СЛУЧАЙ ИЗ ПРАКТИКИ

INTRAMUSCULAR ARTERIOVENOUS MALFORMATION IN RIGHT MASSETER WITH TURKEY WATTLE SIGN

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Purpose. To discuss a rare tumor known as arteriovenous malformations (AVMs), which is a condition where an artery and a vein are directly connected. The masseter muscle is the commonly affected site, with 5% of cases being reported. This condition can be confirmed through radiological diagnostic methods such as ultrasonography and cone-beam computed tomography.

Materials and methods. A clinical case of a 46-year-old female patient who has had swelling in her right cheek region for two years, accompanied by intermittent pain while chewing and enlargement of a mass when lying down on her right side. Orthopantomography was advised to rule out odontogenic origin, followed which CBCT and ultrasonography was advised.

Results. Cone-beam computed tomography revealed multiple calcified nodules involving the masseter region measuring roughly 2x2mm in size. US revealed an irregular soft mass in the inferior border of the right masseter muscle measuring roughly 17x9mm with clusters of venous channels noted within it, as well as areas of calcification.

Discussion. Arteriovenous malformations (AVMs) are rare tumors that are uncommon in the head and neck region, and less than 1% of vascular tumors are found in muscles, with 15% in the head and neck muscles. Since intramuscular vascular malformations are found deep inside the muscle, it is challenging during physical examination to arrive at the diagnosis.

Conclusion. Cone-beam computed tomography and ultrasonography can significantly improve the reliability and accuracy of observers in detecting arteriovenous malformations (AVMs). A multimodal approach to diagnosis is feasible for detecting AVMs.

Keywords: arteriovenous malformation, phlebolith, cone-beam computed tomography, ultrasonography, masseter.

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ВНУТРИМЫШЕЧНАЯ АРТЕРИОВЕНОЗНАЯ МАЛЬФОРМАЦИЯ ПРАВОЙ ЖЕВАТЕЛЬНОЙ МЫШЦЫ С ПРИЗНАКОМ «ИНДЮШАЧИЙ ПЛЕТЕНЬ»

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Цель исследования. Проанализировать редкую опухоль, известную как артериовенозная мальформация (АВМ), которая представляет собой состояние, когда артерия и вена напрямую соединены. Чаще всего поражается жевательная мышца, в 5% случаев по данным литературы. Это состояние можно подтвердить с помощью таких лучевых методов диагностики, как ультразвуковое исследование и конусно-лучевая компьютерная томография.

Материалы и методы. Представлен клинический случай 46-летней пациентки, у которой в течение двух лет наблюдалась припухлость в правой щечной области, сопровождавшаяся периодической болью при жевании и увеличением образования в положении лежа на правом боку. Для исключения одонтогенного происхождения была рекомендована ортопантомография, затем – конусно-лучевая компьютерная томография и ультразвуковое исследование.

Результаты. Конусно-лучевая компьютерная томография выявила множественные кальцифицированные узелки в области жевательной мышцы размером примерно 2х2 мм. УЗИ выявило неравномерное мягкотканное образование на нижней границе правой жевательной мышцы, размером примерно 17х9 мм, внутри которого отмечались скопления венозных каналов, а также участки кальцификации.

Обсуждение. Артериовенозные мальформации (АВМ) – редкие опухоли, которые нечасто встречаются в области головы и шеи, и менее 1% сосудистых опухолей обнаруживается в мышцах, причем 15% – в мышцах головы и шеи. Поскольку внутримышечные сосудистые мальформации располагаются глубоко в мышцах, при физикальном обследовании поставить диагноз довольно сложно.

Заключение. Конусно-лучевая компьютерная томография и ультразвуковое исследование значительно повышают надежность и точность диагностики при выявлении артериовенозных мальформаций (АВМ). Для выявления АВМ целесообразно использовать мультимодальный подход к диагностике.

Ключевые слова: артериовенозная мальформация, флеболит, конусно-лучевая компьютерная томография, ультразвуковое исследование, жевательная мышца.

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Vascular malformations were first identified by Wardrop in 1818. Later, the International Society for the Study of Vascular Anomalies classified them into two types: tumors and malformations, as proposed by Mulliken and Glowacki [1]. Arteriovenous malformation (AVM) is a type of malformation that occurs due to errors in vascular development during embryogenesis, causing abnormal channels between arteries and veins. This condition is progressive and recurring and can involve angiogenesis or vasculogenesis. It is characterized by shunting directly from the arteries to veins without passing through a capillary bed, through a fistula or nidus, which are abnormal channels between feeding arteries and draining veins [2]. Hemangiomas, on the other hand, are tumors characterized by the rapid proliferation of endothelial cells in early infancy, followed by involution over time. Other vascular abnormalities are malformations that result from abnormal development of vascular plexuses. Both hemangiomas and vascular malformations are part of the scientific term "vasoformative tumors," which have a normal endothelial cell growth cycle and affect veins, capillaries, or lymphatics. One of the common challenges in treating these rare and deeply located tumors is inaccurate preoperative diagnosis and inappropriate treatment planning [3, 4].

Case report.

A 46-year-old female patient has been experiencing swelling in her right lower back region for two years, accompanied by intermittent pain and enlargement of a mass when lying down on her right side. She had no history of trauma in the previous two years but had undergone root canal treatment two years ago in a private clinic. The patient's family, including her husband and two children, were said to be healthy and not suffering from any disease. Upon general examination, the patient appeared healthy with a normal gait, moderate build, and vital signs within normal limits. During a complete clinical examination, a solitary mass measuring approximately 1X1.5cm in size was found 1cm anterior to the right angle of the mandible (fig. 1 A, B). The swelling was soft, fluctuant, mobile, non-pulsating, and

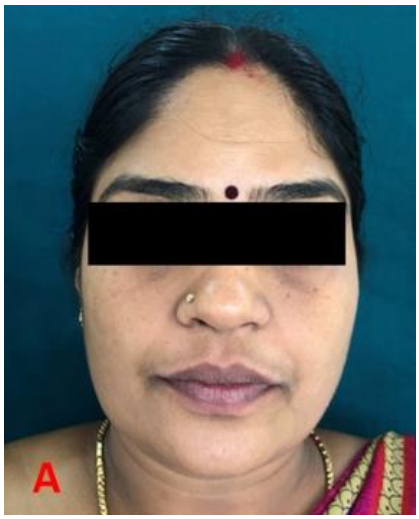
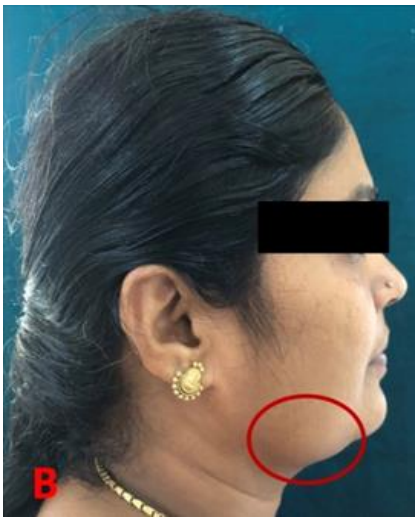


Fig. 1 a (Рис. 1 а)



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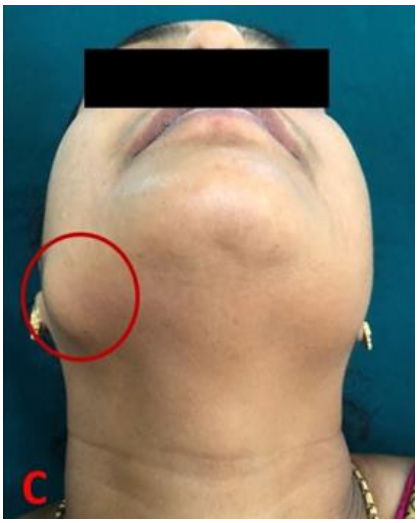


Fig. 1 c (Рис. 1 в)

Fig. 1. Photos.

a – Frontal Profile View of the patient showing mild facial asymmetry on right lower third of the face

b – Right Lateral View of the patient showing solitary swelling measuring about 1*1.5cm anterior to the ramus of the mandible

c – Supine Position – Depicting “Turkey Wattle Sign” in the Right inferior border of mandible Region

Рис. 1. Фотографии.

а – Фронтальный вид пациента, демонстрирующий легкую асимметрию лица в правой нижней трети лица.

б – Вид пациента сбоку справа, с одиночной припухлостью размером около 1х1,5 см в передней части нижней челюсти.

в – Положение лежа – отмечается признак «индюшачий плетень» в области правого нижнего края нижней челюсти.

non-compressible. Tenderness was noted on palpation, and the right submandibular group of lymph nodes was inflamed and tender. The mass was visible and palpable when the patient was clenching or sleeping, which is known as the “turkey wattle sign” (fig. 1 C). Diagnostic imaging was necessary to determine the cause and extent of the abnormalities. An intra-oral periapical radiograph found no bone involvement. According to Orthopantomography, no changes were detected in the right upper and lower tooth region (fig. 2). Further examination

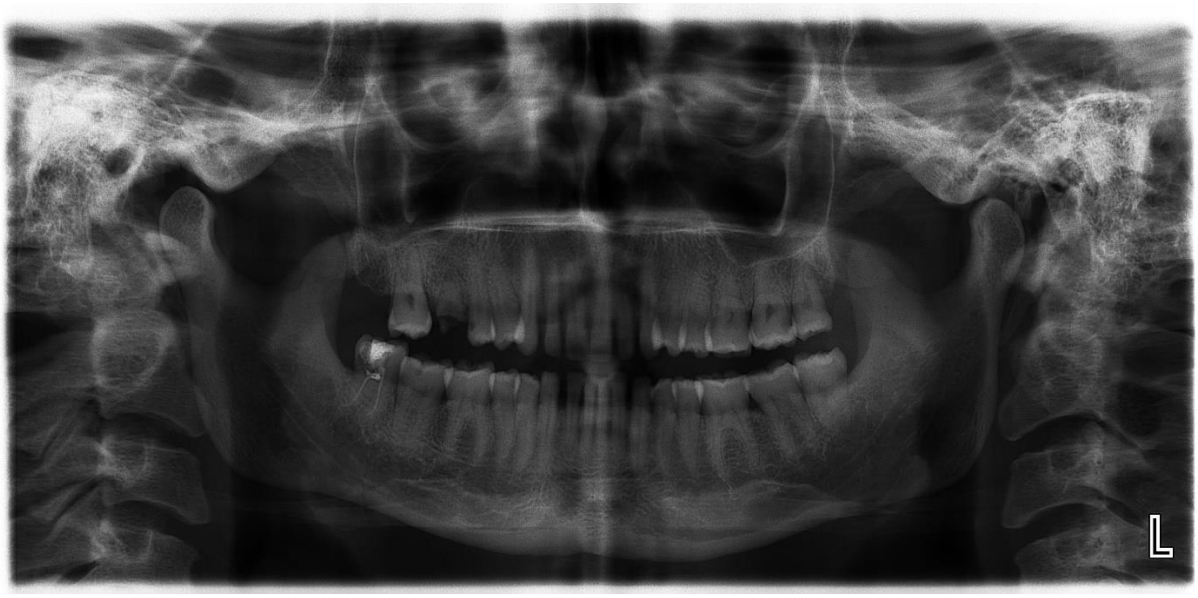


Fig. 2 (Рис. 2)

Fig. 2. Orthopantomogram showing no pathological variations arising from the odontogenic origin.

Рис. 2. Ортопантомограмма, без патологических изменений одонтогенного происхождения.

with cone-beam computed tomography revealed multiple bony nodules involving the masseter region. Three small nodules were noted, each measuring roughly about 2x2mm in size (fig. 3). For further evaluation, the patient underwent USG, which revealed an irregular soft mass in the inferior border of the right masseter muscle. The mass measured roughly about 17X9mm and had clusters of venous channels noted within it, as well as areas of calcification (fig. 4), suggestive of Phleboliths. The impression was an arterial malformation of the right masseter muscle.

Discussion.

Arteriovenous malformations are a medical condition that occurs when an artery and vein connect directly without capillaries. These malformations are typically detected at birth or in early childhood and are often located inside the skull. However, there is another type of AVM called extracranial AVM, which is a more common local aggressive disorder primarily observed during puberty or adolescence [5]. This type of malformation can cause cosmetic and functional disturbances, such as facial asymmetry.

AVM is caused by defects in TGF-beta signaling and a genetic two-hit hypothesis. The development of the mesenchyme primordia is stopped during the undifferentiated capillary network stage, leading to vascular malformation. As differentiation occurs, primitive vessels penetrate deeper into the subcutaneous layer, muscle, or bone tissue. Gradual replacement of the immature plexiform network by mature vascular channels results in a vascular malformation. Angiogenesis is regulated by growth factors such as VEGF, bFGF, TGF-β, and IL-6. This type of vascular tumor is difficult to diagnose due to its cellular nature and firmness. It is called vascular stenosis and has a larger size, is more painful, and is more likely to be diagnosed preoperatively than small vessel tumors [6]. The tumors show more mitotic figures, perineural infiltration, and lymphoid follicles than cavernous types, but less than capillary types. They are only found in 5% of the head and neck region [7].

AVM, or Arteriovenous Malformation, can be triggered by various factors such as trauma, hormonal changes, ischaemic events, and puberty. Skeletal muscle vascular tumors are rare but can occur in the head and neck region, with the masseter muscle being the most commonly affected (accounting for 4.9% of all intramuscular malformations), followed by the sternocleidomastoid and trapezius muscles [8]. While some people may be born with AVM, it

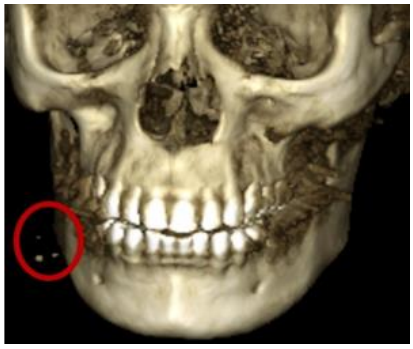
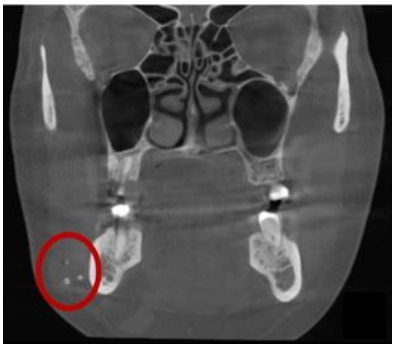


Fig. 3 a (Рис. 3 а)



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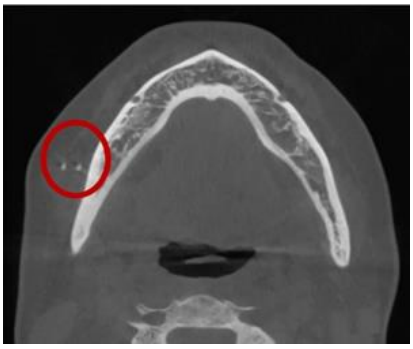

Fig. 3 c (Рис. 3 в)

Fig. 3. Cone-beam computed tomography.

a – section of 3D reconstruction shows 3 specks of calcification measuring roughly about 2x2mm in size 1 cm lateral to the right body of the mandible,

b – coronal Section, c – axial section representing radio-opacity which is 1 cm lateral to the right body of mandible measuring about 2x2mm in size.

Рис. 3. Конусно-лучевая компьютерная томография.

а – на 3D-реконструкции визуализируются 3 кальцината размером примерно 2х2 мм на 1 см латеральнее тела нижней челюсти справа,

б – корональный срез, в – аксиальный срез, визуализируются кальцинаты, которые находятся на 1 см латеральнее тела нижней челюсти справа, размером примерно 2х2 мм.

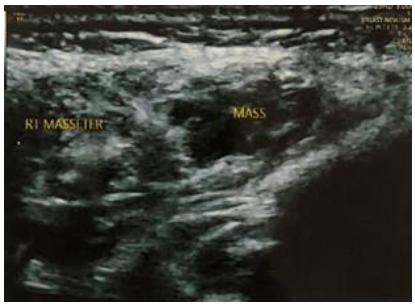

Fig. 4 a (Рис. 4 а)

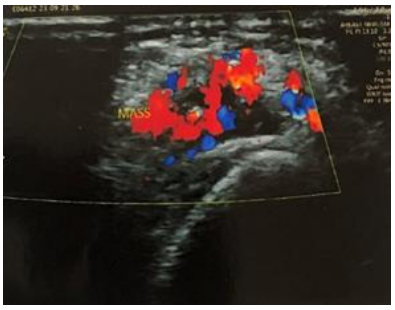

Fig. 4 b (Рис. б)

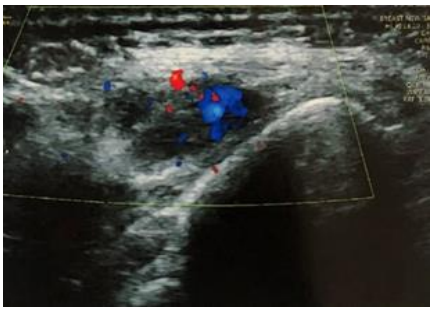

Fig. 4 c (Рис. 4 в)

Fig. 4. Ultrasonography, right masseter region.

Clusters of venous channels noted within the mass measuring 17X9mm with specks of calcification.

Рис. 4. Ультразвуковое исследование, правая жевательная область.

Визуализируются скопления венозных каналов, определяемых внутри образования, размером 17х9 мм с признаками кальцификации.

may not become apparent until childhood. It is most frequently found in the head and neck, followed by the limbs, trunk, and viscera. It can also occur on the anterior two-thirds of the tongue, palate, gingival, and buccal mucosa in the oral cavity.

AVMs can be classified using the Schobinger clinical staging system. Initially, the lesions are latent but develop into a warm pink-bluish skin lesion with a pulsatile thrill in adolescence [9]. Over time, the lesion will expand, leading to dystrophic skin changes, bleeding, ulceration, and tissue necrosis. While the causes of these tumors are still unknown, some evidence suggests that the trophoblast, a placental cell, maybe the origin of vascular malformations [10]. Intramuscular vascular malformations are difficult to detect through physical examination as they are usually located deep within the muscle. The condition is characterized by swelling, pain, and mobile, tense, and pulsatile lesions along the muscle's vertical axis [2, 11].

A pathognomonic symptom of intramasseter and intraparotid hemangiomas is the turkey wattle sign named due to red vascular feature in the male turkey's neck called the "turkey wattle" has the ability to expand in size as it fills with blood. It describes swelling of the lesion accompanied by clenching of the teeth or totally dependent on head posture. Vascular engorgement inside the lesion, which prevents venous return from the head to the superior vena cava, may be the cause of the symptom [16].

Unlike vascular lesions on the skin, these malformations rarely cause skin discoloration and do not grow over time. When they occur in the masseter muscle, they can cause swelling, pain, or facial paralysis [3]. The expansion of the mass can also cause pressure on the nerve, and the lesion becomes more prominent when the Valsalva maneuver or muscle contraction is performed [12]. Tumors can often be mistaken for intramuscular vascular malformations due to their proximity, and they must be distinguished from parotid gland neoplasms. Increased venous pressure can make the tumor more visible. Arteriography is necessary for the detection of vessel communication, while sialography has been replaced by computerized tomography with contrast enhancement, which makes it easier to differentiate between parotid and extra parotid masses [5, 13].

Ultrasound is used to diagnose lesions, while computed tomography (CT) or MRI is better for determining the extent of the lesions. Imaging is crucial in characterizing arteriovenous malformations (AVMs) and planning treatment. Investigations such as Doppler ultrasonography, CT, and MRI can diagnose arteriovenous shunts. Ultrasound and Doppler imaging can reveal high systolic and diastolic flow, arteriovenous shunting, and arterial waveforms. MRI shows AVMs as a mesh of dilated arteries and veins connected by shunts. Magnetic resonance angiography (MRA) shows arterial feeders, shunting volume, and nidus location. Contrast-enhanced CT can provide an accurate structural assessment of arteries, veins, and nidus and also provides flow analysis for preprocedural planning [11]. When dealing with vascular malformations, doctors must examine their flow characteristics to determine the appropriate treatment. Low-flow malformations can be treated with sclerotherapy, laser therapy, or cryosurgery, while high-flow malformations require presurgical embolization followed by aggressive ablative therapy.

The ideal time between embolization and surgical treatment varies based on the goals of embolotherapy. After 2-3 weeks, the benefits of embolization may decrease due to collateral supply and recanalization of vessels. There are several treatment options available, including medical therapy, embolotherapy, sclerotherapy, lasers, cryosurgery, and surgical therapy. The timing of treatment depends on the goals of embolotherapy [14, 15].

Conclusion.

Diagnosis of intramuscular AVMs is challenging and requires a multidisciplinary approach including plastic surgery, vascular surgery, and interventional radiology. In order to improve early diagnosis and patient outcomes, it is imperative that healthcare providers should able to acknowledge the clinical presentations of intramuscular AVMs, particularly the "turkey wattle" sign. Imaging is crucial for diagnosing vascular lesions by identifying the exact location and plane of the lesion. In this case report, Cone beam computed tomography revealed specs of calcification at the site of the right masseter region, leading to further investigation. Ultrasonography helped in specifying the site and size of the lesion and identifying its feeder vessels. To keep an eye out for recurrence and evaluate the effectiveness of the intervention, long-term follow-up is essential. This rare case report adds to the knowledge on intramuscular AVMs by highlighting the significance of a thorough diagnostic imaging and management, when dealing with these unusual vascular diseases.

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