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. 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, they rarely ossify. Osteolipomas may be developed in the interpeduncular region, tuber cinereum, and suprasellar area. 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, with high T1 and T2 magnetic resonance signals comparable to fat. 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...
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. Lipomas occur preferentially along the midline and within the subarachnoid cisterns. Intracranial lipomas occur most frequently in the interpeduncular-, cerebellar-, sylvian- and prefrontal zones, 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 cistern, suprasellar area, and tuber cinereum. In 1977, Fiedler 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 cranioopharyngeal duct. Mackenzie et al. 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 have reported osteolipomas at the same location. Based on these patient-series, Sinson et al. 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 associated anomalies. Considering the interhemispheric locations, there were reports of lipomas containing calcifications and psammoma bodies, however they were not demonstrated as ossification histologically.

Osteolipomas in the tuber cinereum may induce precocious puberty and when they occur in the interpeduncular fossa, facial numbness and weakness, eyelid depression, severe headache and monoparesis may occur. Two autopsy reports describe hypothalamic osteolipomas. 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 palsy. We inferred the case 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 lesion. Intracranial lipoma has been reported to present with headache, dizziness and quadranopia in cases of quadrigeminal cistern lipoma and calcarine fissure lipoma and periorbital headache, ptosis, conjunctival injection and nasal congestion caused by lipoma of the interpeduncular
cistern\(^5\). The symptoms of intracranial lipoma are also related to the symptoms of hemispherical spasm\(^6\), autonomic symptoms\(^5\), cranial nerve palsies and epilepsy. As shown in our case, vessels and nerves are incorporated into lipomas rather than displaced\(^6\). 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 hemispherical 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 dura\(^6\). The meninx primitiva is thought to be a meneshymal 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\(^6,18\). Truwit and Barkovich\(^16\) described the preferential cisternal sites of intracranial lipomas, based on the orderly formation of the cisterns proposed by Osaka et al.\(^5\). 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 Nestska\(^20\) stated that the bony elements are foci of osseous metaplasia following calcification. de Villiers et al.\(^9\) 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 crest\(^19\). 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 intens-