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Case Report

Mature teratoma of the cisterna magna in an adult patient

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ABSTRACT

Background: Teratomas are a rare subgroup of CNS germ cell tumors and are histologically classified into mature teratomas, immature teratomas, and teratomas with malignant transformation. CNS teratomas are infrequently found in the posterior fossa and pure mature teratomas of posterior fossa are rare especially in adults. We present a case of a young adult female with a mature teratoma in the cisterna magna.

Case Description: A 26-year-old female presented to the neurosurgery clinic with headache, nausea and vomiting for the past 1 year. She was found to have dysdiadochokinesia on neurological examination. Brain magnetic resonance imaging scan showed a-well defined lesion, hyperintense on T1 and hypointense on T2-weighted sequences located within the cisterna magna. She underwent a suboccipital craniotomy with resection of lesion. Histopathology confirmed the diagnosis of mature cystic teratoma.

Conclusion: Mature teratomas located in the posterior fossa among adults are rare in the literature. We report the second case of mature teratoma in the cisterna magna of an adult patient.

Keywords: Cisterna magna, Germ cell tumors, Mature teratoma, Posterior fossa

INTRODUCTION

Germ cell tumors, a very rare group of CNS neoplasms, comprise 0.3–0.6% of all primary intracranial neoplasms.[8] They are further divided into germinomas, nongerminomatous germ cell tumors, and mixed germ cell tumors.[8] Teratomas are a subgroup of nongerminomatous germ cell tumors, constituting 0.1–1.5% of all intracranial neoplasms and 18% of all germ cell tumors.[2,3,8,12] Histologically, they are subclassified into mature teratomas, immature teratomas, and teratomas with malignant transformation.[4,8,14] Mature teratoma is the most common subtype and contains well-differentiated, mature tissue elements, while immature teratomas have fetal-like tissue and the rare group teratomas with malignant transformation have a malignant somatic component.[8,14] Containing tissues derived from all three germ cell layers, teratomas are presumed to originate from misplacement or defective migration of pluripotent germ cells and are usually located at or near the midline due to its potential to harbor mislaid embryonic tissue.[2-4,7,8,10,12,14]

These congenital neoplasms, usually presenting during infancy and childhood, are generally located in the supratentorial compartment.[4,14] The two peaks of age are neonatal/infancy periods or the ages between 5 and 14 years, and the most common sites include the pineal

region (51%) and the neurohypophysis or suprasellar region (30%).[2,3,8,12] The occurrence of teratomas in the posterior fossa is uncommon and is extremely rare even so, among adults.[3,4,8,12,] Extensive literature is present to outline the clinical presentation, radiological appearance, and management strategies including surgical and adjuvant chemoradiation for mature teratomas, but due to the uniqueness of these lesions in the posterior fossa, limited data are available to adequately describe the disease process, its clinical and radiological presentation and management options in such cases.

Only 13 cases of mature posterior fossa teratoma in adults have been reported in the literature till now.[2,3-6,9-13,15,16] We report a case of a young female with mature cystic teratoma of the cisterna magna treated with surgical excision. We have reviewed the previous literature with the intent to gain better insight and develop a knowledgeable comprehension of the nature of this disease process.

CASE PRESENTATION

Our patient was a 26-year-old female who presented to the neurosurgery clinic with a sudden onset severe and continuous headache which was localized to the occipital region. It was exacerbated with postural change and associated with vertigo and vomiting. She was being managed along the lines of migraine for the past 5 years and only sought neurosurgical evaluation once her symptoms became severe during the last year. Neurological examination revealed no unusual findings other than a mild dysdiadochokinesia in her right hand.

Magnetic resonance imaging (MRI) demonstrated a 56 × 55 × 47mm, thick-walled infratentorial, and extra-axial lesion within the cisterna magna, which appeared hyperintense on T1-weighted images and hypointense on T2-weighted images [Figure 1]. There was an internal well-defined area at the right lateral aspect showing T1 hypointense rim and a signal dropout on susceptibility-weighted images. A stalk emerging from the posterior aspect of the lesion extended through the occipital protuberance and ended just beneath the skin causing its puckering. The lesion caused cerebellar compression and effacement of the fourth ventricle, thus causing dilation of lateral and third ventricles.

Suboccipital craniotomy was performed with the patient in a prone position. Intraoperatively, a greenish solid cum-cystic lesion was found. The inside contained hair follicles, a tooth, and fluid with mobile-oil-like consistency. An external ventricular drain was placed through a separate incision at Keen's point set at 15 cm of H20.

Histopathology revealed features consistent with mature teratoma. The cyst wall was lined by stratified squamous epithelium with underlying skin appendages. Areas of calcification with hemosiderin-laden macrophages and foreign body type giant cell reaction were also seen. The postoperative course was uneventful and the patient was discharged on the 4th postoperative day.

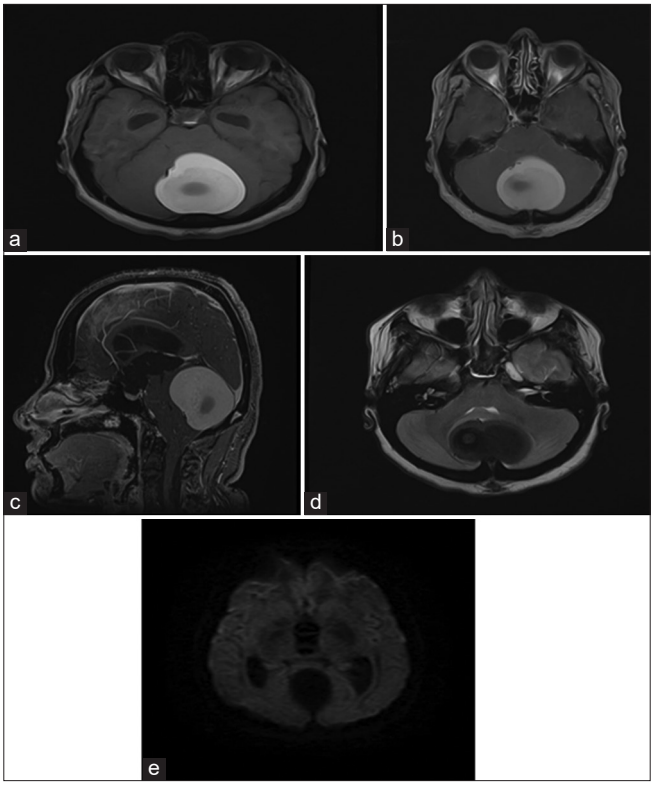

Figure 1: (a) T1-weighted sequence, (b) T1-weighted post contrast axial sequence, (c) T1 weighted post-contrast sagittal sequence, (d) T2 weighted sequence, (e) Diffusion weighted image.

She remained symptom-free on her initial follow-up visit but complained of headache and vomiting on her last follow-up (at 9 months). The MRI scan then showed postoperative changes with a mildly dilated fourth ventricle but no evidence of residual or recurrent disease. Our patient, however, did not follow-up further after this last visit.

DISCUSSION

The etiology and pathogenesis of mature teratoma are not yet understood,[4] but they are thought to arise from abnormally developed primitive streak around the 3rd–5th week of intrauterine life.[14] An overall male predominance is observed in teratomas,[7] but in the 12 previously reported cases of adult mature posterior fossa teratomas, the male-to-female ratio is almost 1:1, with a mean age of 47.7 years (range 19–70 years).[12]

The general presentation of teratomas is nonspecific and depends on the site and size of the lesion.[8] The mass effect is usually mild due to the slow-growing nature of these

tumors.[2] For posterior fossa teratomas, in particular, headache is the most common symptom (55.6%) followed by nausea/vomiting (50%) and gait disturbances (16.7%),[12] as was the case with our patient. Most mature posterior fossa teratomas have been reported in the cerebellar vermis or left hemisphere. Saura *et al.*[11] and Shin *et al.*[12] have each reported a mature teratoma in the right quadrigeminal cistern, whereas Bohra *et al*. have reported the only mature teratoma located in cisterna magna and ours is the second. A sinus tract leading up to the subcutaneous tissue was also found in our patient. These sinus tracts, though not a typical feature, have been found in several previous cases of teratomas.

Radiographic findings are widely inconsistent due to the varying nature of tissues, such as fat, calcification, hair follicles, and cellular debris.[8,12] Computed tomography scans usually demonstrate a cystic lesion of a nonhomogenous character.[3] MRI shows a single irregular, uni, or multilocular mass with mixed intensity signals.[8] Both T1- and T2-weighted sequences describe the internal architecture well and both low and high signals are seen on either sequence. T1 post contrast sequences may show heterogeneous, focal, or peripheral enhancement.[8] Perilesional edema is typically absent, like in the current case due to an intact capsular wall and undamaged blood-brain barrier.[8]

Diagnosis is difficult on radiography alone due to the varying nature of tissue elements; therefore, histopathologic confirmation of all three germ layer components is necessary.[12] Several previous reports have initially misdiagnosed mature teratomas of the posterior fossa as dermoids, epidermoids, or even meningiomas.

All 13 cases of mature posterior fossa teratoma [Table 1] as well as ours were treated with surgical resection alone. It is the recommended treatment for mature CNS teratomas, with the approach depending on the site of the lesion.[8] Radical resection is advocated, whenever possible, and is considered potentially curative due to the benign nature of mature teratomas.[2,4,8] Shin *et al.*[12] speculated that the extent of

Table 1: Literature review of mature posterior fossa teratoma in adults.

Study/Year	Age/sex	Location	Symptoms	Radiographic findings
Strang[13] (1960)	25/Female	Vermis and both hemispheres	Headache, nausea, atactic gait, diplopia (oculomotor palsy)	Not Available
Zavanone *et al.*[15] (2002)	50/Male	Vermis and left hemisphere	Headache, dizziness, nausea, vomiting	CT : heterogenous, calcification
Park *et al.*[5] (2007)	47/Female	Left hemisphere	Headache, nausea, dizziness	T1 low/T2 high/periphery enhancement/no calcification
Beschorner *et al.*[2] (2009)	66/Male	Vermis	Headache, nausea	T1 iso-high/T2 low/mild enhancement and calcification
Coulibaly *et al.*[4] (2012)	42/Female	Vermis and left hemisphere	Progressive headache, vomiting	T1 iso/T2/strong enhancement and calcification
Zhang *et al.*[16] (2012)	70/Female	Right cerebellopontine angle	Headache, vomiting, and gait disturbance	-
Bohara *et al.*[3] (2013)	41/Female	Cisterna magna, Confluence of sinuses around foramen magnum	Facial palsy, hearing loss, vertigo, and dizziness	T1 low to high
Sanyal *et al.*[9] (2013)	28/Male	Foramen magnum, cervicomedullary junction	Headache	T1 low
Saura *et al.*[11] (2014)	19/Female	Right quadrigeminal cistern	Yawning	T1 high/T2 high/no enhancement
Pöschl *et al.*[6] (2015)	59/Male	Midline	Evaluation for metastases	T2 iso
Sattar *et al.*[10] (2021)	24/Female	Right cerebellopontine angle	Cerebellar ataxia, nystagmus, sensorineural hearing loss, decreased palatal movements, decreased facial sensations	Ti high, T2 high, calcification, nodular enhancement
Shin *et al.*[12] (2021)	50/Male	Right quadrigeminal cistern	Headache, syncope	T1 low/T2 high/focal enhancement and calcification
Shin *et al.*[12] (2021)	60/Male	Vermis	Dysarthria, left hemiparesis	T1 high/T2 high/no enhancement and no calcification
Current case	26/Female	Cisterna Magna	Headache, vertigo, vomiting	T1 high/T2 low/no enhancement

CT: Computed tomography

resection (EOR) is directly related to survival. Resection is considered complete with >90% EOR[8] and incomplete if a macroscopic residual tumor is left or in the case of intraoperative tumor rupture.[2] The aim is to remove the entire tumor during the first surgery as revision surgeries are more technically challenging.[10]

Adjuvant therapies have no role as purely mature teratomas are resistant to chemoradiation,[8] though those that contain malignant or immature tissue may require adjuvant radiation or chemotherapy after surgery.[3] Levels of several tumor markers such as alpha-fetoprotein and beta-human choriogonadotropin are also raised in immature and malignant teratomas but mature teratomas lack these markers due to their benign nature and low mitotic activity.[1,8,12,]

Prognosis is good after complete surgical resection, especially in easily approachable tumors.[3] Previous literature describes the survival rate of mature teratomas to be 93–100% in 5 years and 93% in 10 years.[2] The relapse rates were 3–6% after complete resection and 39% after incomplete resection.[2] On rare occasions, a benign mature teratoma may have some immature or malignant components or may undergo a malignant transformation which leads to decreased survival rates.[2]

Out of 13 adult patients of mature posterior fossa teratoma, follow-up data were available for six patients, one of which died after 2 years of surgery, while the rest remained symptom-free or showed no recurrence on follow-up examinations up to 3 years. Our patient remained symptom-free on the initial follow-up visit but later complained of headache and vomiting on the 9 months follow-up. She, however, lost to follow-up after this last visit. Regular follow-up visits with clinical evaluation as well as MRI scanning are recommended since teratomas possess a risk of late recurrence.[4]

CONCLUSION

Mature teratomas of the posterior fossa in the adult age group have been rarely described in the literature. Thirteen cases have so far been reported, mostly in the cerebellum. We report the second case of mature teratoma located in cisterna magna in a young adult patient.

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.

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