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Case Report

Intraoperative discovery of a radiographically occult subependymoma obstructing the obex in a patient with a Chiari malformation – A rare case

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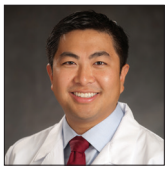

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ABSTRACT

Background: Chiari (type I) malformations are typically congenital. Occasionally, however, tonsillar herniation can arise secondary to cerebrospinal fluid leakage, posterior fossa or intraventricular mass lesions, or other etiologies. We present the first-ever case of an intramedullary subependymoma at the cervicomedullary junction associated with vertebral bone abnormalities and an acquired secondary Chiari malformation.

Case Description: A 60-year-old woman presented with a 3-year history of occipital, tussive headaches. Preoperative imaging was negative for mass lesions but demonstrated a Chiari malformation. She was recommended posterior fossa decompression with tonsillar shrinkage. During surgery, an intramedullary mass was incidentally observed, obstructing the obex at the cervicomedullary junction. Histopathological analysis of the resected lesion revealed a diagnosis of subependymoma.

Conclusion: Subependymomas can sometimes present a diagnostic challenge due to their subtle appearance in neuroimaging. Only rarely are such masses associated with an acquired Chiari malformation. No such case has previously been reported. We present a literature review on acquired Chiari malformations and discuss their management.

Keywords: Atlanto-occipital assimilation, Chiari malformation, Radiographically occult, Subependymoma, Vertebral anomaly

INTRODUCTION

The type I Chiari malformation is characterized by a caudal displacement of the cerebellar tonsils through the foramen magnum into the cervical canal of more than 5 mm. This malformation is generally congenital, affecting approximately 1 in 1000 births. Often asymptomatic in childhood, it can present as tussive headaches or neck pain in young adults. There are, however, cases of acquired Chiari malformations that have been described to arise secondarily, such as lumbar

shunting procedures or spontaneous cerebrospinal fluid (CSF) leakage.[1,3] In rare cases, obstructive lesions in the cervicomedullary junction have caused an acquired Chiari malformation with syringomyelia.[20]

Subependymomas are a benign, indolent subtype of ependymomas that develop from glial cells that line the ventricles and the central spinal canal.[7] We present a rare case of a patient presenting with what appeared to be a symptomatic Chiari malformation, which was found intraoperatively to have a radiographically occult intradural intramedullary subependymoma obstructing the obex with associated vertebral bone abnormalities. To distinguish between the congenital Chiari I malformation and an acquired tonsillar herniation, we use *Chiari malformation* to denote the latter (i.e., without the classifier, type I).

CASE DESCRIPTION

Clinical presentation

A 60-year-old woman with a medical history of gastroesophageal reflux disease, anemia, and knee osteoarthritis status post bilateral knee replacements presented with occipital, tussive headaches for three years. She also noted intermittent subjective left upper extremity weakness and bilateral upper extremity numbness. She denied bowel or bladder issues, vision changes, nausea, or vomiting. Her husband noted nighttime snoring without apneic episodes.

On physical examination, her cranial nerves were grossly intact, and her motor strength was 5/5 in all extremities with intact sensation to light touch. There were no signs of spasticity on examination, with no long tract signs. Cerebellar examination and gait were within normal limits.

Diagnostic results

Computed tomography (CT) of the brain demonstrated crowding at the craniocervical junction with downward displacement of the cerebellar tonsils and a syrinx of the visualized upper cervical cord. Furthermore, there was evidence on CT of partial occipitoatlantal assimilation as well as basilar invagination [Figure 1]. Magnetic resonance imaging (MRI) with and without contrast confirmed inferior descent of the cerebellar tonsils below the level of the foramen magnum by approximately 8 mm [Figure 2], consistent with a Chiari malformation. A well-delineated cystic dilation was also noted in the upper cervical cord, approximately 3.5 cm in the vertical dimension, with some subjacent T2 hyperintensity within the cord parenchyma, suggesting associated cord edema or gliosis [Figure 2]. Of note, there was no enhancement or other radiographic findings to suggest a mass lesion.

Treatment

Given that the patient was symptomatic from tonsillar descent, she was recommended surgical intervention. She underwent a suboccipital craniectomy and a C1 laminectomy for intradural decompression. The C1 lamina was noted to be atretic on one side and occipitalized on the other. It was resected to 1.5 cm from midline bilaterally. The suboccipital fossa bone and dura were noted to be thinned. The cerebellar tonsils were downwardly displaced and immobile. They were shrunken through subpial coagulation and partially resected. Deep to the cerebellar tonsils, a gray, semi-firm, and intramedullary lesion was noted

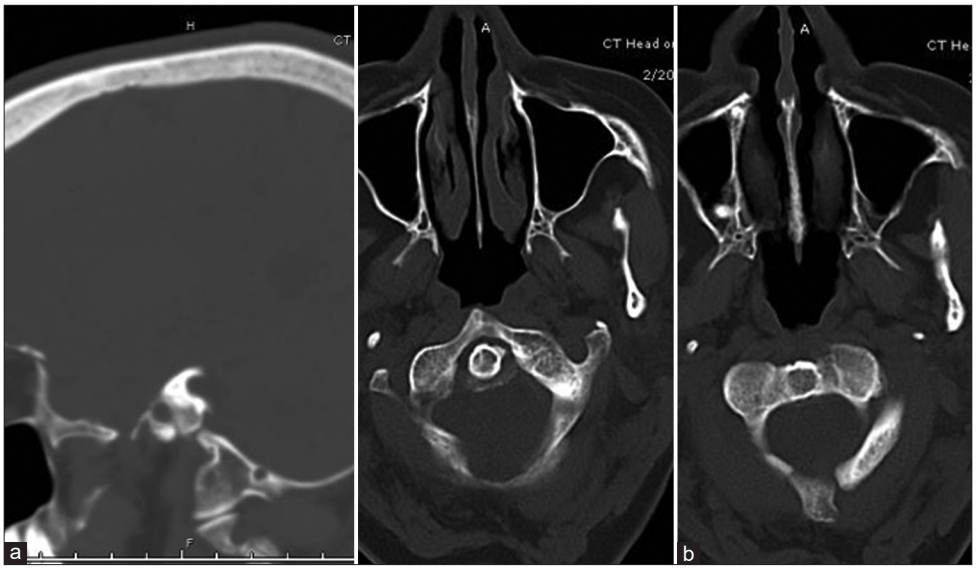


Figure 1: Computed tomography head without contrast (a) sagittal (b) and two axial views with evidence of occipitoatlantal assimilation

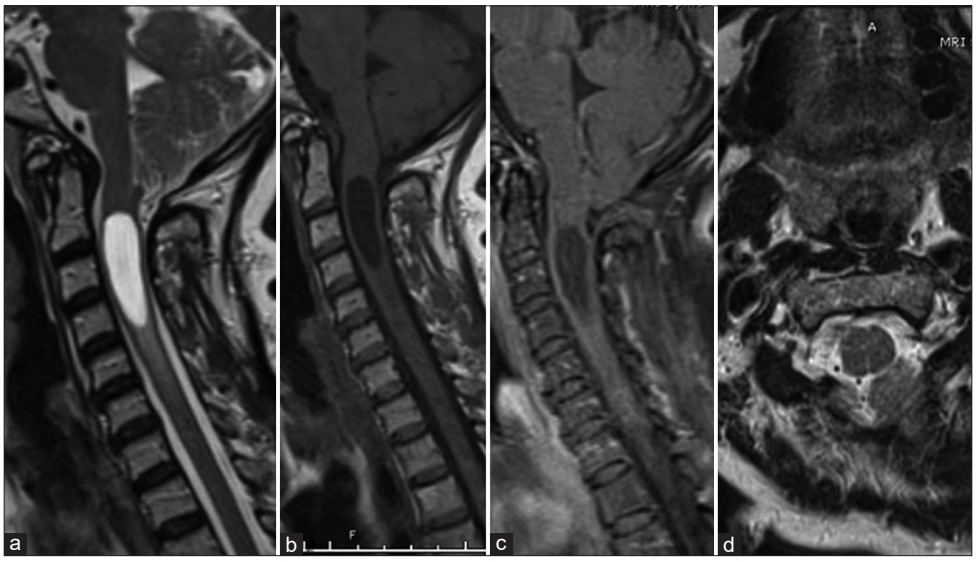


Figure 2: Preoperative magnetic resonance imaging (a) sagittal T2-weighted, (b) T1-weighted , (c) T1 post-contrast, and (d) axial T1 post-contrast views. There is a Chiari I malformation, with inferior descent of the cerebellar tonsils below the level of the foramen magnum by approximately 8 mm.

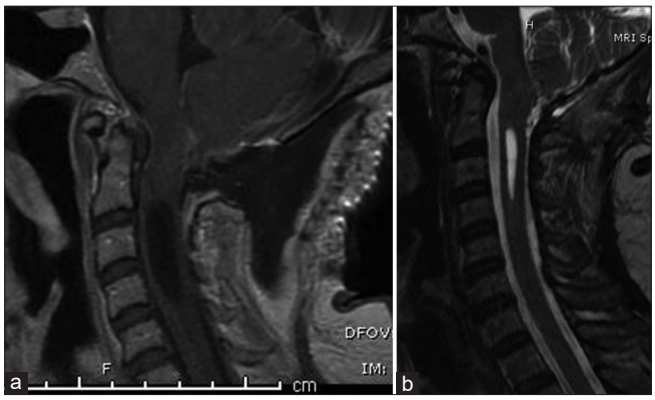
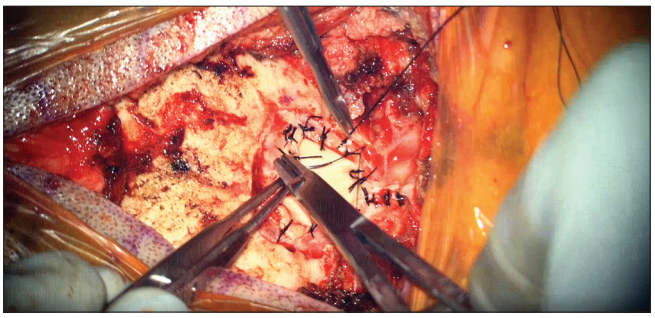


Figure 3: (a) Immediate postoperative magnetic resonance imaging (MRI) sagittal post-contrast sequence demonstrating no visible lesion in the cervicomedullary junction and (b) delayed 4-month postoperative T2-weighted MRI sagittal view with a significant reduction in the size of the cervical syringomyelia.

obstructing the obex, tracking both cephalad and caudad within the central canal [Video 1]. The portion of the mass that could be seen within the surgical field was resected; the portion that extended the caudad was left behind. We note that the lesion was adherent to the surrounding neural tissue. Following debulking, pulsatile CSF flow was noted through the obex. An expansile duraplasty was then performed using bovine pericardium. The intraoperative frozen section showed glial proliferation and two tiny fragments of fibrous tissue with fibrinous material.

Postoperative outcome and follow-up

Postoperatively, the patient reported left upper extremity paresthesias, new blurry vision, dizziness, and intermittent diplopia. Ophthalmology was consulted; both visual and extraocular symptoms were thought to be secondary to cerebellar disturbance from recent surgery. Dexamethasone therapy was started with moderate improvement of her visual symptoms. A postoperative MRI of the brain soon after surgery demonstrated decompression of the suboccipital region and expected postoperative changes [Figure 3]. A delayed MRI four months after surgery showed a significant decrease in size of the cervical syringomyelia [Figure 3].



Video 1: Intraoperative video of microdissection and partial resection of cervicomedullary subependymoma.

Pathological diagnosis

The final pathologic diagnosis of the posterior fossa mass was a World Health Organization (WHO) Grade 1 subependymoma. Histopathological review demonstrated pseudo-lumen rosettes and a nodular pattern [Figure 4]. Immunohistochemistry stained positive for glial fibrillary acidic protein and negative for P53 [Figure 4]. Ki67 positivity was less than one percent [Figure 4].

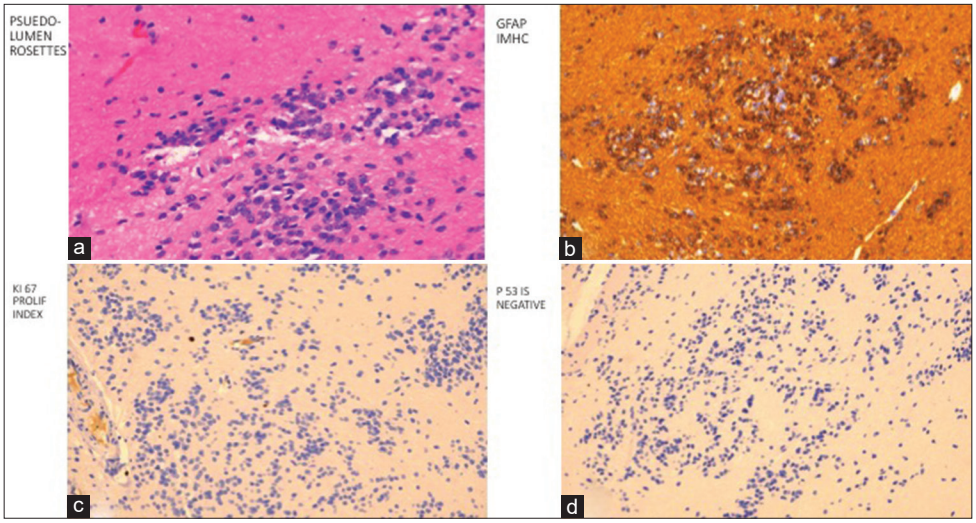


Figure 4: (a) Hematoxylin and eosin stain permanent block demonstrating pseudo-lumen rosettes, (b) glial brillary acidic protein positivity, (c) K167 proliferation index of <1% (c), and (d) P53 negativity.

DISCUSSION

Subependymomas, a subtype of ependymomas, are rare, benign tumors of the central nervous system that is histologically classified as low grade (WHO Grade 1). They represent between 0.07 and 0.7% of all intracranial tumors and arise from bipotent subependymal glial precursor cells.[2] Although, in many cases, they are asymptomatic and found incidentally on autopsy, the incidence of symptomatic subependymomas peaks in the fifth and sixth decades of life.[19] Presentation is typically from hydrocephalus or neurological deficits from progressive mass effect or spontaneous intra-tumoral hemorrhage, depending on the location and the size of the lesion. Subependymomas occur most commonly in the fourth (50–60%) or the lateral ventricles (30–40%) and less commonly in the cervical and cervicothoracic regions of the spine.[21]

Acquired Chiari malformations, defined as arising postnatally, are radiographically and clinically indistinguishable from congenital Chiari I malformations. In the literature, cases of acquired malformations have been associated with lumboperitoneal shunting, spontaneous CSF leak, and lumbar puncture.[13,14] Less commonly, space-occupying lesions have been reported as causes of secondary malformations, with meningiomas (36%) and arachnoid cysts (32%) as the most common offending lesions.[20] Notably, acquired Chiari malformations have a higher rate of associated syringomyelia (82%) compared to congenital Chiari I malformations.[20] We note that our patient had a syrinx as well. To the best of our knowledge, there have been only three other cases of a subependymoma associated with a Chiari malformation reported in the literature, all located in the 4th ventricle.[15,19] One such case was similarly discovered incidentally during the Chiari decompression operation, undiagnosed on preoperative radiology images.[19] We report the only case of an acquired Chiari malformation secondary to an intramedullary subependymoma at the cervicomedullary junction.

In our case, there was no appreciable lesion on preoperative imaging (CT or MRI), as confirmed by neuroradiologists, who reviewed the imaging again retrospectively. Imaging characteristics of intracranial subependymomas vary slightly by their location.[5] Typical MRI characteristics of subependymomas include low-to-intermediate signal intensity on T1-weighted scans and hyperintense in comparison to brain parenchyma on T2-weighted scans.[19] For lateral ventricle subependymomas, minimal contrast enhancement and no calcifications are typical, whereas 4th ventricular lesions typically demonstrate heterogeneous enhancement and calcifications.[5,19] The literature regarding cervicomedullary subependymomas is scarce.

Subependymomas tend to be slow-growing and indolent, evidenced by lower MIB-1 indices than other ependymal tumors, although their clinical course can vary widely based on location, size, and growth rate.[16] Symptomatic lesions often present with signs and symptoms of hydrocephalus and require surgical treatment. However, recurrence, subependymal spread, and central nervous system metastasis have been reported.[8,18] Furthermore, rapid expansion of a formerly asymptomatic subependymoma and sudden death from acute obstructive hydrocephalus has been described.[10,12]

In the cases of a mass lesion associated with an acquired Chiari malformation and syringomyelia, resection of the lesion is generally necessary to restore normal pulsatile CSF flow through the obex and achieve symptomatic improvement. Whether the traditional posterior fossa decompression through suboccipital craniectomy and possible C1 laminectomy is necessary in these cases remains unclear.[20] According to a systematic review by

Wang *et al*., there is no difference in rates of tonsillar ascent, resolution of syrinx, and symptomatic recovery between removal of the lesion only or resection and decompression.[20] In all but one of those patients, the syringomyelia significantly decreased or resolved on the last follow-up.[8,20]

Surgical management is the first-line treatment for symptomatic subependymomas.[19] The literature suggests that subependymomas can be cured with complete resection.[8] However, gross total resection cannot always be safely achieved, as in our case, due to adherence to the underlying brainstem parenchyma and the morbidity associated with pursuing the intramedullary component. For residual or recurrent disease, radiotherapy is a reasonable second-line treatment for these tumors if surgical intervention carries high morbidity. Radiation is typically excellent for local control. However, for symptomatic patients, local control of a slowly growing tumor will not address the presenting symptoms if these are resulting from obstruction of CSF dynamics.

As the recurrence rate is extremely low, there is limited evidence regarding the management of recurrent subependymomas. Ecker and Pollock reported a case of a recurrent fourth ventricular subependymoma that after multiple subtotal resections was ultimately managed with stereotactic radiosurgery with no further recurrence at 54 month follow-up.[6] In contrast, local relapse with subependymal seeding has been described despite radiosurgery to the recurrence after near total resection, reinforcing the idea that these subtotally resected subependymomas should be closely monitored.[18] Furthermore, there is no evidence that chemotherapy is efficacious, although an *ex vivo* functional analysis showed that cytotoxic agents interfering with oncogenes identified in subependymomas may have a clinical impact.[9,17] For all the reasons mentioned above, despite the typically benign clinical course of these tumors, due to presence of residual tumor and potential harmful consequences of a recurrence, our patient will require close clinical and radiological follow-up long-term.

Finally, our patient also had associated vertebral bone abnormalities (C1 atresia and atlanto-occipital assimilation). This finding itself is exceedingly rare. Morselli *et al*. reported a case of intradural extramedullary Grade II ependymoma with C1 partial agenesis.[11] In addition, there is one reported case of a suspected epidermoid cyst causing occipitalization of C1 with a Chiari malformation and a syrinx.[4] No cases of subependymomas associated with atlas bony abnormalities have been published. Our current report is the first described of a subependymoma with associated vertebral bone abnormalities and associated Chiari malformation.

CONCLUSION

Acquired Chiari malformations can be due to CSF shunting or leaking or, less commonly, secondary to a space-occupying lesion in the cervicomedullary junction. We present the only case of an acquired Chiari malformation secondary to an intramedullary subependymoma in the cervicomedullary junction, radiographically occult and incidentally discovered during surgery. Subependymomas can be challenging to diagnose on imaging, as illustrated by our case and another by Varma *et al*., both unnoticed on preoperative radiologic imaging. Most of these lesions are associated with syringomyelia, which nearly always resolves (or at least decreases), as do the neurologic symptoms after removal of the lesion. Therefore, suboccipital craniectomy and shrinkage of the cerebellar tonsils are often unnecessary in this patient population with acquired Chiari malformations. Finally, as subependymomas can recur or cause drop metastases in the central nervous system, close follow-up is required for these patients, especially following subtotal resection.

Ethical approval

The research/study complied with the Helsinki Declaration of 1964.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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