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Intracranial lipoma with extracranial extension through foramen ovale in a patient with encephalocraniocutaneous lipomatosis syndrome

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Abstract We present CT and MR features of a large intracranial lipoma located in the left sphenocavernosal region in a patient with a history of spinal intradural lipoma excision. The lipomatous lesion surrounded the internal carotid artery and trigeminal nerve and extended extracranially into the infratemporal region through a smoothly dilated foramen ovale.

Keywords Magnetic resonance imaging · Intracranial lipoma · Computed tomography

Introduction

Intracranial lipomas are uncommon lesions that comprise fewer than 0.1% of all intracranial tumors [1, 2]. A good proportion of them are located in the interhemispheric fissure, but other subarachnoid locations are also encountered [1, 2, 3]. We report a patient with a left sphenocavernosal lipoma associated with an ipsilateral temporal subcutaneous lipoma and alopecia. This constellation of findings suggested encephalocraniocutaneous lipomatosis syndrome [2, 4, 5, 6]. A distinguishing feature of the case was the extracranial extension of the sphenocavernosal lipoma through the foramen ovale.

Case report

A 9-year-old male patient, who was being treated for paraplegia, was referred for a cranial CT examination due to a congenital left temporal soft-tissue swelling and alopecia (Fig. 1). On physical examination he was found to have severe visual deficit in the left eye secondary to chorioretinitis; he also had mild mental retardation. The medical history revealed excision of a spinal lipoma 9 months previously. Retrospective examination of the MR images showed an extramedullary lipomatous mass located on the dorsal pial surface of the lower thoracic cord (Fig. 2). There was no finding suggestive of spinal dysraphism.

Cranial CT demonstrated a large fat-density lesion measuring –110 HU in the left sphenocavernosal region. The lesion did not contain any calcification or show any contrast enhancement. The cavernosal part of the left carotid artery was encased by the fatty mass. The lesion extended to the infratemporal region through an enlarged foramen ovale (Fig. 3). Also noted was a thickening in the left temporal subcutaneous tissue, consistent with subcutaneous lipoma. MR examination also showed the sphenocavernosal lesion surrounding the internal carotid artery and trigeminal nerve, as well as the infratemporal extension through the foramen ovale (Fig. 4). There was no accompanying congenital cranial malformation. Since there were no symptoms attributable to cranial nerve involvement or mass effect of the lesion, surgery was not considered and the patient was discharged for clinical follow-up.

Discussion

Intracranial lipomas are rare lesions accounting for fewer than 0.1% of all intracranial tumors [1, 2]. Although there are several suggestions with regard to

their pathogenesis, a convincing hypothesis proposed by Truwit and Barkovich states that these lesions result from abnormal persistence and maldifferentiation of the meninx primitiva during embryological development of the subarachnoid cisterns [3]. Thus, these lesions should be accepted as being congenital malformations rather than hamartomas or neoplasms. The authors reach this conclusion from the following observations in their series of 42 patients with intracranial lipomas: First, the most common locations of the lesions corresponded to the temporal resorption of the meninx primitiva. Second, in half of the cases, there was an accompanying congenital brain anomaly such as callosal agenesis or hypoplasia. Finally, these lesions enveloped adjacent nerves and vessels, rather than displacing them, unlike most tumors do [3].

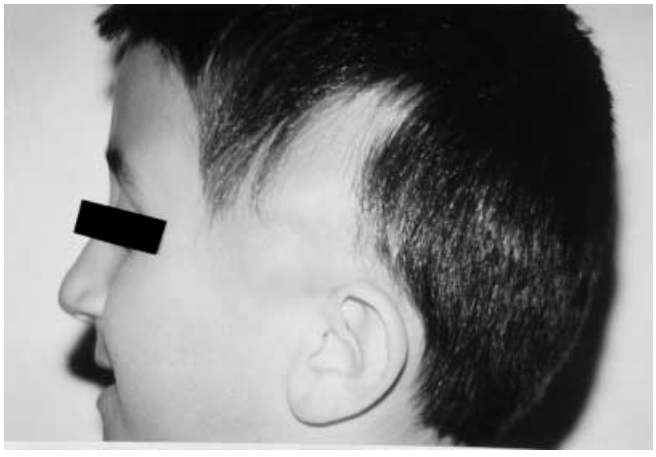

Fig. 1. Photograph of the patient showing the soft-tissue swelling and alopecia in the left temporal region

Intracranial lipomas are very rarely associated with cutaneous lipomas. A few patients have been reported to have accompanying nasal or calvarial lipomas [2]. Intracranial lipomas may occasionally be a component of encephalocraniocutaneous lipomatosis syndrome (ECCL) [2, 4, 5, 6], which was the case in our patient. ECCL is an uncommon congenital neurocutaneous syndrome which primarily involves ectodermal and mesodermal tissues and presents with extensive scalp alopecia, soft subcutaneous craniofacial masses, lipomas, connective tissue nevi of the eyelids and surrounding areas, pterygium-like choristoma of the ocular conjunctiva, mental retardation, motor deficits and seizures [4, 5, 6]. The clinical findings of this syndrome may overlap with other neurocutaneous syndromes, including sebaceous nevus syndrome, oculocraniocutaneous syndrome (OCC), and Proteus syndrome. Sebaceous nevus syndrome and ECCL are regarded as continuums of phenotypic expression. Proteus syndrome, which usually lacks brain anomalies, is accepted as a generalized form of ECCL. Differentiation of ECCL from OCC is facilitated by the absence of facial lipomas and scalp

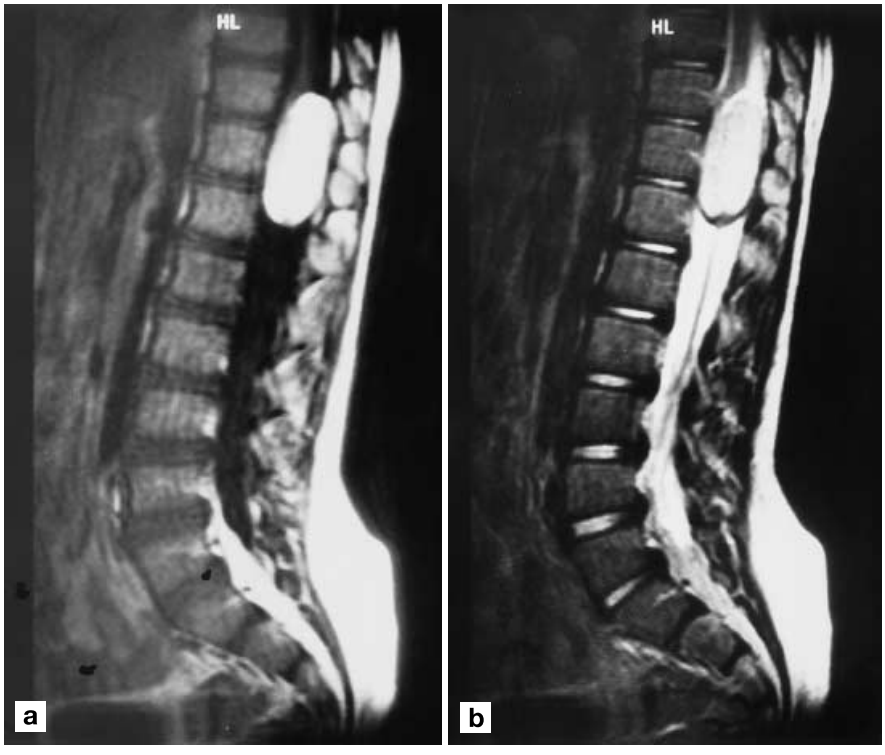

Fig. 2a, b. Sagittal T1-weighted (**a**) and T2-weighted (**b**) images show the intradural lipoma isointense with the subcutaneous fat located in the distal cord

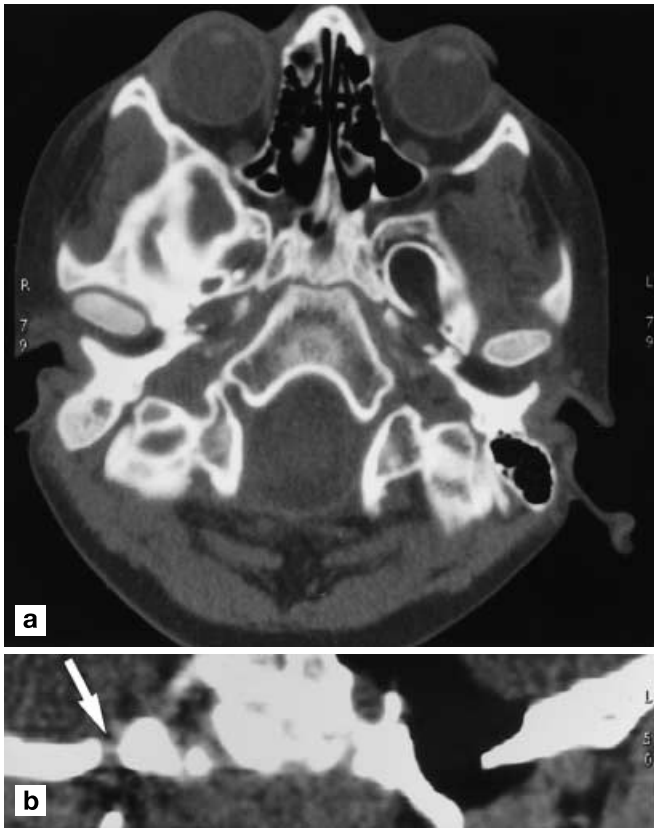

Fig. 3. **a** Axial CT section at the base of the cranium shows the smoothly dilated left foramen ovale by a low density mass. **b** Coronal CT reformatted image shows the infratemporal extension of the sphenocavernosal lipoma through the enlarged foramen ovale. Note the normal-sized right foramen ovale (*arrow*)

alopecia in the latter. Cranial findings reported in ECCL include enlargement of the lateral ventricle, widening of the subarachnoid spaces, arachnoid cyst, lack of normal insular opercularization, dysplastic cortex, corticopial calcifications, thinning of the corpus callosum, and intracranial lipoma. A peculiar feature in our patient was the presence of an intracranial lipoma, which has been described in only a minority of patients with ECCL [5].

Whether an isolated finding or part of a syndrome, the diagnosis of intracranial lipomas is usually straightforward. On CT, they are characterized by a typical fat density ranging from –50 HU to –100 HU. They should be easily differentiated from dermoid or epidermoid cysts, teratomas and occasional cystic meningiomas, which have a more inhomogeneous appearance and an attenuation value above –50 HU [7].

Nearly half of all intracranial lipomas occur in the interhemispheric fissure [3, 8]. The remainder are seen in the quadrigeminal/superior cerebellar cistern, suprasellar/interpeduncular cistern, cerebellopontine angle, Sylvian cistern, and other subarachnoid spaces [3]. This case illustrates an uncommon site of involvement: the

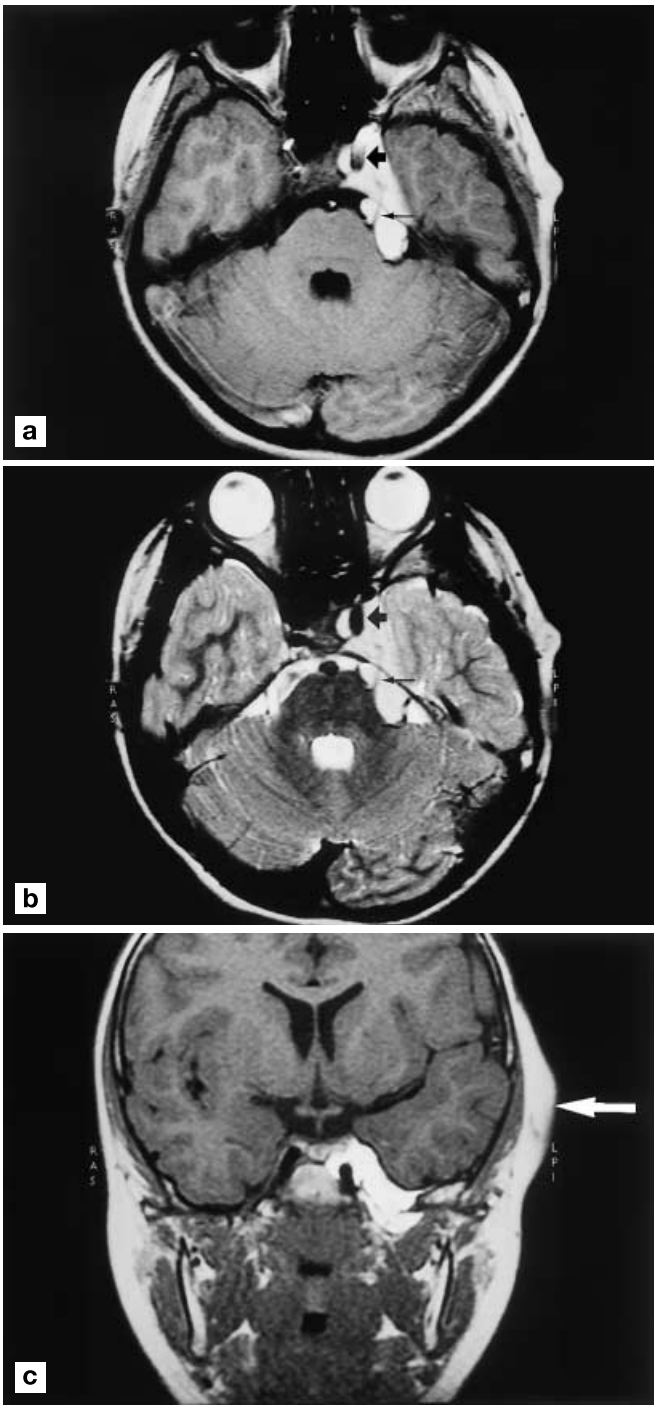

Fig. 4. **a, b** Axial T1-weighted (**a**) and T2-weighted (**b**) MR images show the hyperintense sphenocavernosal lipoma extending to the left cerebellopontine angle and encasing the left carotid artery (*thick arrow*). The left trigeminal nerve passes through the lesion (*thin arrow*). **c** Coronal T1-weighted MR image demonstrates the infratemporal extension of the lesion and the subcutaneous lipomatous mass (*arrow*) in the left temporal region

temporal–parasellar region. A further discriminating feature was the enlargement of the foramen ovale, through which the mass extended extracranially in a rather dumbbell-shaped fashion. This partly resembles previous case descriptions of posterior fossa extension of cervical lipomas through the foramen magnum [7, 9]. Sabates et al. reported a patient with intracanalicular lipoma which had enlarged the optic canal and extended posteriorly to involve the ipsilateral optic chiasm [10].

Intradural spinal lipomas without associated spinal dysraphism are also rare and form fewer than 1% of all spinal tumors [11]. Coexistence of spinal and intracranial lipomas is even more infrequent. Only three patients suspected of having ECCL have been reported to harbor a spinal cord lipoma [4].

To conclude, intracranial lipomas are rare lesions, which may occasionally extend extracranially through potential spaces such as calvarial foramina, in a fashion reminiscent of neurogenic tumors. As in the case described, the presence of accompanying cutaneous and spinal lipomas suggests the diagnosis of a neurocutaneous syndrome with a strong degree of confidence.

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