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SURGICAL TREATMENT OF THE PATIENT WITH STURGE-WEBER SYNDROME- A CASE REPORT

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Abstract

Vascular malformations associated with genetic syndromes lead to dysfunction of human body organs by blood supply impairment. Disorders in the structure of blood vessels walls result in developing pathological lesions called hemangiomas. The case describes a 16-year-old male patient with Sturge-Weber syndrome (SWS) who was treated surgically due to hemangioma of upper jaw. The patient suffered from typical for Sturge-Weber syndrome congenital disorders of other organs. In these cases surgical treatment requires deep analysis and proper procedure planning in the context of hemostasis.

Key words: Sturge-Weber syndrome, hemangioma, cavernous hemangioma, surgery, general anesthesia

Sturge-Weber Syndrome (SWS) is rarely occurring congenital disorder syndrome. It is characterized by impairment of nervous system, eyes and skin disorders. Extra orally SWS manifests by unilateral port-wine stains accompanied by glaucoma, seizures and mental impairment [1]. Oral cavity lesions in the process of SWS manifest in various ways. Diagnosing of their origin is based on clinical examination [2]. Intraoral hemangiomas of upper or lower jaw and port-wine stains on the face occur ipsilaterally[1].

SWS is the third most commonly occurring congenital disorders syndrome, just behind neurofibromatoses and tuberous sclerosis, associated with nervous system and skin malformations. It presents in approximately 1 in 20,000 newborns. SWS is not inherited, occurs sporadically, with the same frequency in males and females, regardless of the race or ethnic origin [2, 3]. Mutation in GNAQ gen is considered to be the genetic cause of SWS and its isolated port-wine stains. If there are any port-wine stains in infant on the upper half of the face the risk of SWS occurring fluctuates from 15 up to 50%. The probability of vascular malformations in brain and eye depends on the extensiveness of port-wine stain [3]. In about 15% of all cases disorders in brain presents bilaterally. Prognosis is unfavourable [4]. Venous plexus lying under leptomeninges is bereft of venous cortical drainage [2].

Observations and screening examinations have a great importance for proper and early diagnosis, what leads to treatment introduction at an early stage. Laser treatment, using eye drops, glaucoma operations, taking low doses of aspirin, anticonvulsants, neurosurgical operations are a few treatment options for the patients with SWS [3]. Also surgical treatment of vascular malformations in other regions of the body ought to be considered.

Taking into consideration all complications, which may occur during treatment and patient's general health condition status, it is indispensable for the doctor diagnosing the disease and surgeon to have a great knowledge of the syndrome of congenital disorders and coexisting diseases [2].

Case report

16-year-old patient was referred to the Oral Surgery Department for the consultation and the treatment of the upper jaw tumor in the region of tooth 23.

In patient's medical history, Sturge-Weber Syndrome with coexisting glaucoma, seizures, deep mental impairment and port-wine stains on the left cheek. No drug allergies. In 2007 the patient underwent glaucoma operation in the left eye. Due to deep mental impairment the patient had undergone all the procedures of the extraction of deciduous and permanent teeth in the general anesthesia. Patient was taking anticonvulsants and eye drops.

In extraoral examination diagnosed disorders of cranium development and the port-wine stain on the left cheek (Fig. 1).

Intraoral examination showed numerous missing teeth. In the region of tooth 23 on the alveolar process crest localized hypertrophic tuberous lesion, 1 cm diameter, soft, covering decayed tooth 24 (Fig. 1).

After a deep analysis of the patient's medical history, clinical examination and an assessment of possible complications, surgery was planned – resection of the upper jaw tumor together with decayed tooth 24 extraction.

The surgery operation plan along with possible complications was presented to the patient and his guardian. All the surgery permissions were collected. Pre-operative medications and laboratory blood tests were recommended. A day of the surgery was appointed.

The patient and his guardian were informed about the details of postoperative recommendations and obligatory follow-up the day after surgery.

In general anesthesia the tumor of the upper jaw in the region of the tooth 24 was excised and the decayed tooth 24 was extracted (Fig. 2). Pathological lesion was excised with clinical margins. Particular attention was paid to proper hemostasis. Excised tissues were sent to histopathological examination.

Intra- and postoperative course were without complications. Patient in a general good condition was discharged from the clinic in guardian's care.

Control examination was performed the day after operation. Proper healing of the wound was observed. The follow-up and sutures removing were recommended for three weeks.

The histopathological examination confirmed the initial diagnosis of a cavernous hemangioma, excised completely. Currently the patient stays under Oral Surgeon's care.

Discussion

Sturge-Weber Syndrome is rarely occurring syndrome of neurocutaneous congenital disorders. Port-wine stains on the face skin innerved by the first branch of trigeminal nerve, leptomeninges vessels disorders, epileptic attacks, glaucoma, increased risk of a stroke, mental impairment are main features of SWS. It is assumed that somatic mosaic mutation is the cause of Sturge-Weber Syndrome and port-wine stains manifestation on the skin. Symptoms and the advancement of the syndrome depend on the period of embryogenesis, in which the mutation occurred [5].

Understanding the disease process in the patients with SWS has been improved since the first description of Sturge-Weber Syndrome in 1879. Last research results report about locating the somatic mutation in GNAQ gen as a causative factor of SWS and port-wine stains on the skin face. Current medical reports show growing problems in patients with SWS, which are associated with otorhinolaryngological, endocrinological, behavioral and emotional aspects. Neuropsychology and imaging examinations of nervous tissue allow observing the changes in the disease process of SWS over the years. An early diagnostics and implementation of proper proceeding in the patients with SWS is indispensible for an appropriate care and control of the whole treatment process [6].

SWS is also described as a encephalotrigeminal angiomatosis with the prevalence of vascular component. Intraorally it manifests by the presence of gingival hemangiomatosis uni- or bilaterally in the region of upper and lower jaw. The second variant occurs more rarely [7].

In the case described in our article, intraoral manifestation of SWS was the hemangioma of upper jaw on the left side. Ipsilaterally a port-wine stain manifested on the cheek. A proper assessment of the tumor and a suspicion of hemangioma allowed for an appropriate excision of the lesion. A central vessel supplying the tumor was located and banded.

An ability of bleeding control is a key factor for each practitioner during surgery procedures. Hemostasis may be obtained by applying local surgery dressing or in case of failure by different hemostatic medications, as an addition to basic methods [8].

In the presented case an appropriate vessel banding resulted in its closure and stopped bleeding. Excised tumor was diagnosed in its histopathological examination as cavernous angioma, which is a rare finding pathological lesion in the oral cavity. Cavernous malformations of vessels consist of dilated vessel canals [9]. Due to their infrequent occurrence in the oral cavity, cavernous hemangiomas require special experience and care in the proceedings of oral surgeons.

Despite the fact that patients with Sturge-Weber Syndrome are relatively small group of surgery patients, the planning of surgery procedures ought to be prepared with particular thoroughness. Because of vascular malformations, which occur in SWS, a surgeon should take into consideration the potential bleeding and provide a proper protection of hemostatic agents. Any

tumor-like lesions in the region of oral cavity and in the other regions of human body in patients with SWS ought to be diagnosed as potential hemangiomas. Worth noticing is the fact that hemangiomas are supplied by the central vessel which locating and banding has a great influence on the success of the operation. Due to many human organs disorders the patients with Sturge-Weber Syndrome are a kind of challenge for oral surgeons in surgery proceedings.

Conclusions

Each tumor-like lesion in patients with vascular malformations syndroms ought to be considered as a potential hemangioma.

Due to a great risk of massive haemorrhage during angiomas surgery operations, all surgical procedures should be performed with presence of haemostatic agents.

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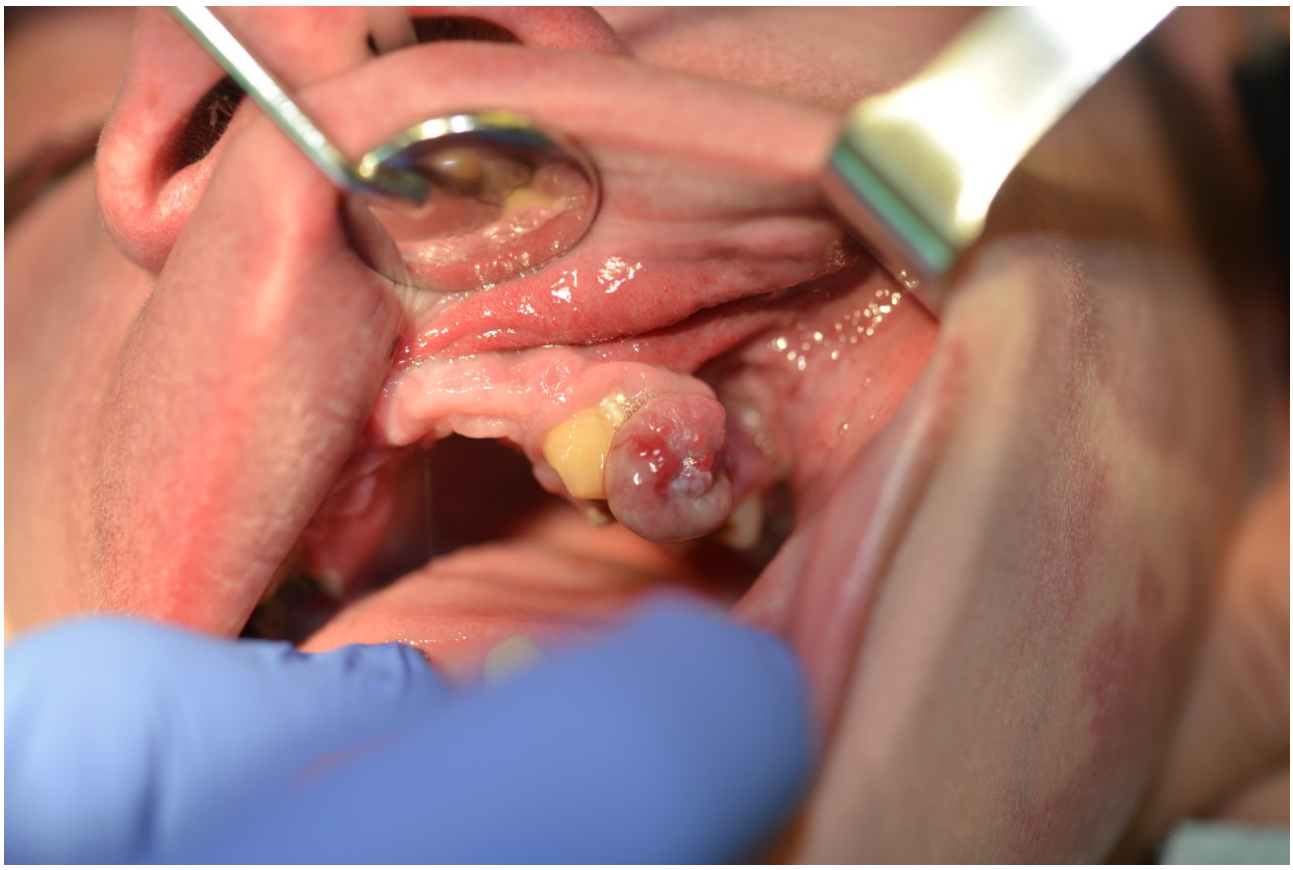
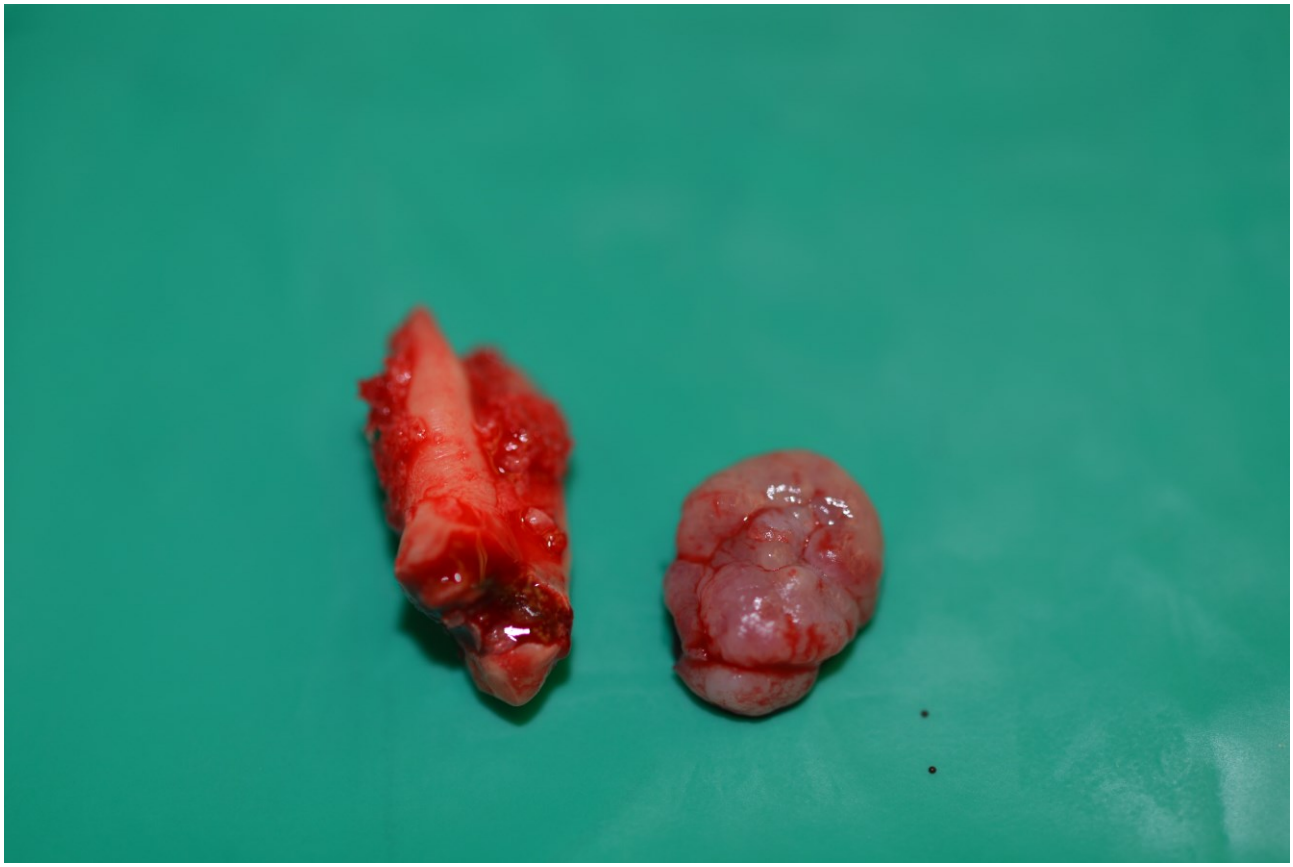


1. Port-wine stain on the left cheek.



2. Tumor of upper jaw along with decayed tooth 24.