1). The younger brother has a hemifacial microsomia on the left, the auricle on the left was presented in the form of a weakly expressed cartilaginous ridge without a lobe, the auditory meatus is absent, the auricle on the right is deformed, there is a preauricular skin process (Fig. 2).

Results and Discussions

Patient S. in the department underwent a comprehensive examination: ECG: sinus rhythm, increased electrical activity of the right ventricle. Chest X-ray: no focal infiltrative changes were found. Ultrasound of internal organs: liver, gallbladder, spleen, adrenal glands without pathology. General and biochemical blood analysis, general urine analysis without pathology. Ophthalmologist's consultation: no pathology. ENT doctor's consultation: congenital anomaly of the left ear (grade III microtia, atresia of the external auditory canal, grade III hearing loss). On the right, hearing is not impaired. Recommended: observation of an ENT doctor and maxillofacial surgeon. Taking into account the age of patient J., the patient was sent to the regional multidisciplinary children's clinical hospital in the city of Samarkand, with subsequent examination by specialists.

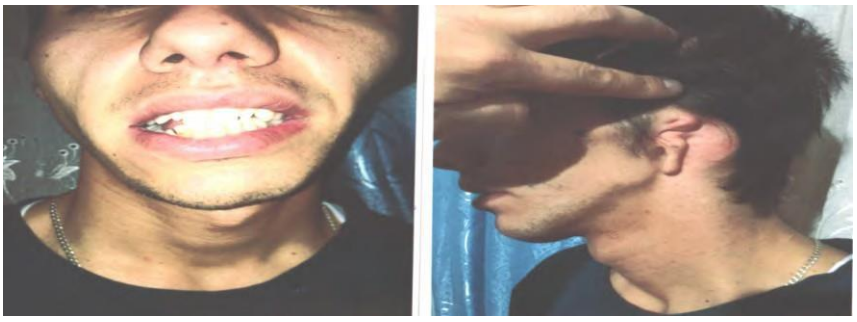

Fig. 1. Patient S. 31 years old. On external examination, the older brother has a pronounced asymmetry of the face due to hypoplasia of the muscles of the face and underdevelopment of the body, branches of the lower jaw, as well as the temporomandibular joint on the left, aplasia of the auricle and atresia of the auditory meatus on the left

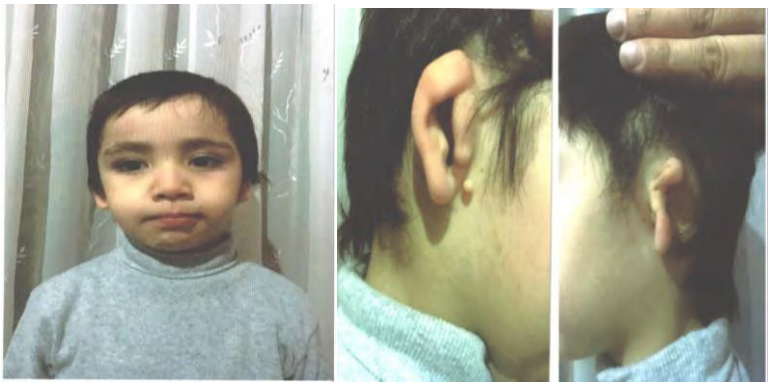

Fig. 2. Patient J. 8 years old. a. front view, b. Side view. The younger brother has a hemifacial microsomia on the left, the auricle on the left was presented in the form of a weakly expressed cartilaginous ridge without a lobe, the auditory canal is absent, the auricle on the right is deformed, there is a cutaneous preauricular process

Conclusion

A clinical example indicates the need for a comprehensive examination and a thorough collection of anamnesis of the disease in patients with Goldenhar syndrome for the timely implementation of appropriate treatment and rehabilitation measures and improving the quality of life of children.

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