

# A pediatric posterior neck venous malformation with an endocranial extension

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## Abstract

Venous malformations are frequently localized in the head and neck region. However, a cervical localization with an endocranial extension is rather a very uncommon occurrence. We present a case of a 4-year-old child who presented with a large posterior cervical mass evolving for a year, firm and painful at palpation. Imaging was required, revealing a posterior cervical mass with an extension to adjacent structures, a destruction of the occipital bone and an endocranial extension. A macrobiopsy of the mass showed numerous irregular vessels. A surgical treatment was performed due to the extension of the mass, the esthetic prejudice it caused and the uncertain diagnosis. Venous malformation diagnosis was confirmed by a histological examination of the resected piece. Surgical management was not associated with the mass recurrence in our case. Here, we aim at identifying the clinical and radiological features of venous malformations, and at describing the different therapeutic features of this condition.

## Keywords

Vascular abnormalities, venous malformation, neck

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## Introduction

Venous malformations (VMs) are low flow vascular malformations based on the International Society for the Study of Vascular Anomalies (ISSVA) classification of vascular anomalies.<sup>1</sup> They are caused by an error in the venous morphogenesis, leading to the development of a dysfunctional and dilated venous network.<sup>2,3</sup> They are predominately located in the head and neck region. They are present at birth. However, they can remain asymptomatic until a late age.<sup>3</sup> Diagnosis of VMs is mainly clinical. Magnetic resonance imaging (MRI) or computed tomography (CT) can be necessary in large VMs or when a complication is suspected. VMs can result in life-threatening complications.<sup>2</sup> Treatment of VMs is either conservative, based mainly on sclerotherapy or surgical. Recurrences are frequently encountered especially in extensive forms.<sup>2,4</sup>

## Case report

A 4-year-old female was referred to our department with an occipital mass evolving for more than 1 year progressively increasing in size associated with a delayed growth. Physical

examination revealed an ecchymotic left posterior cervical mass, firm and painful at palpation, extending to the sub-mandibular and parotid region, filling the retro-auricular groove displacing the pinna laterally. CT scan revealed a posterior cervical mass measuring 63 mm × 75 mm × 50 mm reaching the superficial cervical fascia, the parotid gland and the sternocleidomastoid muscle with an endocranial extension and a destruction of the occipital bone and the retro-auricular temporal bone and with punctiform calcifications within the lesion (Figure 1). MRI confirmed the presence of an irregular and heterogeneous mass with well-defined limits, centered on the occipital bone with a high signal on T2- and T1-weighted images with an endocranial extension. It

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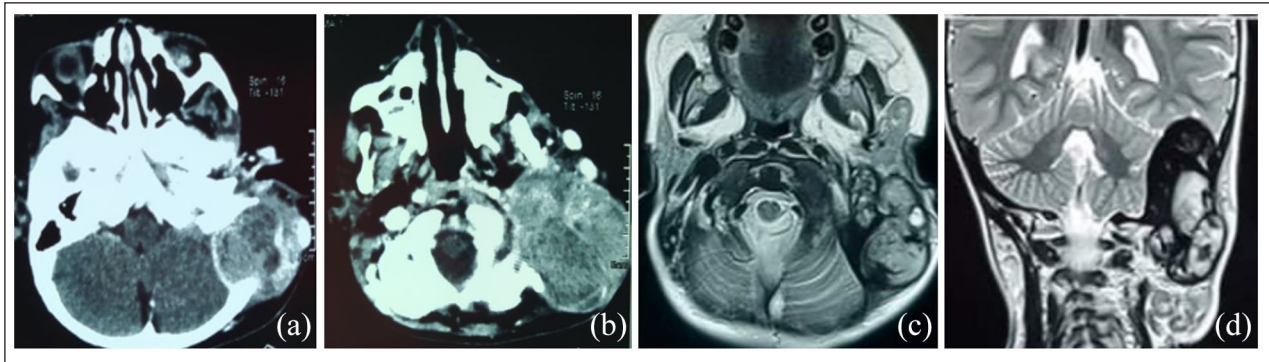
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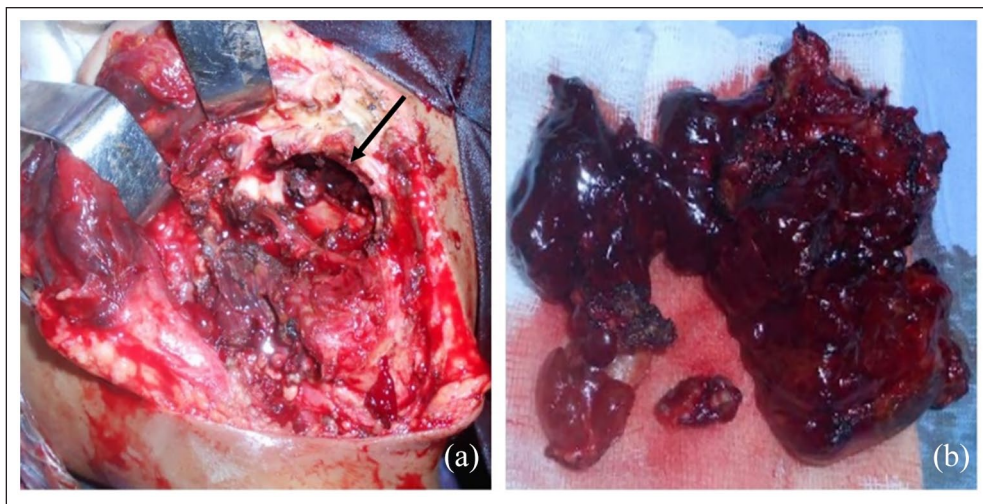
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**Figure 1.** (a and b) Axial cervical contrast-enhanced CT scan showing a left heterogeneous posterior cervical mass, with a massive destruction of the occipital bone without a periosteal reaction, associated to an endocranial extension repressing the left cerebellum without invading it. (c and d) Cervical MRI, axial and frontal T2-weighted images showing a hyper-intense irregular and heterogeneous mass centered on the occipital bone, surrounded by a hypo-intense halo with a loco-regional and endocranial extension repressing the left hemi cerebellum.



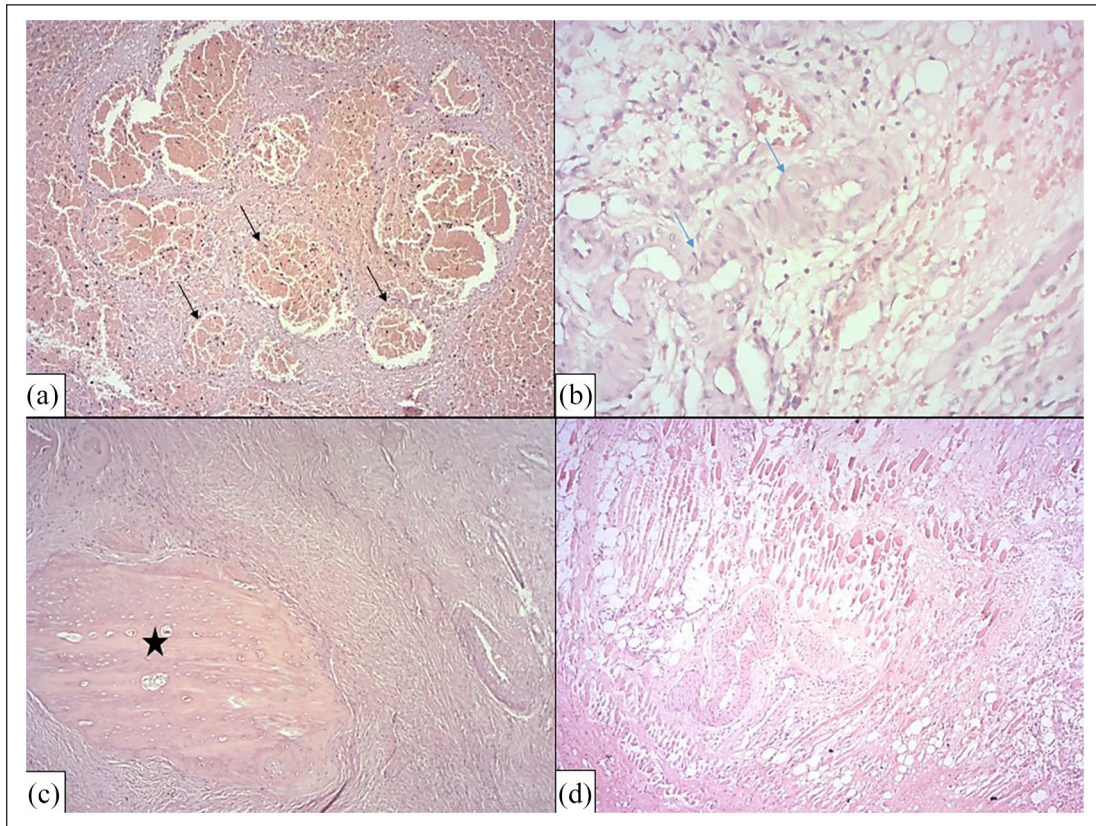
**Figure 2.** (a) Perioperative image after the mass resection, showing the occipital bone lesion (black arrow). (b) Macroscopic view of the resected cervical mass, showing a hemorrhagic mass 8 cm × 6 cm × 3 cm.

showed a significant enhancement, predominantly on the peripheral parts (Figure 1). Imagery was suggestive of a primitive bony lesion or a sarcoma. We performed a macrobiopsy under general anesthesia that only showed the presence of numerous irregular vessels.

Surgical treatment was performed and consisted of an excisional resection of the mass. Perioperatively, we noted the presence of a cervical mass that constituted predominantly of multiple hematomas that was completely resected by our team. The endocranial part of the mass was then totally resected. We noted the presence of an important lesion at the occipital bone reaching and repressing the left cerebellum, yet the dura mater was intact (Figure 2). The surgical procedure lasted for 120 min. No perioperative complications were noted, and the estimated perioperative blood loss was 100 mL. Postoperatively, no complications were recorded and the total length of hospitalization in our

department was 5 days. The diagnosis was made on the basis of the macroscopic anatomopathological examination of the mass which indicated the presence of multiple capillary and venous vessels with varying sizes and proportions haphazardly arranged, dilated capillaries, and thick muscle walls veins. We did not record the presence of arterial vessels with their various parietal tunics and elastic laminae (Figure 3). Although the histological appearance was not suspicious of malignancy, an immunohistochemical study was performed on the previous biopsy as well as on the excision specimen; it did not show a clone of tumor cells.

Control after 5 months revealed a total regression of the cervical mass with a partial regression of the cutaneous ecchymosis. A CT scan performed 3 months postoperatively did not demonstrate any evidence of a persistent or a recurrent disease.



**Figure 3.** The lesion consists of multiple vessels of varying sizes and proportions, haphazardly arranged: dilated capillaries (black arrows) ((a): H&E 50 $\times$ ) and thick muscle walls veins (blue arrows) with regular endothelium ((b) H&E 200 $\times$ ). Vessels dissociate occipital bone ((c) H&E 100 $\times$ ) and extend to subcutaneous fat and skeletal muscle ((d) H&E 100 $\times$ ).

## Discussion

VMs are the most frequent vascular malformations.<sup>2</sup> They are caused by an error in the embryogenesis resulting in abnormal veins with thin walls as well as deficient and dilated smooth muscles. This causes a progressively expanding and dysfunctional venous network leading to clotting and flow stagnation.<sup>2,3</sup> VMs occur sporadically in 90% of cases, but familiar forms were also reported.<sup>2</sup>

VMs occur in the head and neck region in between 40% and 50% of cases affecting most commonly the oral cavity, the neck, the masseter, buccinators and tongue muscles and the airway.<sup>5-7</sup> Craniofacial skeleton involvement is dominated by facial bones while cranial bones are not commonly affected.<sup>2</sup> Extensive forms spread to adjacent structures including parotid gland, cervicofacial musculature, oral cavity and respiratory tract.<sup>2,5</sup> Our patient presents a rare case of a cervical VMs associated with an occipital bone destruction and an endocranial extension. To the best of our knowledge, this could be the first reported case of a venous malformation that was complicated by a destruction of the occipital bone and an endocranial extension.

VMs are present at birth. However, unlike other arteriovenous malformations, they do not regress spontaneously

and rather expand compressing and infiltrating surrounding structures.<sup>2,3,5</sup> VMs, especially deep forms, may remain asymptomatic until they grow enough to cause a local deformity or a complication.<sup>3</sup> In our case, the VM only manifested at the age of 3 years old.

VMs appear as a single lesion that is either localized or diffused.<sup>3,8</sup> Symptoms depend on the size, location and local expansion of the mass.<sup>7</sup> Local swelling and pain occur due to thrombosis and phleboliths formations within the mass.<sup>9</sup> In our patient, VM manifested by a painful and ecchymotic posterior cervical mass rapidly expanding in size. We attributed this abrupt size growth to an infection of the mass and to the trauma that could have been caused by the macrobiopsy. Hormonal changes, hemorrhage, valsalva maneuvers, infections and trauma were reported as the factors that can possibly lead to a change in VMs size. Physical examination in VMs reveals a soft, compressible and non-pulsatile mass with engorgement at the valsalva maneuver and undefined boundaries.<sup>2,6,7</sup> Skin can be either normal or purple due to the dermis involvement. In some cases, palpation reveals local phleboliths.<sup>10</sup> Large VMs cause an esthetic prejudice, bleeding and local complications due to the local expansion and the compression of adjacent structures.<sup>6</sup>

VMs can be associated with coagulation abnormalities such as the elevation of plasma D-dimers.<sup>5,11</sup> Systematic thrombosis is not commonly reported.<sup>8</sup>

Diagnosis is usually established based on history and examination especially in superficial VMs, whereas imaging can be necessary in deep forms. Ultrasound can prove the presence of phleboliths within the lesion.<sup>12,13</sup> CT is indicated when a bony destruction is suspected revealing bony lesions and the presence of phleboliths within the lesion which are characteristic of VMs.<sup>3</sup> MRI is indicated in large and deep lesions in order to confirm diagnosis, define the extension toward adjacent structures and make the difference between VMs and arteriovenous malformations.<sup>2</sup> VMs manifest as hyper-intense and hypo-intense lesions on T2- and T1-weighted images, respectively.<sup>3,8</sup> As physical examination was not sufficiently contributive in our patient, MRI and CT were performed revealing a bony destruction with an endocranial extension.

Histopathological examination is rarely necessary to confirm diagnosis. It shows irregular venous-type channels lined by irregular layers of smooth muscles, focally absent. Vessels' lumina frequently contain thrombi or phleboliths.<sup>14</sup> In our case, the diagnosis was confirmed based on the histopathological examination of the resected piece.

Treatment of VMs depends on the localization, extension, associated complications, esthetic prejudice, team experience and on technical availability.<sup>6,7</sup> It aims at curing VMs definitively and reducing symptoms. VMs management can be conservative using either sclerotherapy or laser.<sup>4,6</sup> Sclerotherapy is the mainstream treatment of VMs; it is used alone in small forms, or preceding surgical resection. It also helps to treat pain.<sup>7</sup> However, it is associated with a risk of recurrence mainly related to the VM size and extension. Sclerotherapy can cause allergic reactions and local necrosis.<sup>6</sup>

Surgical management is mainly indicated in deep small and well-limited VMs and in the case of esthetic and functional complications. It can also be considered in large VMs with well-definite limits are MRI.<sup>5</sup> It can be proceeded by sclerotherapy to reduce the size of extensive and deep VMs and reduce perioperative bleeding.<sup>2</sup> Surgical treatment can result in an important perioperative bleeding requiring multiple transfusions.<sup>6,15</sup> However, complete resection is rarely achievable and can be associated with a higher risk of disfigurement and injury to surrounding structures.<sup>5</sup>

Recurrences are possible in the case of incomplete resection, requiring repeated procedures. In the case of our patient, due to diagnosis uncertainty, the size of the mass, its extension, the esthetic prejudice and its well-limited borders, we opted for a surgical treatment. It was efficient without recurrence after a follow-up period of 5 months.

## Conclusion

VMs occurring in the head and neck region can be associated with an extension toward the adjacent structures. However, a

cervical localization of a VM with an endocranial extension is a very rare occurrence. Clinical presentation of VMs depends on the depth, the size and the local extension of the mass. Diagnosis is mainly established based on history and clinical examination while imaging can be necessary in deep forms. Treatment tends to cure VMs and to reduce symptoms; it can be either conservative based on sclerotherapy or surgical. Treatment modalities are related to the size, the extension to the surrounding structures, the esthetic prejudice and the technical availability. Due to its rarity, the treatment of a VM with an endocranial extension is not well defined in the literature. Surgical resection can be efficient in large, well-limited and extensive forms.

## Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

## Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.


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## Informed consent

Written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article.

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