

Case Report

Interhemispheric Osteolipoma with Agenesis of the Corpus Callosum

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Osteolipoma is an ossified lipoma with distinct components of fat and bone. We present a case of interhemispheric osteolipoma associated with total agenesis of the corpus callosum. A 20-year-old man complained of severe headache, nausea and vomiting. Brain computed tomography showed a low-density mass in an interhemispheric fissure, with high T1 and T2 magnetic resonance signals compatible with fat. The mass measured 4.9 × 2.9 cm in size and showed peripheral calcifications. There was another small piece of same signal mass within the lateral ventricular choroid plexus. The interhemispheric lesion was removed by an interhemispheric approach. Osteolipoma is rare in interhemispheric region, however, it should be a differential diagnosis of lesions with fat intensity mass and calcifications.

KEY WORDS : Agenesis · Corpus callosum · Intracranial · Lipoma · Osteolipoma.

INTRODUCTION

Intracranial lipoma is a congenital malformation that develops by abnormal differentiation of normally present primitive tissues^{1,21}. Interhemispheric lipoma is the most common intracranial lipoma, and is often associated with agenesis of the corpus callosum.

Osteolipoma is an ossified lipoma with distinct components of fat and bone. Although intracranial lipomas occasionally contain amorphous and/or punctate calcifications^{11,22}, they rarely ossify¹⁷. Osteolipomas may be developed in the interpeduncular region¹⁹, tuber cinereum^{1,12,14,20} and suprasellar area¹⁷. However, radiological and histological evidence does not clearly confirm reports of interhemispheric osteolipomas.

Here, we present a case of interhemispheric osteolipoma associated with total agenesis of the corpus callosum and discuss its developmental pathogenesis.

CASE REPORT

A 20-year-old man was presented with a one-month history of severe headache accompanied by intermittent nausea and vomiting. At the time of presentation, he was serving an obligatory term in the military. He had no history of seizure, febrile convulsion, or other significant event, and general physical and neurological examinations showed unremarkable findings. Prior to joining the military, he had graduated from high school without any physical problems and worked as a cook. Brain computed tomography (CT) showed a low-density mass in an interhemispheric fissure (Fig. 1A), with high T1 and T2 magnetic resonance signals comparable to fat (Fig. 1B, C). The mass measured 4.9 × 2.9 cm and showed peripheral calcification. A small portion of the same mass corresponding to that signal appeared within the lateral ventricular choroid plexus. We also noted agenesis of the corpus callosum. These findings suggested the presence of a lipoma or rupture of a dermoid cyst. We thought that the headache was related to the mass and that surgical removal could relieve the headache.

The interhemispheric lesion was removed piecemeal fashion by an interhemispheric approach. The lesion had encased and adhered to both anterior cerebral arteries, and formed multiple calcifications. Interspersed fibrous tissue

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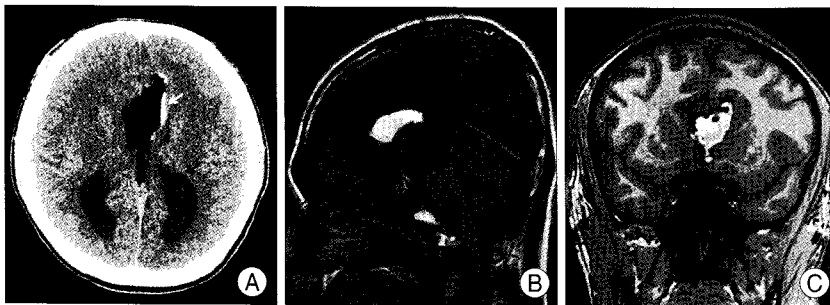


Fig. 1. Computed tomography (A) shows a fatty mass with peripheral calcification in the interhemispheric fissure (white arrow), and a separate lipoma in the left lateral ventricle (black arrow). Magnetic resonance T1-non-enhanced sagittal (B) and coronal (C) images show corpus callosal agenesis and vascular voids.



Fig. 2. Microscopic view of the tumor showing lobules of mature adipose tissue separated by strands of fibrous connective tissue and lamellar bone islands (H & E, $\times 100$).

adhered firmly to both sides of the mesial hemisphere. The mass was partially removed, and the patient's headache improved thereafter.

Histologically, the specimen consisted of mature adipocytes separated by thin fibrous strands and lamellated bone. A few osteoblasts without atypia were found. The histology of the lesion was consistent with osteolipoma (Fig. 2).

DISCUSSION

Locations and symptoms of lipomas and osteolipomas

Intracranial lipomas are rare, accounting for about 0.1% to 0.5% of all intracranial tumors^{6,11}. Lipomas occur preferentially along the midline and within the subarachnoid cisterns. Intracranial lipomas occur most frequently in the interpeduncular-, cerebellopontine angle-, sylvian- and prepontine cisterns^{10,18}, and interhemispheric lipoma is the most common type.

Osteolipomas are lipomas associated with bone. Reports

suggest a rather limited distribution for these tumors in the interpeduncular cistern¹⁹, suprasellar area¹⁷ and tuber cinereum^{1,12,14,20}. In 1977, Friede⁸ reviewed 24 lipomas of the tuber cinereum, based on autopsy findings, and found that 11 of the 24 lipomas were partially ossified. He concluded that osteolipomas are displaced elements of the mesenchyme and encompassing walls of the craniopharyngeal duct. Mackenzie et al.¹²

reported an occurrence of osteolipoma at the tuber cinereum that presented with headache, blurred vision and unilateral facial numbness. Their specimen showed mature adipose tissue containing small blood vessels and delicate fibrous septae, with a thin outer cortex of compact lamellar bone and a central core of marrow. The bone contained osteocytes, and the central marrow showed evidence of erythropoiesis and myelopoiesis. Other authors^{1,12,14,17,19,20} have reported osteolipomas at the same location. Based on these patient-series, Sinson et al.¹⁷ concluded that lipomas in the suprasellar and interpeduncular areas are resilient to ossification. The lesions in all of these cases were pea- or bean-sized, located in the suprasellar/interpeduncular fossa, and closely attached to adjacent neural structures. The reports did not mention of associated anomalies. Considering the interhemispheric locations, there were reports of lipomas containing calcifications and psammoma bodies^{9,21}, however they were not demonstrated as ossification histologically.

Osteolipomas in the tuber cinereum may induce precocious puberty¹ and when they occur in the interpeduncular fossa, facial numbness and weakness, eyelid depression, severe headache¹⁷ and monoparesis¹⁹ may occur. Two autopsy reports describe hypothalamic osteolipomas^{12,20}. In contrast to osteolipomas, intracranial lipomas induce many symptoms. Although usually benign and asymptomatic, lipomas may present with neurologic symptoms such as epilepsy, headaches, behavioral disturbances, and cranial nerve palsies. We inferred the cause of headache in our patient from previous cases of lipoma. Our patient complained of sudden severe headache, which subsided after partial removal of the osteolipoma. A previous report described cluster-like headache caused by a forehead lipoma, and the patient's headache disappeared after removal of the lesion². Intracranial lipoma has been reported to present with headache, dizziness and quadrantanopia in cases of quadrigeminal cistern lipoma and calcarine fissure lipoma⁷ and periorbital headache, ptosis, conjunctival injection and nasal congestion caused by lipoma of the interpeduncular

cistern⁵). The symptoms of intracranial lipoma are also related to the symptoms of hemifacial spasm¹⁶, autonomic symptoms⁵, cranial nerve palsies and epilepsy. As shown in our case, vessels and nerves are incorporated into lipomas rather than displaced¹⁸. Incorporated vascular or nervous structures, adjacent membranous structures or the mass itself can cause these symptoms. It remains unclear why the headache could be relieved by partial removal of the lesion. However, since those symptoms seemed to be related to neuronal irritation by the lipoma, such as hemifacial spasm, periorbital headache, and epilepsy, volume reduction could be helpful for relieving headache.

Embryopathogenesis

Intracranial lipomas are congenital malformations stemming from abnormal differentiation of the persistent meninx primitiva, which occupies the inner level of the pia arachnoid and dura⁴. The meninx primitiva is thought to be a mesenchymal derivative of the neural crest. The inner meninx is normally resorbed in an orderly process, as the subarachnoid cisterns expand into the space around it^{15,18}. Truwit and Barkovich¹⁸ described the preferential cisternal sites of intracranial lipomas, based on the orderly formation of the cisterns proposed by Osaka et al.¹⁵ They reported that since the meninx primitiva dissolves last in the region dorsal to the lamina terminalis, the chance that the meninx will differentiate abnormally is greatest in the interhemispheric area, which makes it the most common site for intracranial lipomas to occur. If the meninx primitiva regresses incompletely, adjacent neural structures may fail to develop or would develop improperly to form lipomas. Considering calcification and bone formation, Zettner and Netsky²² stated that the bony elements are foci of osseous metaplasia following calcification. de Villiers et al.³ described seven patients with lipoma of the corpus callosum associated with a frontal cranial defect, and suggested that the primitive mesenchyme gives rise to the meninx primitiva as well as to the fronto-facial skeleton, and that both of these lesions may stem from a disturbance in the neural crest. There is general consensus that all of the dermal bone contributing to the viscerocranium originates in the neural crest¹³. Based on these reports, we suggest that the interhemispheric osteolipomas accompanied by agenesis of the corpus callosum may generate from disturbance of neural crest development.

CONCLUSION

Osteolipoma is rare in interhemispheric area, however it should be a differential diagnosis of lesions with fat inten-

sity mass and calcifications.

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