Frontal Lobe Lipoma Associated with Cortical Dysplasia and Abnormal Vasculature

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SUMMARY – *Intracranial lipomas (ICLs) are rare lesions, the vast majority encountered as incidental findings on imaging studies. ICLs are generally pericallosal midline lesions and thought to be asymptomatic and can be accompanied by additional intracranial congenital malformations. We describe a 17-year old male with an unusual case of ICL on the frontal lobe associated with cortical dysplasia and abnormal vasculature mimicking arteriovenous malformation on magnetic resonance images.*

Introduction

Intracranial lipomas (ICLs) are very rare congenital malformative lesions accounting for about 0.1-1.7% of all intracranial tumours [1,2]. The majority of the ICL occur at or near the midline, mostly in the callosal cisterns [1]. Other involved locations include the quadrigeminal-ambient cistern, suprasellar, interpeduncular, cerebellopontine and sylvian cisterns [2]. Lipomas on the cerebral surface are extremely rare [2-7]. A variety of brain malformations have been reported to be associated with ICLs, mostly agenesis or dysgenesis of the adjacent structures with the lesions located at midline [8] and cortical dysplasia with lesions located at the cerebral surface [2].

Herein we describe a rare case of cerebral surface lipoma with an unusual location in the inferior medial part of the right frontal lobe in association with cortical dysplasia and abnormal vasculature mimicking an arteriovenous malformation (AVM) on initial magnetic resonance (MR) scan.

Case Report

A 17-year-old male patient was admitted to our hospital with a prior diagnosis of cerebral AVM. He had a history of syncope after a head trauma two months previously while playing soccer. Unenhanced cranial computed tomography (CT) showed a hypodense lesion outside the cortex in the right medial frontal region, with attenuation (–55 - –65 HU) characteristics of adipose tissue without calcification. No associated haemorrhage was detected (Figure 1).

A lesion in the right inferior frontal region was detected on MR scan performed immediately after the trauma in emergency conditions and considered an AVM. The patient had no episodes of epileptic seizures. The physical and neurologic examinations were unremarkable. We re-evaluated the patient for the purposes of differential diagnosis.

Cranial magnetic resonance imaging (MRI) was performed to evaluate the lesion and adjacent cerebral parenchyma. MRI revealed an extra-axial lesion which was hyperintense on T1-weighted (Figure 2A), T2-weighted (turbo spin-echo, TSE) (Figure 2B) images. Because of the signal intensity of the lesion on the T1 and T2-weighted images we performed fat-saturated T2-weighted images to rule out fat presentation. On fat-saturated T2-weighted, images suppression of the lesion was detected (Figure 2C). The CT and MRI characteristics of the lesion were compatible with lipoma. The lipoma adhered to the medial inferior aspect of the gyrus rectus and medial orbital gyrus. Thickness of the cortex adjacent to the mass was increased and had an abnormal sulcal pattern consistent with cortical dysplasia (Figure 2D).

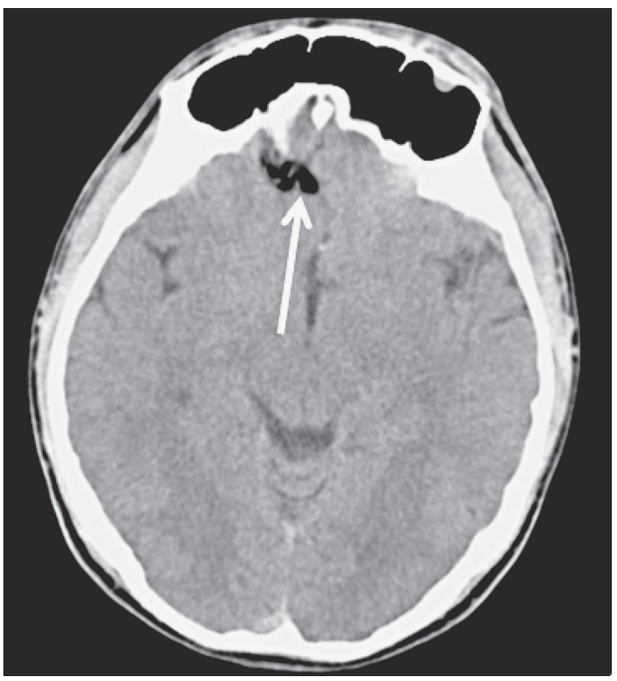

Figure 1 Unenhanced cranial CT scan showing a well-defined lobulated hypodense lesion (arrow), with no evidence of calcification in the right medial frontal region, with attenuation (–55 to –65 HU).

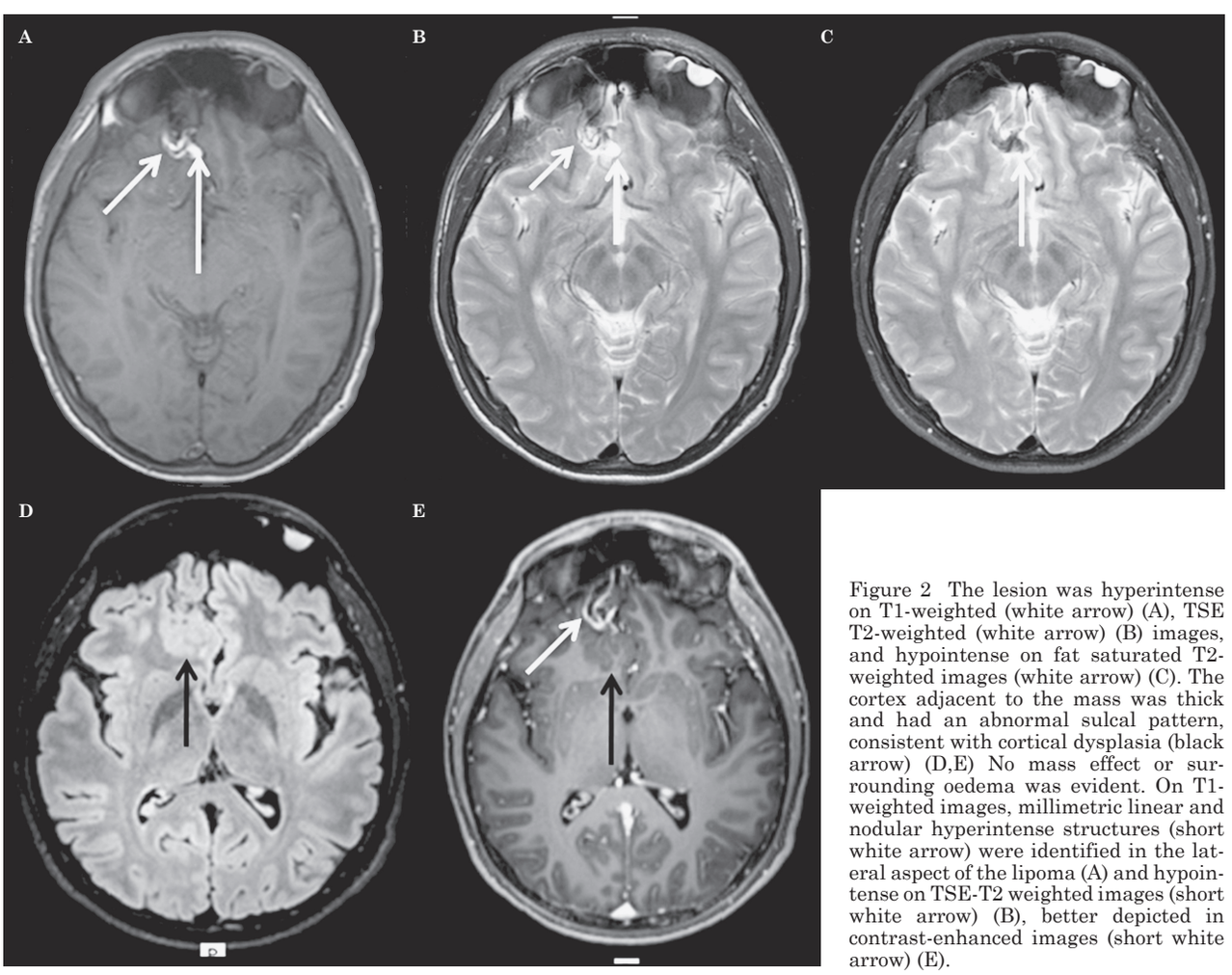

Figure 2 The lesion was hyperintense on T1-weighted (white arrow) (A), TSE T2-weighted (white arrow) (B) images, and hypointense on fat saturated T2-weighted images (white arrow) (C). The cortex adjacent to the mass was thick and had an abnormal sulcal pattern, consistent with cortical dysplasia (black arrow) (D,E) No mass effect or surrounding oedema was evident. On T1-weighted images, millimetric linear and nodular hyperintense structures (short white arrow) were identified in the lateral aspect of the lipoma (A) and hypointense on TSE-T2 weighted images (short white arrow) (B), better depicted in contrast-enhanced images (short white arrow) (E).

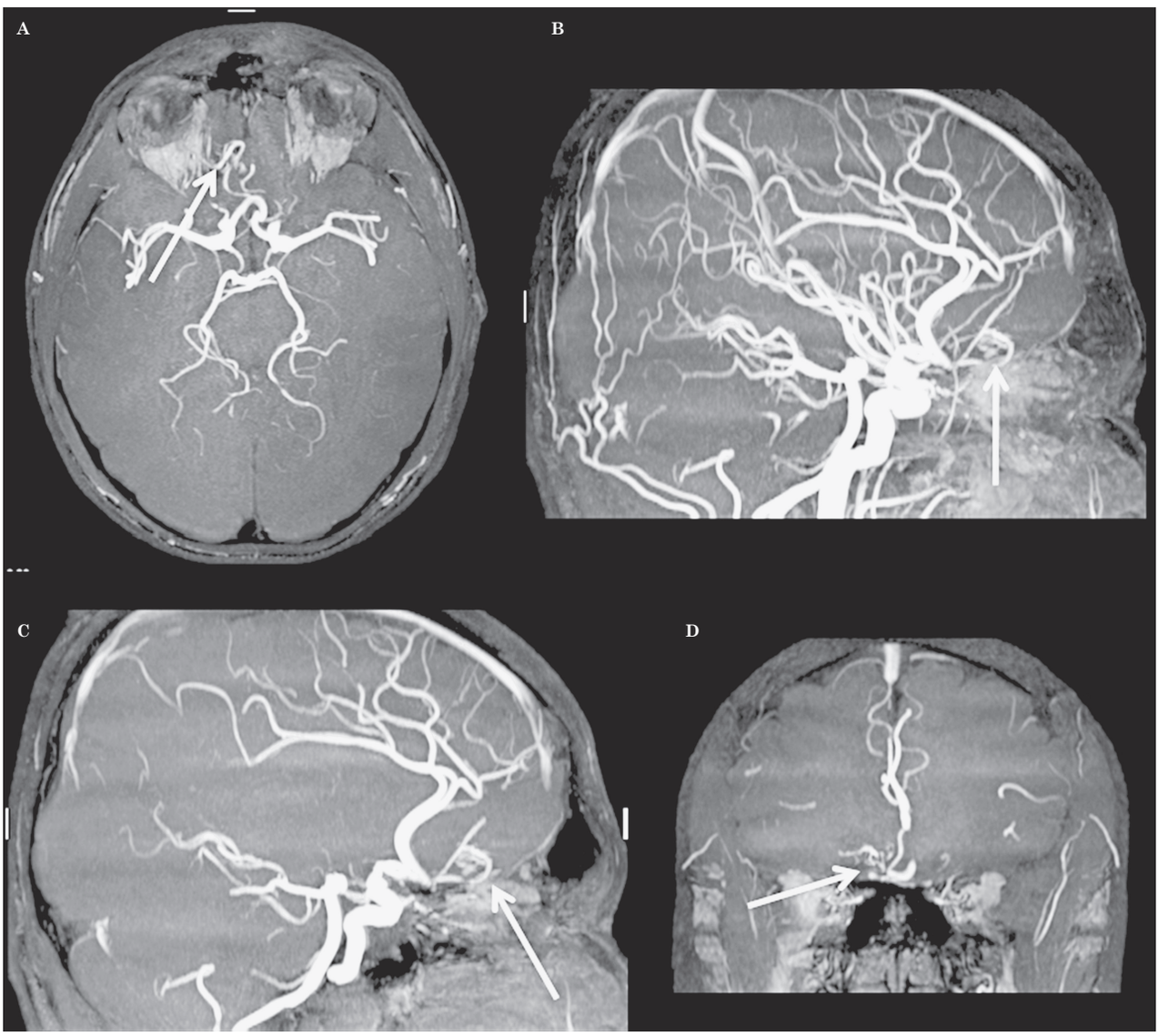

Figure 3 TOF MRA. Abnormal arterial branches (arrows) originating from the anterior cerebral artery in axial (A), sagittal (B,C), coronal (D) images.

Additionally, millimetric linear and nodular hyperintense structures were identified in the lateral aspect of the lipoma (Figure 2A) on T1-weighted images, which were hypointense on TSE-T2-weighted images (Figure 2B). These were better seen in contrast-enhanced images (Figure 2E).

Magnetic resonance angiography (MRA) performed using the time-of-flight (TOF) technique confirmed the presence of abnormal arterial branches originating from the anterior cerebral artery (Figure 3). No abnormally large vessel was detected that could represent an enlarged drainage vein. Nonetheless, there was no evidence of an AVM on TOF MRA.

Considering the limitations of TOF MRA, a conventional cerebral angiogram was performed to rule out a vascular malformation. The angiogram revealed an enlarged polar frontal branch of the right anterior cerebral artery feeding the region of the cortical dysplasia and lipoma seen on the MRI (Figure 4). There was no evidence of an early draining vein or any venous anomaly. In the absence of a vascular malformation or arteriovenous fistula, these findings were considered suggestive of an arterial dysplasia.

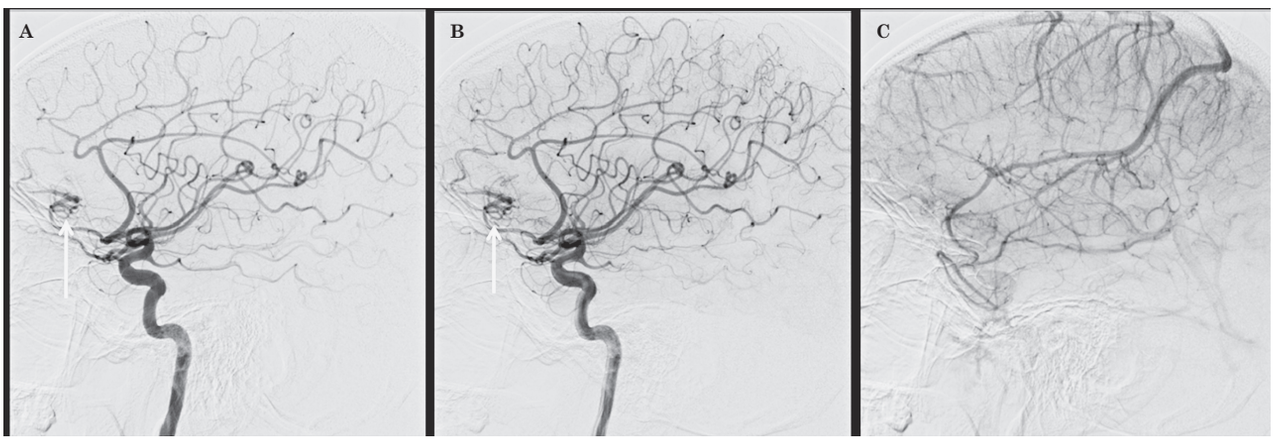

Figure 4 Cerebral angiography. Lateral projection of right internal carotid artery injection showing the abnormal vasculature (arrows) (A,B). There is no evidence of a draining vein (C).

Discussion

The first descriptions of intracranial lipomas (ICLs) were mainly through incidental findings at autopsy. ICL was first described in 1818 by Meckel, who found a chiasmatic lipoma, while in 1856 Rokitansky first described a pericallosal lipoma associated with corpus callosum agenesis [9].

ICLs are rare lesions. The majority of the lesions are interhemispheric and more than 50% are located in the callosal cisterns [1]. Other typical localizations are the quadrigeminal plate (25%), suprasellar-interpeduncular cisterns (14%), and the cerebellopontine angle (9%). The sylvian cistern is a rare location for lipomas having a reported incidence of 3.4% [1], and 5% [8]. Cerebral hemispheric lipomas located on the cortical surface are extremely rare [2,6,7]. In our case, the lesion was located superficially, overlying the right inferior medial frontal lobe.

ICLs are mostly asymptomatic, the vast majority of them appearing as incidental findings on imaging studies. However, they can cause symptoms differing according to their localization and comprising seizures, headache, paresis and cranial nerve impairments [8]. Hemispheric lipomas are more likely to be symptomatic than lipomas in the midline [6].

ICLs are neither hamartomatous nor neoplastic lesions. These are assumed to result from abnormal persistence and maldifferentiation of the meninx primitiva, the mesenchymal precursor of the leptomeninges, during the development of the subarachnoid cisterns [8]. These lesions should be accepted as being congenital malformations [8]. Various brain malformations are often seen in association with ICLs, including dysplasia or agenesis of the corpus callosum, absence of the septum pellucidum, spina bifida, encephalocele, myelomeningocele, malformation of the cortex and abnormal intracranial vessels [8,2]. There are a few cases of cortical malformations associated with ICLs in the literature, including pachygyria-like abnormalities and subcortical nodular heterotopia [10], an abnormal sulcal pattern-thickened cortex [4]. Various vascular anomalies have also been described in association with lipomas such as venous angioma, dilatation and prominent tortuous arteries, and saccular aneurysm [4,10,11].

In our case, the lipoma under the frontal lobe was also associated with cortical dysplasia and abnormal vasculature. To our knowledge, only four cases of ICLs on the cerebral surface have been reported with cortical dysplasia and abnormal vasculature in or around them [2-5]. It is not clear whether cortical dysplasia and lipoma arise together or not and if abnormal vasculature is caused by lipoma or cortical dysplasia. There have been two ideas for the combination of lipoma and anomaly, one is that an anomaly arises primarily whether with a lipoma or not, and the other is the anomaly occurs secondary to the preexistence of a lipoma [2,8]. Truwit et al. [8] suggested that ICLs are the result of abnormal persistence and maldifferentiation of the meninx primitiva which as a rule dissolves to form the primitive subarachnoid space by the end of eighth gestational week. Most cortical dysplasias are considered to result from injuries in the first half of the second trimester [2].

These concepts and the abnormal vascularization around the lesions support the idea that ICLs arise first, physical interruption and focal perfusion insufficiency due to preexistence of the lipomas might contribute to the formation of the focal cortical dysplasia [2].

In this case, we observed abnormal sulcal pattern and thickened cortex, consistent with

focal cortical dysplasia (Figure 2D), and tortuous and prominent vascular structures within and adjacent to the lipoma (Figures 2E and 3). There was no evidence of a feeding artery, early draining vein or any venous anomaly associated with an AVM. In the absence of a vascular malformation or arteriovenous fistula, these findings were considered suggestive of an arterial dysplasia. The other rare possibility of a spontaneously thrombosed AVM could be considered. However, there were no signs of arteriovenous shunt in the angiogram.

On CT and MR images, lipomas show a typical appearance. ICL appears as a homogenous low-density mass with attenuation characteristics similar to adipose tissue (–50 - –100 HU) on CT scan. MRI is sufficient to establish the diagnosis. On MRI, ICLs demonstrate a fat signal with short T1 and T2 relaxation times, i.e., a bright homogeneous signal on T1-weighted images and an intermediate signal on T2-weighted images. Moreover, fat may appear isointense or hyperintense on TSE T2 sequences. Also with MRI, fatty tissue has become easily detectable with the fat saturation techniques. MRI findings can be useful for differential diagnosis and detecting associated malformations.

ICls are rare benign malformations and primarily detected incidentally during radiological examinations but sometimes they can cause symptoms. They are easily diagnosed using CT or MR. Cortical and vascular malformations can be associated with ICLs located in the cortical sulci, therefore, a careful radiologic evaluation for any associated anomalies is necessary. Differential diagnosis should be made especially for vascular malformations that may need interventional procedures.

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