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CASE REPORT

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Not all roses are sweet—Pediatric ependymoma

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Abstract

Ependymomas are rare malignant neoplasms that originate from radial glial cells within the central nervous system. Within pediatric central nervous tumors, ependymomas constitute the third most common entity with the majority occurring within the posterior fossa. Over the past decade, there have been monumental strides in classifying and grading central nervous tumors, specifically ependymomas. Revised classifications now identify ependymomas by anatomic location, histopathological and genetic subgroups with varying levels of symptom presentation and disease progression. Standard care of therapy remains surgical resection with post- operative radiotherapy.

KEYWORDS
brain tumor, emergency department, pediatric

1 | CASE

A 5-year-old female presented to the emergency department with a chief complaint of headache, vomiting, and abdominal pain. Vitals signs on arrival were blood pressure 115/72 mm mercury, heart rate 74 beats per minute, respiratory rate 23 breaths per minute, temperature 36.1°C, oxygen saturations 100% on room air, and weight 20.7 kg. Her abdominal pain had started earlier that day and was described as generalized, non-radiating, intermittent, and without aggravating or alleviating factors. She had been experiencing vomiting in the morning for approximately 1 week before developing abdominal pain and had been prescribed ondansetron from her primary care physician without relief of symptoms. She had been experiencing worsening headaches for approximately 1 week and described them as frontal, non-radiating, and not alleviated or exacerbated by anything.

Past medical history was significant for cyclic vomiting syndrome and constipation, for which the patient had been prescribed ondansetron and famotidine. The patient had an unremarkable birth history. The review of systems was as described and negative otherwise. The physical examination was significant for bilateral nystagmus. Laboratory results were unremarkable, other than positive for COVID-19. Computed tomography (CT) of the head revealed obstructive hydrocephalus secondary to a large hyperdense heterogenous 4 cm mass in the fourth ventricle, highly suggestive of an ependymoma (Figure 1). The patient was made nothing by mouth and started on D5W normal saline solution. Neurosurgery and pediatric critical care were consulted. The patient underwent suboccipital craniotomy for resection of tumor, and a follow-up lumbar puncture demonstrated no evidence of residual tumor cells.

2 | DISCUSSION

Ependymomas are rare malignant glial neoplasms that originate within the ependymal linings of the central nervous system (CNS). The incidence is higher within the pediatric population compared to the adult

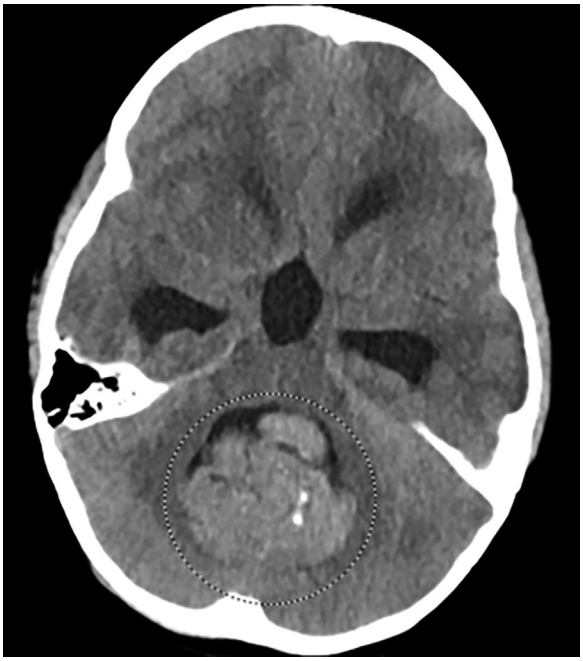

FIGURE 1 Computed tomography revealing obstructive hydrocephalus secondary to large hyperdense heterogenous 4 cm mass in the fourth ventricle indicating an ependymoma.

population. With an annual incidence of 0.26 per 100,000 children, ependymomas account for 10% of the total pediatric CNS tumor burden with a high mortality rate of 45%.[1,2] The clinical course is variable, with time to initial recognition and early treatment being a crucial prognostic factor.[1]

Posterior fossa ependymomas present with symptoms of increased intracranial pressure such as nausea and vomiting, secondary to obstructive hydrocephalus, ataxia, and cranial nerve palsies whereas supratentorial ependymomas present with headaches, seizures, and focal neurological signs secondary to cortical involvement.[3,4] Because of nonspecific and variable presentations, especially in young children, the chance of missing the diagnosis can be high and emergency physicians need a high degree of clinical acumen in making the diagnosis. Initial imaging in the ED can be performed using the CT with definitive diagnosis carried out using magnetic resonance imaging (MRI) showing T1 hypointensity and T2 hyperintensity with heterogenous enhancement on T1 sequences post-gadolinium.[3] Given that diagnosis is largely based on physical exam findings of increased intracranial pressure and imaging, there are no standard laboratory markers used routinely in diagnosis of this condition. Treatment of ependymomas is surgical resection and radiotherapy.[2,3,5] In patients with complete resection followed by radiotherapy, the overall survival rate can vary between 67%–93% with a progression-free survival rate of 51%–82% whereas in patients with incomplete resections, the survival rate decreases to 22%–52% and progression free survival rate to 0%–41%.[4]

3 | CONCLUSION

Ependymomas constitute a rare subgroup of malignant central nervous system neoplasms among the pediatric population. Primary evaluation includes imaging with MRI and treatment with gross total resection and radiotherapy. Improved awareness about the presentation and management of these tumors can assist emergency physicians in making the diagnosis, facilitate early detection, and improve patient outcomes.

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