Giant intracranial and extracranial cavernous malformation

Case report

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Massive enlargement of an extracerebral cavernous malformation and extension across tissue planes is very uncommon. The authors present the case of a 49-year-old woman with a giant cavernous malformation in the left frontotemporal area. It progressively enlarged during several decades, extended through the calvaria to the extradural space, and was surgically treated. The lesion may have originated in the soft tissue or the skull. The locations of cavernous malformations in various parts of the body are reviewed and their mechanisms of growth are discussed. Surgical excision is the treatment of choice.

KEY WORDS • cavernous malformation • skull • soft tissue

Vascular malformations are developmental anomalies that occur when embryonic vascular networks fail to differentiate.35,36 Cavernous malformations (CMs), which are also termed cavernous hemangiomas, cavernous angiomas, and cavernomas, are one type of vascular malformation. They are found within the parenchyma of the central nervous system, in intracranial, extraxial locations, in bone, especially the vertebrae and skull, and in soft tissues of the body. Published reports typically deal with solitary lesions involving only one area of the body. Multifocal CMs affecting the same organ system occur to a lesser extent. Eighteen percent of CMs in the brain are multiple, and multiple soft-tissue CMs occur in Maffucci’s syndrome.3,46 More extensive, simultaneous involvement of adjacent tissues is unusual. In this report we present a patient with a giant frontotemporal CM involving the extracranial soft tissues, skull, and extradural space.

Case Report

History. This 49-year-old right-handed woman was born with a red birth mark on her left preauricular area. She had been told it was initially the size of a nickel. It enlarged somewhat during her first pregnancy and then stabilized. At 22 years of age she was admitted to a local hospital with a 6-week history of increasing lesion size and ipsilateral tinnitus. On examination, the skin abnormality measured 3 × 2 cm. It was red with an overlying “papular eruption,” nontender, and had a palpable thrill. At the time of surgery the mass was found to extend through the temporalis muscle into the underlying skull. Vigorous bone bleeding was encountered, which resulted in a loss of 8 U of blood. The removal was described as nearly complete and the surgeon believed the resected tissue represented a “hemangioma with arteriovenous fistula.” The pathological specimen contained only striated muscle with several large vessels. During the ensuing 27 years, the lesion recurred and gradually enlarged.

Examination. At the time of the patient’s initial presentation to our institution, there was a 10-cm mass in her left frontotemporal area, which produced a significant cosmetic deformity (Fig. 1). The patient did not report headache or tinnitus but did notice occasional muscular spasms in the left infraorbital area and cheek. The overlying skin was coursed with several ectatic superficial vessels. The lesion was painless, soft, and pulsatile but had no bruit. She was neurologically intact. A computerized tomography (CT) scan of the head, with and without intravenous contrast medium, demonstrated a large subgaleal mass with two underlying areas of expansion of the squamous temporal and frontal bones, producing small extradural components (Fig. 2). The overlying soft tissues were involved and the lesion extended deep to the zygomatic arch. In the left parietal area, there was an adjacent focus of bone erosion and a larger associated extradural mass that produced midline shift (Fig. 2). An angiogram revealed a vascular lesion with exclusive blood supply.
FIG. 1. Intraoperative photograph. The large mass extends from the frontotemporal area to below the zygoma. Note the ectatic superficial vessels on the inferior portion of the mass and cheek.

from the left external carotid artery (Fig. 3). An attempt at preoperative embolization was unsuccessful because of difficulty in cannulating the feeding vessels.

Operation. The external carotid artery was exposed in the neck to allow for proximal control. The mass was subgaleal and the temporoparietal fascia was reflected, sparing the frontal branch of the facial nerve. The malformation involved the temporalis muscle, and several feeding vessels coursing under the zygoma through this muscle were ligated and divided. The areas of calvarial involvement in the temporal, frontal, and parietal bones were adjacent but not directly contiguous. The extradural component did not involve the dura and separated cleanly from it. All of the apparent malformation was resected including a small rim of surrounding bone. The cranial defects were repaired with methylmethacrylate.

Histological Examination. The specimen consisted of multiple pieces of gray, white, and red soft tissues and bone. Several of the pieces had a focally spongy texture because of the presence of numerous vascular channels. Histologically, most of the thick-walled vascular channels had relatively large lumina with walls composed of endothelial cells surrounded only by connective tissue (Fig. 4). The vascular channels were often closely apposed without other intervening tissue, as is typical of CMs. In some areas, there were foci of intraluminal tufting consistent with intravascular papillary endothelial hyperplasia.

Postoperative Course. Although the resection appeared complete, during the next 2 years the patient developed a limited extracranial recurrence. On postoperative CT scanning, this was seen to involve the masseter muscle. She has declined further surgical intervention, and the lesion has remained stable for the past 4 years.

Discussion

Location of CMs

Within the central nervous system the cavernous form represents 8 to 15% of vascular malformations.\(^\text{16,17}\) They are most commonly found subcortically near the fissure of Rolando or basal ganglia or in the brainstem. They are made up of vascular channels with an endothelial lining, but no internal elastic lamina or smooth-muscle layer, and no intervening parenchyma. The frequent presence of hemosiderin-laden macrophages suggests previous bleeding, and accounts for the characteristic surrounding rim of low signal intensity seen on T1-weighed magnetic resonance images.\(^\text{19}\) They occur equally in males and females and generally present when the patient is in the 3rd and 4th decades of life.\(^\text{42}\)
Giant cavernous malformation

Intracranial, extraaxial CMs are uncommon, making up 0.4 to 2% of all vascular malformations.60 Most reported cases are found in the middle fossa, arising within the cavernous sinus.31,38,22,23,27,28,32,41 They occur predominantly in females (11:1). They have been suggested to represent a true neoplasm rather than a vascular malformation,22,23 but we have included them in the category of vascular malformations because of histological appearance.31,38,33 Other reported sites of origin include the subarachnoid space,20 the tentorium,20,29,86 the convexity dura,13,21,33,43,44,45,53 the cerebellopontine angle,26 Meckel’s cave,8 and other cranial nerves.23,25 Dural CMs found outside the middle fossa have no gender predilection.26 Erosion of adjacent bone is often found with all these lesions, and they are histologically similar to intraparenchymal malformations.51,23 Radiographically, they resemble meningiomas.

Hemangiomas arising in bone account for 0.7% of all osseous tumors. The vertebral column is the most common site of occurrence, followed by the calvaria and the mandible.12 Skull hemangiomas most frequently occur in the parietal and frontal bones and represent 0.2% of all bone tumors and 10% of all primary benign skull tumors.5,12,35,54 the majority are CMs.3,5 They are three times more common in females, with a peak incidence in the 4th decade. They arise in the diploë, expand the outer table, and generally spare the inner table. More extensive involvement of the inner table and extradural space is unusual. Most are solitary, and one-third have perilesional sclerosis.24 On plain skull x-ray films, they are seen as a rounded area of rarefaction that has a honeycomb appearance.5,35,54 A “sunray” pattern of radiating trabeculae is sometimes present on tangential views. Microscopic examination reveals dilated sinusoidal channels that are lined by a single layer of flattened endothelium and interspersed among bony trabeculae.12,35,54

Historically, the nomenclature for soft-tissue vascular lesions was varied and confusing. The modern classification system of Mulliken and Glowacki,10,31 however, arranges these lesions according to their clinical and histological characteristics. In this system, hemangiomas are present at birth in only 33% of patients; the lesions grow rapidly during the 1st year and more than 40% involute during the patient’s childhood. They are three times more common in females. During growth they exhibit endothelial cell proliferation with increased mitotic activity and form syncytial masses with and without lumina. Vascular malformations, on the other hand, are always present at birth, grow in proportion to the child, and never involute. Both sexes are equally affected. They are not hypercellular, and the vascular channels are lined by flat endothelial cells with normal mitotic activity. There are low-flow and high-flow subtypes.

There are other reports of pediatric head and neck lesions that may fall into this vascular malformation category.14 The authors, although they term them cavernous hemangiomas, describe involution that does not occur or is incomplete, and their microscopic appearance consists of thin-walled cavernous vessels lined by endothelium. Cavernous malformations are known to arise in many other parts of the body. They can be found throughout the gastrointestinal tract and make up 5 to 10% of benign tumors of the small intestine. They represent the most common benign tumor of the liver and spleen.23 The cavernous malformation is also the most common primary benign orbital tumor in adults,21 and retinal lesions are seen in neurocutaneous syndromes.3,8,53 Cavernous malformations are occasionally found as intrinsic tumors of peripheral nerves.45 Those in the liver and orbit are more common in females.15,46

Origins of CMs

The tissue from which the CM originated in our patient is uncertain. In the patient’s infancy it was visible on the skin, but involvement of deeper structures is unknown. At the first surgery it was located in the subcutaneous soft tissue and the underlying skull. By the time of the second surgery, the lesion extended from the soft tissue through the skull to the extradural space. The skull appeared to be involved in three adjacent areas, and there was expansion of the diploë as well as bone erosion.

The malformation may have begun in the soft tissue and subsequently invaded structures underneath. Sinonasal and cavernous sinus CMs have been shown to erode adjacent parts of the skull.10,19,22,32 Okada, et al.34 reported a frontal skull cavernous hemangioma with components extending extracranially as well as extradurally. A patient born with a large facial cavernous hemangioma was described by Pásztor, et al.34 This hemangioma was substantially removed in adult life, but the patient developed vision changes 2 years later from hemangiomatous extension to the underlying skull, with formation of an extradural middle fossa and parasellar mass.

The possibility also exists in this case that several structures were involved simultaneously rather than by progression of one into another. In 1928, Dandy4 reported a 1-month-old infant with a large intradural posterior fossa cavernous hemangioma that extended through the dura and skull into the subgaleal space. An adult woman was found to have CMs within four mid-thoracic vertebrae, with separate involvement of the adjacent epidural space and an overlying area of skin.3 A similar case has been reported in which CMs were present within the thoracic spinal cord and in the subcutaneous tissue of the adjacent chest wall.51 Eighteen percent of cerebral CMs are multi-
ple, and the number is much higher in familial cases. Multifocal calvarial CMs have been reported, as have simultaneous but separate lesions in the dura and the brain.

Growth of CMs

The malformation affecting our patient underwent substantial growth over the course of several decades. Although made up of nonneoplastic embryonic vascular pathways, CMs are not always static. Cerebral CMs are believed to grow by recurrent internal microhemorrhages and organization. The extraxial CMs generally do not exhibit evidence of prior hemorrhage. These lesions found within the cavernous sinus and dura, as well as those in the skull, enlarge by other means. Their growth may be the result of capillary budding, vascular ectasia, or vascular thrombosis and organization. Hormonal factors may also be involved in the growth of these lesions. Expansion of a very large CM involving the soft tissues, skull, and extradural space. Cavernous malformations can progress from one tissue into another or arise in adjacent tissues simultaneously. It enlarged during a pregnancy and increased progressively following subtotal removal. Its growth was thus related both to hormonal effects and to changes resulting from the first surgical procedure, such as alterations in its vascularity or collateral supply. The patient was successfully treated with a nearly complete removal and her condition has remained stable over several years. As the present case demonstrates, extracerebral CMs may undergo significant growth and cross tissue planes. They may cause cosmetic deformity and produce mass effect on the brain. Surgical excision is the treatment of choice.

Treatment Options

Cerebral CMs present with seizures, focal neurological deficits, headaches, or hemorrhage, and the primary treatment for accessible symptomatic lesions is surgical resection. cavernous malformations in an intracranial, extraxial location generally present with mass effect. Their operative characteristics are similar to those of meningioma. They are attached to the inner surface of the dura and separate cleanly from the underlying brain. Cavernous malformations arising in the cavernous sinus are not so straightforward. Patients with these highly vascular lesions had a perioperative mortality rate of 38% prior to 1984. The close association with neural and vascular structures in this area increases the complexity of removal. Preoperative embolization and the application of modern skull base techniques have improved the success of treatment. Both preoperative and postoperative radiation therapy have also been used to reduce vascularity and shrink these lesions. For CMs confined to the calvaria, en bloc resection including a small margin of the surrounding normal bone is generally curative.

Surgical resection is recommended for soft-tissue vascular malformations of the head and neck that produce significant cosmetic deformity, ulcerate with infection or bleeding, or affect function such as obstructing vision, oral intake, or breathing. Intraoperative blood loss is also a problem with these lesions, especially because they frequently affect the pediatric population. Preoperative embolization can be helpful in this regard. Injection of sclerosing agents has been useful in the nonoperative management of some vascular malformations. Radiation therapy was used in the past as a form of conservative treatment, but it is now infrequently prescribed because the amount of radiation needed to obliterate the lesion produces a significant rate of complications.

Conclusions

It is difficult to be certain of the tissue of origin of this very large CM involving the soft tissues, skull, and extradural space. Cavernous malformations can progress from one tissue into another or arise in adjacent tissues simultaneously. It enlarged during a pregnancy and increased progressively following subtotal removal. Its growth was thus related both to hormonal effects and to changes resulting from the first surgical procedure, such as alterations in its vascularity or collateral supply. The patient was successfully treated with a nearly complete removal and her condition has remained stable over several years. As the present case demonstrates, extracerebral CMs may undergo significant growth and cross tissue planes. They may cause cosmetic deformity and produce mass effect on the brain. Surgical excision is the treatment of choice.

References

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