Resection of a rare giant chondromesenchymal hamartoma of the scalp with intracranial blood supply in an adult woman: illustrative case

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BACKGROUND Nasal chondromesenchymal hamartomas (NCMHs) are benign, slow-growing lesions formed by mesenchymal and cartilaginous components. They occur predominantly in male infants at the nasopharynx and orbit. Rare cases have been reported in adults. Ectopic NCMHs occurring in other head regions without the typical nasopharyngeal or orbital involvement have not been previously described.

OBSERVATIONS The authors presented the case of a 40-year-old woman with a giant mass in the left frontoparietal region that started to enlarge progressively after the patient’s first pregnancy at the age of 21 years. The tumor caused intense headaches, nausea, vomiting, asthenia, and syncope.

On admission, the neurological examination revealed no abnormalities. Brain magnetic resonance imaging showed a solid homogeneous tumor without intraaxial involvement extending inferiorly to the left zygomatic arch, with a significant mass effect on the adjacent bones but no infiltration. Remarkably, digital subtraction angiography demonstrated that the tumor received blood supply from superficial as well as intracranial branches of the left vertebral artery. After tumor resection, histopathological analysis revealed characteristics indistinguishable from an NCMH.

LESSONS The authors described a rare NCMH of the scalp with intracranial blood supply in an adult patient. A case with similar characteristics had not been reported before.

https://thejns.org/doi/abs/10.3171/CASE22249

KEYWORDS hamartoma; chondromesenchymal hamartoma; scalp tumors; neurosurgery

Hamartomas are tumor-like lesions formed by an aberrant mixture of mesenchymal tissue and other components of the organ in which they occur as a result of abnormalities in their quality, organization, and differentiation. These lesions are benign and often show a slow enlargement. Head cartilaginous hamartomas have increased in frequency despite being considered rare. Indeed, lesions such as the chondromesenchymal hamartoma now figure in the World Health Organization classification of head and neck tumors along with osteochondroma, chondroma, chondroblastoma, and chondromyxoid fibroma, among other cartilaginous tumors.

Chondromesenchymal hamartomas are rare, benign, slow-growing, and locally destructive lesions that contain mixed mesenchymal and cartilaginous components. These tumors occur predominantly in male infants, commonly at the paranasal sinus and nasopharynx. Thus, they are named nasal chondromesenchymal hamartomas (NCMHs). Importantly, these lesions can extend to the orbital cavity and intracranially through the ethmoid cribiform plate. Morphologically, these lesions are analogous to other mesenchymal hamartomas and consist of islands of chondroid tissue. A few NCMHs affecting the head have been reported in adults, most of which were small with typical nasopharyngeal, orbital, or intracranial involvement.

Here, we report the case of a woman in her 40s presenting with a giant mass in the left temporal region of the head. After resection and histopathological analysis, the characteristics of the lesion were indistinguishable from an NCMH. Strikingly, the tumor’s location was extraxial with no nasal, orbital, or intraxial involvement. However,
the lesion’s vascularization depended on extra- and intracranial branches of the left vertebral artery. Finally, the tumor cells expressed progesterone receptors. To the best of our knowledge, a case with similar characteristics has not been reported before. Hence, we provide clinicopathological insights regarding this extremely rare tumor relevant to pathologists, clinicians, and surgeons.

Illustrative Case

A 43-year-old woman attended our center after an episode of syncope preceded by a recent history of anorexia and fluctuations in level of consciousness. Since 1995, 6 months after giving birth to her first child at the age of 21, she experienced a gradual and slow increase in volume in the left temporal region, which caused a pulsatile and persistent intense headache that sometimes woke her up at night. Over time, the headache was accompanied by nausea, vomiting, phonophobia, and asthenia. The rest of her family and medical history was not relevant.

On admission, the patient was oriented to person, time, and space. His vital signs and anthropometrics were within normal ranges. The physical examination revealed an evident mass in the left frontotemporal region of the scalp that extended to the zygomatic arch, measuring 18 × 17 cm in horizontal and vertical diameters, respectively. The neurological assessment showed a Glasgow Coma Scale score of 15 with no cranial nerve abnormalities. The ophthalmological evaluation did not reveal significant changes. No alterations in muscle strength, deep tendon, and plantar reflexes were observed. The Romberg and finger-to-nose tests were normal, and meningeal signs were absent.

Initial blood tests were within normal ranges. A brain magnetic resonance imaging study revealed an extraaxial tumor placed over the external cortical layer of the left temporal and parietal bones isointense in T1, with a significant volume effect but without bony invasion or calcifications (Fig. 1A and B). The tumor also extended inferiorly to the level of the maxilla, passing under the zygomatic arch. However, the lesion did not involve intracranial structures and had no relationship with any cranial cavity. Interestingly, digital subtraction angiography of the brain demonstrated that the lesion was irrigated by multiple superficial branches of the left vertebral artery as well as by some intracranial ramifications from the vertebral territory that crossed through the skull bone to get into the tumor (Fig. 1C).

Tumor excision was performed without complications. During surgery, we confirmed that the lesion was completely extracranial but tightly attached to the left temporal muscle, causing erosion to the adjacent bones, including the left zygomatic arch. Notably, the inferior parts of the lesion had bony insertions but did not penetrate the maxillary sinus, nasal cavity, and orbit (Fig. 2A). A solid, homogeneous, and highly vascularized tumor of approximately 300 g was resected (Fig. 2B). We removed part of the scalp flap to improve the aesthetic outcomes, preserving the natural distribution of hair-bearing areas and hair insertion lines. The histopathological analysis of the excised mass showed a mixed composition with muscular tissue, adipose tissue, and multiple hyaline cartilage islands containing scarce spindle cells, consistent with an NCMH (Fig. 2C). The immunohistochemical analysis showed S100 expression in the muscle and adipose tissue and a very low proliferative

![Fig. 1. Preoperative images of the tumor. A: Transverse T1-weighted magnetic resonance imaging of the head shows a giant extracranial homogeneous mass with rich vasculature compressing the left frontal and parietal bones. B: The tumor extended inferiorly, passing under the zygomatic arch to the level of the maxilla, sparing the left orbit and maxillary sinus. C: Digital subtraction angiography demonstrated that the left vertebral artery provided the main irrigation of the tumor through multiple superficial muscular branches (white arrows) but also by some intracranial ramifications crossing the skull bones (black arrow).]
activity, according to Ki-67. We also evaluated the expression of sex hormone receptors. Interestingly, the tumor cells showed nuclear expression of the progesterone receptor but were negative for estrogen receptors. The patient was discharged because of clinical improvement, and she reported high satisfaction with the aesthetic outcomes of the procedure (Fig. 3).

**FIG. 2.** Resection and histopathological analysis of the tumor. **A:** Resection of a giant extracranial tumor of the skull. (1) Positioning of the patient and a left frontotemporoparietal incision with midline extension was marked. (2) Surgical asepsis and placement of sterile fields. (3) Skin incision and soft tissue dissection to the bone plane, having the inferior edge of the lesion as the limit for the dissection. (4) The skin and muscle flap are lifted. (5 and 6) The subperiosteal dissection is continued, separating the tumor from the bone until exposing its inferior insertion in the zygoma. (7) Complete tumoral resection. (8) Surgical wound closure by planes. **B:** Macroscopic analysis showing an ovoid mass of brown rugous appearance and rich vasculature on the surface. The medial aspect of the tumor displayed multiple fascicles of connective tissue and vascular spaces. **C:** Histopathological analysis of the resected tumor showing a mixed chondromesenchymal, adipose, and muscle composition (first and second left-upper panels). Several vessels can be observed within the chondromesenchymal matrix (third-from-left upper panel), containing scarce spindle cells without atypia and mitosis (right upper panel). The tumor was negative for Ki-67, whereas S100 could be detected in some muscle fascicles and adipose tissue by immunohistochemistry. Tumor mesenchymal cells were negative for estrogen receptors but showed nuclear reactivity to progesterone receptors.
vessels, these atypical vascularization of the tumor and involvement of intracranial tumors despite no evident intracranial tumor extension. Together with the feature of the case was that the patient presented neurological symptoms. Accordingly, a remarkable intracranial vascular system. However, in most instances, such masses depend on an arteriovenous malformation. Notably, despite the complexity and dimensions of the tumor, a gross total excision was achieved without major vascular complications, thus preventing future relapses. The aesthetic outcomes were also commendable, impacting the patient’s quality of life significantly.

The rarity of the case raises several questions regarding the etiology of the tumor. Hamartomas, particularly NCMHs, are considered congenital lesions because they are predominantly observed during early childhood, some even associated with genetic syndromes. Other hamartomas of the scalp with a possible congenital origin have also been reported before. Nonetheless, they were small tumors, some with a clear epithelial origin, whereas others contained tissue elements derived from neural crest cells such as bone, cartilage, muscle, nerve, ganglion, and even meningothelial tissue. Also, these lesions predominated in children.

In contrast, a recent report of a maxillary sinus NCMH in an elderly patient with persistent sinusitis indicates that hamartomas could emerge spontaneously in response to chronic insults. Interestingly, the patient’s tumor started to grow until her first gestation, suggesting an acquired origin perhaps promoted by the effects of sex hormone. In fact, 90% of pulmonary hamartomas contain mesenchymal cells that express receptors for estrogens and progesterone, indicating that sex hormones participate in the development and growth of these tumors. Here, we show that our patient’s tumor expressed progesterone receptors, a finding not previously reported in NCMH of the head. However, we cannot rule out the possibility that the patient’s tumor was present since birth before being apparent. Furthermore, the unusual intracranial vascular supply of the lesion could have originated during fetal life. Therefore, in our case, it is plausible that an inborn tissue defect of very slow growth remained untouched during childhood and adolescence until a second hit by sex hormones provoked rapid enlargement.

In sum, we described a rare NCMH of the scalp with intracranial blood supply in an adult patient. To the best of our knowledge, a case with similar characteristics has not been reported before.

Discussion

Observations

We described an unusual head tumor that, to our knowledge, is the first of its class reported in the literature. The case is unique for the following reasons: (1) this is a cartilaginous hamartoma of the scalp with tissue components indistinguishable from an NCMH; (2) the lesion showed no direct connection to the nasopharynx or orbit or intracranial involvement; (3) the mass received blood supply from extracranial and intracranial arterial branches from the posterior cerebral circulation; (4) the tumor occurred in an adult woman with no apparent history of tumor enlargement during childhood but progressive tumor growth since her first pregnancy; and (5) the size of the tumor exceeded what has been found with most NCMHs. Thus, this report could provide insights for clinicians, pathologists, and surgeons. Accordingly, a remarkable feature of the case was that the patient presented neurological symptoms despite no evident intracranial tumor extension. Together with the atypical vascularization of the tumor and involvement of intracranial vessels, these findings required management by a neurosurgical team. Remarkably, other scalp masses have been found connected to the intracranial vascular system. However, in most instances, such masses depend on an arteriovenous malformation. Notably, despite the complexity and dimensions of the tumor, a gross total excision was achieved without major vascular complications, thus preventing future relapses. The aesthetic outcomes were also commendable, impacting the patient’s quality of life significantly.

Lessons

Scalp masses are frequently encountered in daily neurosurgical practice. They originate from a broad spectrum of different disorders and are usually managed by observation or excision. NCMHs are rare head tumors of infants that involve the nasopharynx, orbit, and intracranial spaces, most of them with a possible congenital origin. No reports of hamartomas of the scalp with histopathological characteristics of NCMH have been described before in the literature. Furthermore, little literature exists on the clinical characteristics and origin of NCMH in adults. Rarely, scalp masses receive intracranial blood supply, unless they depend on a congenital vascular malformation or an intracranial tumor with bone infiltration. The case presented here illustrates a well-differentiated extracranial tumor-like lesion with intracranial blood supply, a finding rarely observed in practice. Nonetheless, it is an essential factor to be considered while managing this kind of tumor to prevent vascular complications.

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Disclosure
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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Conception and design: Guadarrama-Ortiz, Capi-Casillas, Román-Villagómez, Almendárez-Sánchez, Choreño-Parra. Acquisition of data: Guadarrama-Ortiz, Capi-Casillas, Román-Villagómez, Alcocer-Villanueva, Choreño-Parra. Analysis and interpretation of data: Guadarrama-Ortiz, Choreño-García, Román-Villagómez, Alcocer-Villanueva, Almendárez-Sánchez. Drafting the article: Guadarrama-Ortiz, Choreño-Parra. Critically revising the article: Guadarrama-Ortiz, Choreño-Parra. Reviewed submitted version of manuscript: Guadarrama-Ortiz, Capi-Casillas, Román-Villagómez, Alcocer-Villanueva, Almendárez-Sánchez, Choreño-Parra. Approved the final version of the manuscript on behalf of all authors: Guadarrama-Ortiz. Study supervision: Guadarrama-Ortiz, Choreño-Parra.

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