Edson da Silva
(Organizador)

Avanços na Neurologia e na sua Prática Clínica 2

Atena Editora
Ano 2019

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APRESENTAÇÃO

A coleção “Avanços na neurologia e na sua prática clínica” é uma obra com foco principal na discussão científica por intermédio de trabalhos multiprofissionais. Em seus 21 capítulos o volume 2 aborda de forma categorizada e multidisciplinar outros trabalhos de pesquisas, relatos de casos e revisões que transitam nos vários caminhos da formação em saúde à prática clínica com abordagem em neurologia.

A neurologia é uma área em constante evolução. À medida que novas pesquisas e a experiência clínica de diversas especialidades da saúde avançam, novas possibilidades terapeutas surgem ou são aprimoradas, renovando o conhecimento desta especialidade. Assim, o objetivo central desta obra foi apresentar estudos ou relatos vivenciados em diversas instituições de ensino, de pesquisa ou de assistência à saúde. Em todos esses trabalhos observa-se a relação entre a neurologia e a abordagem clínica conduzida por profissionais de diversas áreas, entre elas a medicina, a fisioterapia e a enfermagem, além da pesquisa básica relacionada às ciências biológicas e da saúde.

Temas diversos são apresentados e discutidos nesta obra com a proposta de fundamentar o conhecimento de acadêmicos, profissionais e de todos aqueles que de alguma forma se interessam pela saúde em seus aspectos neurológicos. Compartilhar a evolução de diferentes profissionais e instituições de ensino superior com dados substanciais de diferentes regiões do país é muito enriquecedor no processo de atualização e formação profissional.

Deste modo a obra Avanços na neurologia e na sua prática clínica apresenta alguns progressos fundamentados nos resultados práticos obtidos por pesquisadores e acadêmicos que desenvolveram seus trabalhos que foram integrados a esse e-Book. Espero que as experiências compartilhadas neste volume contribuam para o enriquecimento de novas práticas com olhares multidisciplinares para a neurologia.

Edson da Silva

SUMÁRIO

CAPÍTULO 20

TEMPORAL SUBCUTANEOUS CAVERNOUS HEMANGIOMA: CASE REPORT AND REVIEW

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ABSTRACT: Background: The cavernous hemangioma is considered by the World Health Organization a benign vascular tumor that more frequently involves the skin. However, it can develop in any part of the body.

Case description: An 80-year-old woman experienced subtle headache associated with a progressive growing mass in the temporal region, and right moderate otalgia. During the following 25 days after initial presentation of this tumor, the otalgia and right temporal headache got worse, and the temporal region mass had an important growth in size. She had local pain, which was aggravated by touch. It was unresponsive to conventional analgesia. The patient underwent gross total microsurgical resection of the tumor without complications or presence of any neurological deficits.

Conclusion: The evolution of this type of vascular anomaly as well as the predisposing factors is not yet fully understood. Cavernous hemangioma should be promptly resected when symptomatic or when rapid growth or unacceptable cosmetic appearance is observed.

KEYWORDS: Cavernous Hemangioma, Hemangioma, Neurosurgery.

HEMANGIOMA CAVERNOSO SUBCUTÂNEO DE REGIÃO TEMPORAL: RELATO DE CASO E REVISÃO DE LITERATURA

INTRODUCTION

Hemangiomas are benign vascular tumors that more frequently occur in the superficial skin layers (dermis and epidermis), but may also be present in subcutaneous tissue, muscles, tendons, bones, among others.[11] The progression and malignancy are rarely seen in this tumor.

Cavernous hemangiomas or cavernomas are circumscribed mulberry-shaped vascular anomalies characterized by clusters of enlarged, capillaries with increased permeability. They are mostly located in the brain and then referred to as cerebral cavernous malformations. These lesions occur in 0.1–0.8% of the general population and comprise 10–20% of all vascular malformations in the central nervous syndrome.[7] Cavernous hemangiomas (CH) are more prevalent in women, and the CH of the subcutaneous tissue are more prevalent in childhood. However, their diagnostic in elderly individuals has also been reported, due to growth of preexisting lesion after birth.[3,11,13] Furthermore, only 20% of these vascular tumors are originated at head and neck.

The hemangiomas are classified in capillary and cavernous subtypes according to their etiology.[13] However the cause of hemangiomas remains unclear although previous reports suggest hormonal or congenital genetic factors may be involved.[5] Among these, cerebral cavernous malformations may caused by the loss-of-function mutations in KRIT1, CCM2, or PDCD10. The proteins encoded by these genes are involved in four partially interconnected signaling pathways that control angiogenesis and endothelial permeability. Cerebral cavernous malformations can occur sporadically, or as a familial autosomal dominant disorder with incomplete penetrance and great inter-individual variability.[7]

Clinically, subcutaneous cavernous hemangiomas usually present as a red-blue painless masses with slow growth, despite its capability to bleed, generate local acute pain that may be associated with local hematoma.[12,13] Due to its rarity and diverse clinical presentation, more than 90% of cases are not diagnosed.[4]

Although cavernous hemangiomas do not usually regress with time, the surgical treatment should be considered in situations of subtle growth, acute uncontrollable pain and to restore the physiological contour in situations related to cosmesis. The most common treatment is surgical resection of the tumor, which has a good long-term prognosis.[11]

The goal of this article is to describe the diagnosis and surgical approach in a case of temporal subcutaneous cavernous hemangioma.

CASE REPORT

An 80-year-old female patient came for consultation at emergency unit

complaining of subtle headache associated with a progressive growing mass located at the right tempora and moderate ipsilateral otalgia, within a few hours of its onset.

She referred a poorly controlled hypertension, congestive cardiac insufficiency with 25% ejection fraction and significant cardiomegaly, grade IV chronic renal failure, hypothyroidism, gout disease and hypertriglyceridemia. Furthermore, the patient referred she had underwent previous drainage of subgaleal hematoma of the right temporal region 40 years ago.

She was initially diagnosed with otitis and treated with antibiotics with no improvement after treatment. After ten days, she sought new medical treatment, again. However, due to congestive heart failure presentation, additional investigation of the progressive growing mass at the temporal region of the head was contraindicated.

During the following 25 days, the temporal region mass had an important growth, extending inferiorly from the zygomatic arch to the superior temporal line at the right side of the head. In addition, the patient developed dyspnea and productive cough with worsening of her general condition. The patient was hospitalized and, where she underwent a 10 days treatment for pneumonia and congestive cardiac heart failure.

A neurosurgical team evaluated the growing mass at the temporal region. The patient presented with intense otalgia and right temporal headache with visual analogic scale of pain (8/10) which had worsened with superficial touching, both unresponsive to conventional analgesia by painkillers or anti-inflammatory medications.

She presented an expansive non-pulsatile subcutaneous mass at the temporal region with softened content at superficial palpation, adhered to deep planes and painful to vigorous mobilization (Figure 1). MRI scan showed a heterogeneous subcutaneous mass with predominant hyposignal in T2 weighted images and predominant T1 hypersinal, suggesting methemoglobin content and previous bleeding (Figure 2). This fact complicated the interpretation of the images due to the hematic content that was detected. Hemangiomas classically show T2 weighted images hypersinal. In Time of Flight (TOF) sequences the vascularization of the lesion could not be seen (Figure 2), which is compatible with cavernous hemangioma since these hemangiomas are low flow vascular malformations. The surgical team preferred not to administer gadolinium due to critical renal functional status of the patient.

The patient underwent an uneventful and successful microsurgical resection of the tumor. Considering her clinical status and comorbidities, the surgical team together with the anesthesiology team opted to program for awake surgery with light sedation and local anesthesia. A straight incision was made one centimeter forward to the tragus extending to the superior temporal line at the right side of the head. Intraoperatively an expansive lesion at the subcutaneous tissue was evidenced. It presented irregular contour, yellow-reddish heterogeneous coloration, soft consistency, with a clear cleavage plane with the superficial fascia of the temporal

muscle. After cautious dissection of the lesion, it was resected on en bloc fashion. The pathology confirmed the diagnosis of cavernous hemangioma of the subcutaneous tissue, revealing on macroscopical analyses a well-defined lesion with vascular dilatations. Microscopically, the pathology evidenced thick wall vessels, without layer distinctions associated with fibrosis and the presence of intraluminal thrombus inside some vessels. The vessels were involved by subcutaneous tissue (Figure 3). The patient was discharged from the hospital, six hours after surgery without neurological deficits. Control MRI six months after surgery showed gross total resection of the lesion (Figure 4).

DISCUSSION

Hemangiomas are benign vascular malformations of congenital origin that are characterized by vessels proliferation. The cavernous hemangiomas of the subcutaneous tissue are described as a red-blue spongy mass, formed by great vascular cavernous spaces filled with blood and separated by a small amount of connective tissue.[12,13] Clinical features depend exclusively on its topography, size of the lesion and presence of acute bleeding. Although generally they present as painless tumors, they can cause symptoms if they compress nearby structures or if they bleed.[13]

Hemangioma's etiology is still very discussed in the literature, although the previous trauma history has been suggested as one of the predisposing factors, the majority of authors believe that hemangiomas arise from genetic mutations or hormonal alterations.[7,9]

Subcutaneous hemangiomas might occur both in adults and children, and about 73% of the cases are already present at birth. By the end of the first year of life this estimative reaches 85% of cases. Its onset at older ages is unusual and according to some authors is related to the expansion of an already existing hemangioma at birth. Most hemangiomas often grow up to 15 years of age.[3,11,13]

The diagnosis is made through the CT scan, MRI scan and TOF imaging.[1] Computed tomography is useful to define the shape, size and anatomical tumor relationship, but MRI is the method of choice in defining the vascular nature of the tumor. Certain radiological features seem to suggest the presence of hemangiomas such as high signal intensity on T2 weighted image, the presence of endothelial channels separated by linear fibrous and/or fat tissue in lesion larger than 2 centimeters, and the presence of thrombus areas, fibrosis, deposition of hemosiderin and/or calcification. [4] Therefore, the most important image feature is to make the differential diagnosis of the lesion with malignancies,[9] once they are easily distinguished from other soft tissue tumors by CT scan, MRI scan and arteriography.[4] Gadolinium enhancement

of the lesion as well as vascular enhancement inside and around the tumor would be expected. The main differential diagnoses include neurofibroma, lipoma, dermoid cyst, enlarged lymph nodes, soft tissue sarcomas, myositis ossificans and temporal arteritis.[9] Cavernous hemangiomas may be sporadic or genetic (familial autosomal dominant disorder). Description of an unusual association between two independent hereditary diseases of confirmed genetic origin included multiple familial cerebral cavernous malformations (FCCMs) and Wilson disease. Genetic analysis revealed single nucleotide polymorphisms in genes CCM2 and CCM3, associated with cavernous malformations, and homozygote mutation in the ATP7B gene, responsible for Wilson disease.[6] Dural arteriovenous shunts in the central nervous system and cavernous hemangiomas may be comorbidities.[10] Cavernous hemangioma was sporadic in our patient.

As cavernous hemangiomas do not usually regress, treatment is usually necessary,[11] and surgical resection seems to be the best choice for bleeding or expansive lesions. Other treatment options are radiation therapy which is avoided in children because of their side effects and arterial embolization which is usually not done in isolation.[2,8,13]

CONCLUSIONS

Subcutaneous cavernous hemangioma of the temporal region of the head is a very rare vascular neoplasm. Although the subcutaneous hemangioma is commonly presented as a benign asymptomatic mass, bleeding or compression of adjacent structure with pain may occur. The differential diagnosis with soft tissue tumor or malignancies should be considered when rapid growth or painful presentation are observed. The best imaging options for its diagnosis in the subcutaneous tissue of the head is certainly TOF imaging and MRI with gadolinium.

Although subcutaneous hemangiomas do not metastasize, follow up is necessary because the progression of the lesion may lead to potential risks of bleeding, and mass expansion with the possibility of causing compression of adjacent structures.

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FIGURES

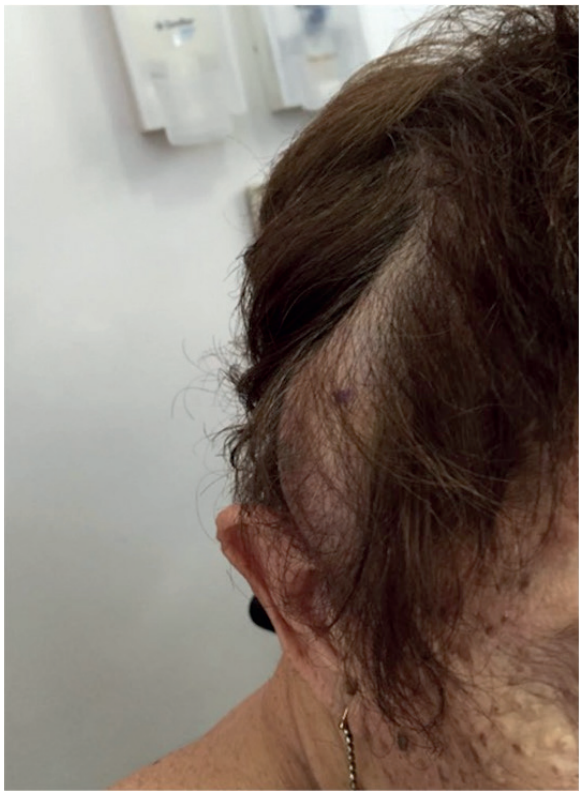

Figure 1. Inspection image during preoperative physical exam: tumor extending inferiorly from the zygomatic arch to the superior temporal line at the right side of the head.

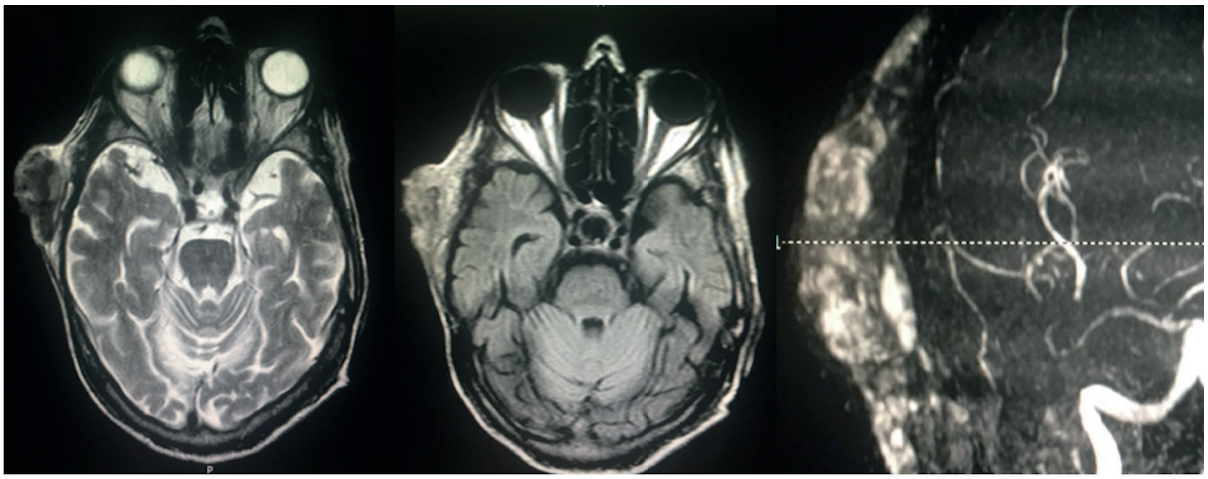

Figure 2. On the left T2 weighted MRI image with predominant hiposinal in the lesion. On the right, T1 weighted MRI image with predominant hypersinal. This presentation suggests previous bleeding. On the right side TOF imaging shows an avascular lesion.

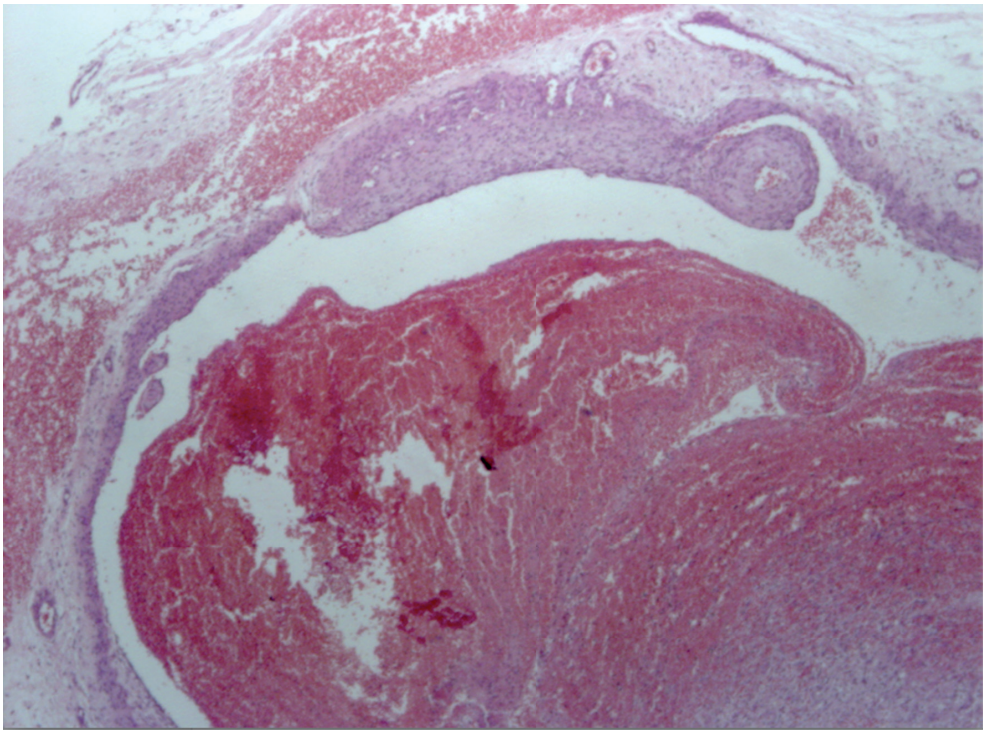

Figure 3. Hemangioma associated with intraluminal thrombus with vessels involved by subcutaneous tissue (hematoxylin and eosin stain, 40x).

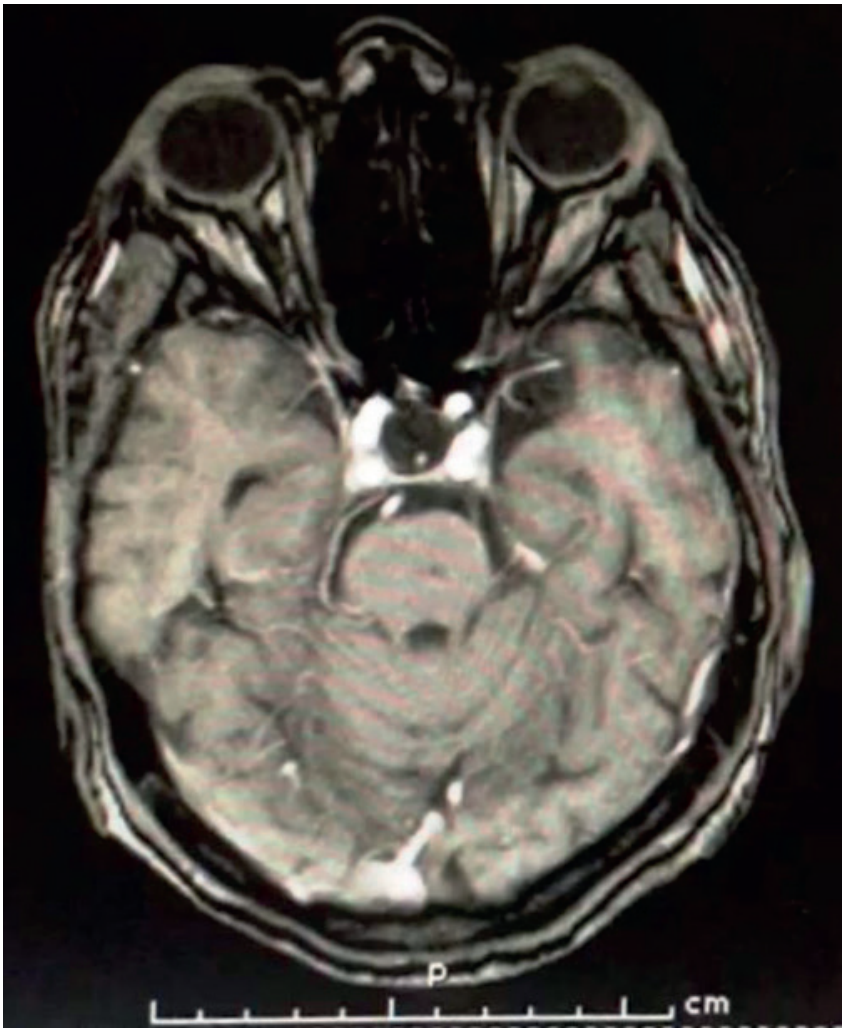

Figure 4. T1 weighted MRI image, axial plane, showing the complete resection of tumor in postoperative.

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