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Case Report

Giant cervical teratoma and cerebral arteriovenous malformation: Coincidence or association?

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Abstract. Giant congenital cervical teratomas and cerebral arteriovenous malformations (AVMs) are both rare and life threatening. The co-occurrence of these conditions has not previously been described, however new theories regarding the etiology of cerebral AVMs include a "response to injury". We describe a patient with a vascular giant cystic cervical teratoma diagnosed in fetal life and a cerebral hemorrhage from an ipsilateral AVM later in childhood. We postulate that the development of the AVM may have been related to vascular steal from the tumor in utero. We raise awareness of the potential association of vascular neck masses with intracranial AVMs.

Keywords: Fetal MRI, EXIT procedure, fetal neck mass, cystic teratoma, intracranial AVM

1. Introduction

Giant cervical teratomas are rare and increasingly diagnosed antenatally. Their incidence is consistently reported as between one in 20,000 and one in 40,000 live births [1]. They often cause antenatal esophageal and airway obstruction, which can lead to polyhydramnios. In this article, we report a child with an antenatally diagnosed tumor successfully resected in the neonatal period, who presented at the age of 10 yr with an intracranial hemorrhage secondary to an underlying ipsilateral arteriovenous malformation (AVM).

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2. Case report

The patient, whose perinatal course we had previously reported [2], was first found to have a solid and cystic vascular right sided neck mass on antenatal ultrasound at 20 wk gestation. There was borderline polyhydramnios at that time. The diagnosis of a giant cervical teratoma was confirmed on fetal magnetic resonance imaging (MRI) at 30 wk (Fig. 1). As there was evidence of airway obstruction, the infant was delivered by ex-utero intrapartum treatment (EXIT) procedure at 35 wk with successful intra-procedural intubation.

Postnatal MRI with magnetic resonance angiography further delineated the anatomy prior to surgical resection (Fig. 2). The solid and cystic tumor centered on the infratemporal fossa extended into the oro, naso and hypopharynx. It was supplied by a large branch of

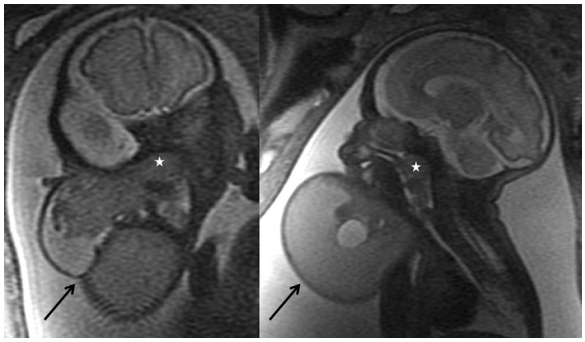

Fig. 1. Coronal and sagittal T2-weighted images of the fetal head and neck at 30 wk. The large solid and cystic right sided neck mass (black arrow) extending into the pharynx (white star) is demonstrated.

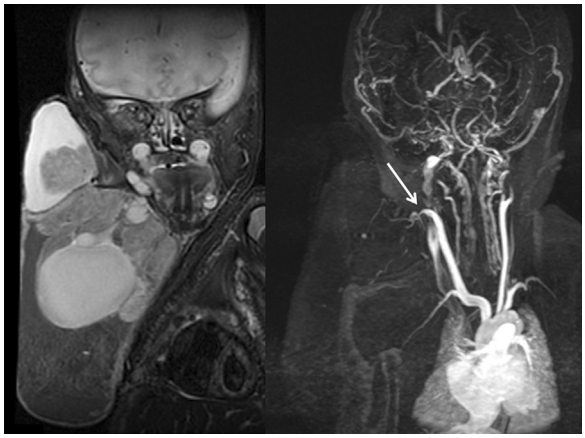

Fig. 2. Coronal T2-weighted magnetic resonance imaging and time of flight magnetic resonance angiography maximum intensity projection of the neonatal neck. The right sided neck tumor and its supply off the right carotid artery (arrow) are demonstrated.

the external carotid artery. Initial subtotal resection at 1 wk of age was followed by two further resections in the first year of life to remove the residuum at the skull base. The histopathology confirmed a mature teratoma. The right external carotid artery, right half of the pharynx and of the soft palate were sacrificed at surgery.

Over the subsequent year, the patient had variable problems related to his velopalatal insufficiency, but there was no evidence of tumor recurrence. At the age of 9 yr he represented acutely with a right sided intracerebral hemorrhage secondary to a right frontal AVM delineated on computed tomography angiography (Fig. 3). The malformation was fed by the right middle cerebral artery and drained into the internal cerebral

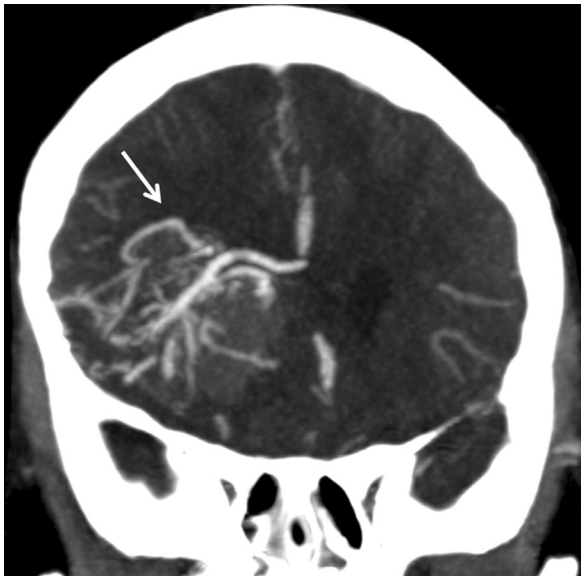

Fig. 3. Coronal computed tomography angiography image demonstrating the high density right frontal hematoma, and the large underlying arteriovenous malformation (arrow).

veins. The hematoma was evacuated and the AVM resected, with no residuum seen on formal follow-up angiography. Retrospective review of the neonatal preoperative magnetic resonance angiography showed relative enlargement of the right common carotid artery, and of the right external carotid artery feeding the tumor (Fig. 2).

3. Discussion

The co-occurrence of a giant cervical teratoma and an ipsilateral cerebral AVM has not previously been documented.

Although they have potential for delayed malignant transformation, mature teratomas are benign tumors and are typically cystic. They can be very vascular and contain tissue elements of one to three germ layers. They are the most common germ cell tumor in the fetus and neonate, and most often occur in the sacrococcygeal region [3]. Cervical teratomas are much more rare with an incidence consistently reported as between one in 20,000 and one in 40,000 live births [1]. The majority are now diagnosed antenatally. They often cause esophageal and airway obstruction, which can lead to polyhydramnios. Fetal MRI allows accurate delineation of the anatomy, and planning of delivery. Delivery by EXIT procedure, which allows access of the airway while the

fetus remains on placental support has significantly improved postnatal outcome [1]. Associated intracranial anomalies are extremely unusual with the only reported cases being one of agenesis of the corpus callosum [4] and one of direct extension into the middle cranial fossa [5].

Cerebral AVMs are also relatively rare. The prevalence has been reported as lying between 0.02–0.2%, with a new detection rate between 0.89 and 1.34 cases per 100,000 [6]. They usually present as an acute cerebral hemorrhage and account for the majority of childhood hemorrhagic strokes [7]. Despite this, their etiology remains unclear. Although they can be associated with certain genetic syndromes, such as hereditary hemorrhagic telangectasia, the majority are sporadic. There is little evidence for them being a congenital lesion arising during embryonic development. Recently Kim et al. [8] have proposed a "response-to-injury" paradigm to explain the pathogenesis of these sporadic brain AVMs based on findings from clinical research studies of AVM patients and animal models investigating AVM formation. They propose that inciting events may include sequelae of even modest injury (e.g. traumatic, mechanical, and inflammatory). Whereas the normal response would involve angiogenesis, endothelial mitogenesis and vascular stabilization, if superimposed on an underlying microscopic vascular structural defect or an underlying genetic predisposition such as a mutation in angiogenic genes, the normal injury response shifts towards an abnormal dysplastic response.

In the case we report, the patient had two separate and rare pathologies which presented metachronously. The fact that they were linked by occurring in a common vascular territory leads us to suggest that they may be associated rather than a random coincidence. Although this has not previously been documented, we suggest that the presence of the hypervascular cervical teratoma fed by the right external carotid artery during fetal development may have provided a sufficient insult to the cerebral vascular territory distal to this to have led to the formation of the AVM as proposed by Kim et al. [8] in their "response to injury paradigm for brain AVM pathogenesis.

In conclusion, we describe the co-occurrence of an antenatally diagnosed giant cervical teratoma with a large arterial supply from the external carotid artery with a late presenting cerebral AVM fed by the ipsilateral middle cerebral artery. We postulate that there may be an etiological association between the two, with the AVM having arisen as a "response to injury" [8] and suggest that screening of the brain at birth with MRI by extending the diagnostic imaging of the neck to include the brain may be worth considering in patients with giant neck lesions with high vascularity in order to document any possible associated intracranial vascular malformation.

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