Ectopic Cerebellar Tissue in the Occipital Bone: A Case Report

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Abstract

Ectopic cerebellar tissue has only been described in isolated case reports, with only two reported cases in adult patients. We report the case of a 63-year-old woman with progressive, medically refractory headaches. A scan showed an intraosseous lesion of the midline occipital bone. Surgical resection of the soft tissue lesion was undertaken. Her headaches ceased postoperatively. Histopathological analysis revealed cerebellar cortical tissue with a surrounding meningothelial cell layer, characteristic of cerebellar ectopia. This is the second reported case of an intraosseous location of this lesion, and only the third case described in an adult patient. Our findings illustrate a rare cause of headaches and support the therapeutic roles of surgical treatment for this extremely rare condition.

Keywords

- ectopic cerebellar tissue
- glioneuronal ectopia
- intraosseous lesion
- headache
- occipital bone
- craniectomy

Introduction

Ectopic cerebellar tissue is an extremely uncommon finding, occurring as both an isolated finding and in conjunction with other neurological disorders. The condition remains poorly understood due to its rarity and diverse clinical presentations. Only two cases have previously been described in adults, and only one in an intraosseous location.1 We describe the case of an adult patient presenting with headaches which were found to be associated with a small nodule of ectopic cerebellar tissue in the midline occipital bone. Cessation of the headaches upon removal of the lesion suggests a causative relationship. We present clinical, operative, radiographic, and histopathologic findings and their implications for our understanding of this rare lesion.

Case Report

A 63-year-old Hispanic woman with no significant past medical or neurological history presented with several years of progressive, medically refractory occipital headaches. The pain was of a pounding quality and associated with nausea. Symptoms occurred daily, without obvious aggravating or alleviating factors. Work-up of primary and secondary headache disorders, with empiric medication trials where appropriate, had been negative over the preceding 2 years. Noncontrast computed tomography (CT) and magnetic resonance imaging (MRI) of the brain showed a 1-cm, well-demarcated spherical lesion in the occipital bone just to the left of midline. There was a small opening into the posterior fossa. The lesion appeared to be lytic on CT without obvious abnormalities in the surrounding bone. It paralleled cerebrospinal fluid (CSF) on all MRI sequences (►Fig. 1).

An exploratory suboccipital craniectomy for resection of the lesion was undertaken. Upon removal of the outer cortical bone overlying the lesion, a smooth-walled bony cavity was found. Inside the cavity laid a small mass of red and off-white friable tissue with a thin translucent surrounding membrane. The inner cortical bone was eroded revealing the underlying cerebellar dura. The dura appeared intact, and the lesion was...
confirmed to be anatomically isolated from the dura or brain. The lesion was carefully resected en bloc with a curette. The patient tolerated the procedure without any complications and returned home on the first postoperative day. Her headaches substantially decreased in frequency and intensity immediately postoperatively and remained controlled at the last follow-up 9 months later.

Histopathological analysis showed non-neoplastic cerebellar cortex comprising molecular and granular layers with a small number of Purkinje cells. A thin layer of fibrous tissue with overlying meningotheelial cells was also noted. The surrounding bone demonstrated reactive changes and benign spindle cells with immunohistochemical staining positive for epithelial membrane antigen, supporting meningotheelial origin (►Fig. 2).

**Discussion**

Ectopic cerebellar tissue, also known as cerebellar ectopia, was first reported in 1973 by Billings and Danziger, in a 9-month-old infant presenting with hydrocephalus, macrocephaly, and developmental delay. The patient was found to have a 1-cm

![Fig. 1 Preoperative radiographic findings. (A) Noncontrast head CT shows a well-demarcated lytic lesion of the occipital bone just to the left of midline (arrow), with a small opening into the posterior fossa. Noncontrast MRI of the brain on a 1.5 Tesla magnet shows that the lesion parallels CSF on (B) T1-, (C) T2-, and (D) T2-weighted fluid-attenuated inversion recovery sequences. CSF, cerebrospinal fluid; CT, computed tomography; MRI, magnetic resonance imaging.](image)

![Fig. 2 Histopathological findings following operative resection. (A–C) Hematoxylin and eosin staining at 200x magnification shows cerebellar cortex comprised of molecular (*) and granular (**) cell layers with occasional intervening Purkinje neurons (arrow). Overlying fibrous and meningotheelial cell layers (open arrow) with adjacent reactive bone (****) are also seen. (D) Benign spindle cells showing positive immunohistochemical staining for epithelial membrane antigen (open arrow).](image)
focus of ectopic cerebellar tissue in the roof of the fourth
ventricle that was resected successfully. To our knowledge,
this condition has been documented in only two previous
adult cases and 14 pediatric cases, without obvious gender
predilection. The anatomic location of the ectopic tissue in
prior cases is extremely heterogeneous, including the orbit, anterior fossa, suprasellar region, posterior fossa, and even the
spinal cord.

The diagnosis of ectopic cerebellar tissue requires a
high degree of suspicion due to its protean clinical manifesta-
tions. The lesion may be associated with abnormalities
such as teratoma, intracranial cysts, Chiari I malformation, or
spinal dysraphism. In these situations, presenting symptoms
are likely due to the associated abnormality, and the cerebel-
lar tissue is discovered intraoperatively and/or upon histo-
pathological examination. In other cases, the lesion may be
an incidental finding in screening studies or the work up of
other conditions. Alternatively, ectopic tissue may cause
symptoms related to mass effect (proptosis, hydrocephalus,
syringohydromyelia, etc.), impingement on structures
such as the optic nerve, or cortical irritation causing epilep-
sy. The radiographic appearance of the lesion may resemble
gray matter and/or CSF when occurring in isolation, or the
surrounding pathology when associated with other
abnormalities.

The pathogenesis of ectopic cerebellar tissue is not
known, but may relate to abnormal cell migration or differen-
tiation during development, such as in other forms of
glioneuronal ectopia and heterotopia. The condition has
also been hypothesized to arise from herniation of mature
tissue through a pial defect. Given the surrounding menin-
gothelial cell layer in our case, it is also possible these lesions
could represent congenital meningoencephaloceles or cystic
intraosseous arachnoid granulations/diverticula that subse-
sequently disconnect later in life. We propose that these
hypotheses may not be mutually exclusive; the underlying
mechanism may instead depend upon the anatomical loca-
tion and associated pathologies, converging upon histologi-
cally identical differentiated cerebellar tissue.

Only two prior reports have documented headache in
association with ectopic cerebellar tissue; the patients were
5 and 15 years of age, respectively. Interestingly, in both cases
the ectopic tissue was in the supracerebellar region and
connected to the cerebellum via a stalk, without associated
hydrocephalus or other abnormalities. Surgical resection
was both diagnostic and therapeutic in the former case,
while the diagnosis was made by diffusion tensor imaging
and the lesion managed expectantly in the latter. Our report
thus represents the first case of an adult presenting with
headache in this condition.

Similarly, there are only two prior reports of ectopic
cerebellar tissue in adults. Matyja et al described a 25-year-
old woman with complex partial seizures; midline deformities
including hypertelorism, low-set hairline, and nose deforma-
tions; and a large mass at the base of the mesial frontal lobes.
Resection of the mass disclosed ectopic cerebellar and heter-
genous neuroglial tissue and scattered thin-walled, dilated
vessels. Subsequently, Kawashima et al described the case of a
46-year-old woman who was incidentally found to have a
small lesion in the right side of the occipital bone, which was
excised during suboccipital craniotomy for hemifacial spasm.
This is also the only prior report of an intraosseous location of
ectopic cerebellar tissue. The radiographic, gross, and histo-
pathological characteristics were quite similar to the findings
in our case.

Given that the lesion was asymptomatic in the other
intraosseous case, it may be possible that our patient’s
headaches were due to a different cause and abated indepen-
dently of surgical resection. However, the mechanism
underlying pain in this condition remains unknown, and the
prior report of surgical resection curing a pediatric patient’s
headache must also be taken into account. Indeed, it is
possible that slow growth of the lesion over time caused it to
become symptomatic and accounted for the delayed presen-
tation in our patient.

The pathogenesis, natural history, and symptoms associ-
ated with ectopic cerebellar tissue remain poorly
understood. However, this condition must remain in the
differential diagnosis of isolated intraosseous masses in
adults. Surgical resection plays a critical role in providing
definitive tissue diagnosis to distinguish the lesion from
intraosseous neoplasms such as meningioma, lymphoma,
dermoid cyst, and hemangioma.

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Conflict of Interest
The authors have no conflicts of interest to declare.

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