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LETTER TO THE EDITOR

Ectopic schwannoma of the lateral ventricle: case report and review of the literature

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Introduction

Schwannomas, which originate from Schwann cells, are mostly benign entities, accounting for approximately 8% of the tumors of the central nervous system (CNS), with acoustic neuroma (vestibular schwannoma) being the most common. Less than 1% of schwannomas occur in the brain parenchyma or ventricle [1, 2]. The nerve fibers in the CNS are not wrapped in Schwann cells. Consequently, schwannomas that occur in the parenchyma or ventricle of the brain are also known as ectopic schwannomas (ES) [3]. ES of the lateral ventricle are very rare. Since the first report describing ES of the lateral ventricle by David in 1965 [4], there have been only 21 cases reported in the English literature. The ages of the patients in these cases were between 8 and 68 years, with a median age of 21 years. Only one case involved malignant biological behavior in the literature [5], with metastasis to the cerebellar angle and cerebellum occurring seven months after the operation. We report the case of a woman with ES in the right lateral ventricle who experienced dizziness and headache for two years, with exacerbation of the symptoms over one month. The present study aimed to review the relevant literature of all ES of the lateral ventricle, and to summarize and analyze the clinical manifestation, pathological and magnetic resonance imaging (MRI) features, to improve the understanding of ES of the lateral ventricle.

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Case presentation

A 51-year-old woman, with no significant medical history, experienced intermittent dizziness and headache during the previous two years, which had worsened in the past month. The patient experienced no significant weight loss or other symptoms of discomfort; furthermore, laboratory tests revealed no obvious abnormalities. Contrast-enhanced MRI revealed that the lesion, measuring approximately 3.5 cm × 4.3 cm × 2.9 cm in size, was located in the right lateral ventricle. The solid components of the tumor exhibited equal signal on T1-weighted imaging (T1WI) and slightly lower signal on T2-weighted imaging (T2WI) (Fig. 1a, b). Cystic components with low signal on T1WI and high signal on T2WI were apparent in the center of this lesion. Diffusion-weighted imaging and apparent diffusion coefficient images demonstrated a slightly higher signal. After contrast-enhanced T1WI, the solid part of the lesion was inhomogeneously enhanced (Fig. 1c, d). The patient then underwent resection of the lesion in the lateral ventricle. During the operation, the tumor, measuring approximately 4.0 cm × 4.0 cm × 3.5 cm in size, was found to be located in the right lateral ventricle and exhibited reddish color, irregular shape, fragile texture, and normal in terms of blood supply. On pathological analysis, the tumor was composed of bundle-shaped and braided spindle-shaped cells in a biphasical pattern, some tumor cells grew densely and arranged closely, forming a palisade structure and the Antoni A region, and some tumor cells were sparsely arranged with mucus and loose edema area, forming Antoni B region (Fig. 1e). On immunohistochemical staining, the tumor cells were positive for S-100 (Fig. 1f), vimentin and Ki-67 (< 3%), and were negative for glial fibrillary acidic protein (GFAP), cytokeratin (CK), and epithelial membrane antigen (EMA). Based on the pathological features, a diagnosis of ES of the lateral ventricle was established. The course of

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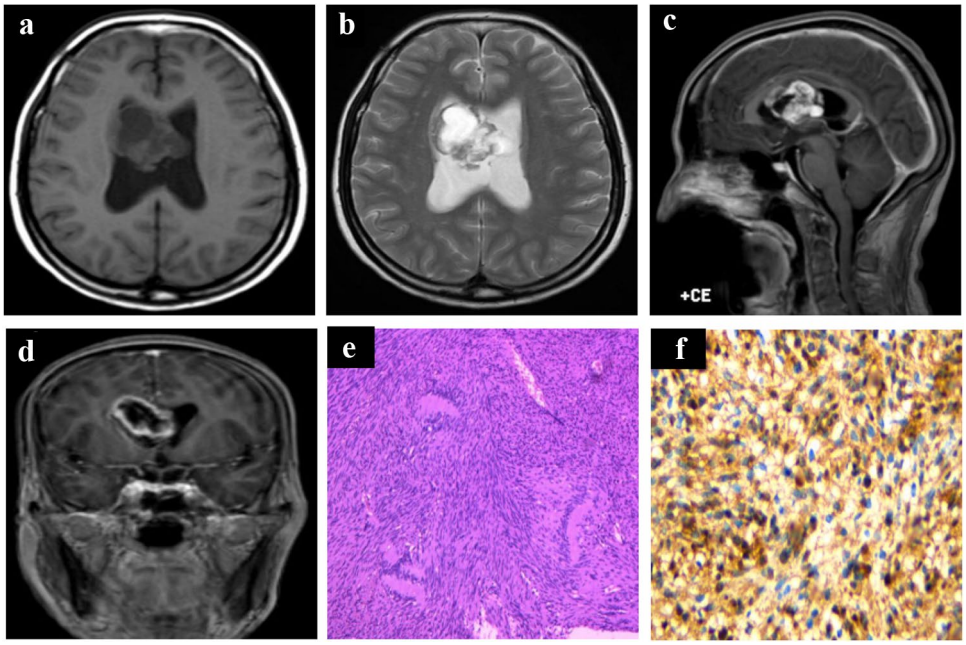

Fig. 1 A 51-year-old woman with ES of the right lateral ventricle. **a**, **b** Axial T1WI and T2WI uncontrast enhanced scan showed an approximately 3.5 cm × 4.3 cm × 2.9 cm irregular cystic-solid lesion in the right lateral ventricle, and the solid components exhibited equal signal on T1WI and slightly low signal on T2WI, cystic components with low signal on T1WI and high signal on T2WI are apparent in the center, mild edema is apparent near the body of the lateral ventricle. **c**, **d** Sagittal and coronal contrast-enhanced T1WI showed heterogeneous and obvious enhancement of the solid part of the mass, with no significant enhancement of cystic components. **e** The tumor is composed of bundle-shaped and braided spindle-shaped cells in a biphasical pattern, Antoni A region and Antoni B region can be seen (H&E × 100). **f** Immunohistochemical analysis of the tumor showed diffuse positive staining of S-100 (×400)

the disease was smooth, and there was no recurrence after 12 months of follow-up.

Discussion

Schwannomas are benign tumors originating from nerve sheath cells, which are common in the head, neck, and extremities. Intracranial schwannomas account for approximately 8% of CNS tumors. Less than 1% of these tumors occur in the cerebral parenchyma or ventricle [1, 2]. Generally, only after the cerebral nerve passes through the pia meninges are Schwann cells found. There are no Schwann cells in the nerve fibers of the CNS, consequently, schwannomas that occur in the parenchyma or ventricle of the brain are often known as ES. Although the origin of ES remains controversial [6], three main theories have been proposed, (a) misplaced neural crest remnants, (b) differentiation of multiple mesenchymal progenitor cells and (c) Schwann cells of the surrounding perivascular nerves undergo neoplastic transformation [6, 7]. Moreover, some studies [8] have demonstrated the presence of nerve tissues in the choroid plexus, cerebral vessels, and subependymal zone, inferring that Schwann cells may be distributed around these nerve axons. Accordingly, we speculate that ES of the lateral ventricle may be derived from Schwann cells around the perivascular nerve plexus in the choroid plexus or ependyma. Susceptibility to cystic degeneration is the main feature of the disease [6, 7, 9], which may be caused by secondary hemorrhage or necrosis in the tumor. Peritumoral edema is another major feature of ES. Feigen et al. [10] attributed this to the degeneration of white matter or proliferation of glial cells around the tumor tissue. However, some investigators believe that there is a certain correlation between this and the expression of vascular endothelial growth factor.

Immunohistochemical staining is very important in the diagnosis of ES. The positive expression of S-100 and vimentin is characteristic, while the expression of GFAP, EMA, and CK is often negative [7, 9]. Microscopically, the tumor cells are often arranged in fusiform and palisade patterns, and typical Antoni A and Antoni B regions are observed. In 22 cases of lateral ventricular ES [Table 1], the average age at onset was 28.33 years, with the youngest at 8 years [11] and the oldest at 68 years [7]. There were 6 females and 16 males. Four cases of ES were located in the left lateral ventricle, 18 cases in the right lateral ventricle. Therefore, we speculate that the disease is more common in the right lateral ventricle, and more common in males than in females. The clinical manifestations of the disease are not specific. Most patients experience dizziness and headache

Table 1 Ectopic schwannoma of the lateral ventricle reported in the English literature

No.	Year	Author/References	Age	Gender	Located in the lateral ventricle (left/right)	Clinical symptoms	Follow-up
1	1965	David et al. [4]	15	M	R	Headache,vomiting,left hemiplegia	No recurrence
2	1975	Ghatak et al. [15]	63	F	R	Eilepsy, left hemiplegia	No recurrence
3	1975	Van Rensburg et al. [16]	21	M	R	Epilepsy	No recurrence
4	1988	Pimentel et al. [11]	8	M	R	Headache, vomiting, hemiplegia	No recurrence
5	1990	Ost and Meyer [17]	44	M	L	Right hemianopia	Missing
6	2001	Barbosa et al. [18]	13	F	R	Headache	No recurrence
7	2003	Erdogan et al. [19]	21	M	R	Left eye vision loss	No recurrence
8	2004	Dow et al. [20]	16	M	R	Asymptomatic papilledema	No recurrence
9	2007	Leveque et al. [21]	16	M	R	Epilepsy	No recurrence
10	2008	Benedict et al. [22]	15	M	R	Headache	No recurrence
11	2009	Vasconcellos et al. [23]	21	F	L	Headache	No recurrence
12	2013	Luo et al. [6]	24	M	R	No	No recurrence
13	2013	Jaimovich et al. [24]	16	M	R	No	No recurrence
14	2013	Alberione et al. [25]	41	F	R	Headache, nausea	No recurrence
15	2015	Glikstein et al. [12]	34	M	L	Headache	No recurrence
16	2015	Curran-Melendez et al. [26]	20	M	R	Epilepsy, lower limb weakness	No recurrence
17	2016	Abdolhosseinpour et al. [9]	9	M	R	No	No recurrence
18	2016	Salazar et al. [27]	16	M	L	Headache, blindness in the left eye	No recurrence
19	2017	O'Reilly et al. [28]	52	M	R	Headache	No recurrence
20	2018	Kouitcheu et al. [7]	68	F	R	Headache, dizziness, left hemianopia	No recurrence
21	2020	Chiba et al. [29]	26	M	R	Headache and left hemianopsia	No recurrence
22	2020	Our case	51	F	R	Headache	No recurrence

as the main manifestations, and some can exhibit signs of epilepsy, hemiplegia, fatigue, nausea, hemi-blindness, and other symptoms. The patient described in the present case was a 51-year-old woman who had a lesion located in the right lateral ventricle. Postoperative pathology revealed staggered Antoni A and Antoni B regions, with positive immunohistochemical staining for S-100 and vimentin.

In terms of imaging, the MRI features of ES of the lateral ventricle have specific characteristics. Based on the present case and the relevant literature, these findings are summarized as follows. Cystic changes: cystic and solid changes are a characteristic manifestation of the disease. The cystic part is often characterized by a low signal on T1WI and high signal on T2WI, while the solid part often exhibits slightly low signal on T1WI and high signal on T2WI, which corresponds to the Antoni A and Antoni B regions on microscopy, respectively. After a contrast-enhanced scan, the solid component is clearly enhanced, but the cystic part is not. Edema: peritumoral edema is believed to be a characteristic manifestation of benign schwannoma. ES exhibits varying degrees of peritumor edema [12]. Calcification: it has been reported that part of the lateral ventricle ES may appear calcified [13], which, to a certain extent, can serve as providing the basis for the diagnosis of the tumor. The patient in the present case exhibited cystic and solid changes, and mild edema was apparent around the lateral ventricle, which is consistent with features reported in the literature.

Surgical resection is the best therapeutic modality for lateral ventricular ES. Oertel et al. [14] reported that no recurrence was found in the longest follow-up period (10 years). The total resection rate of the disease is high, with fewer postoperative complications, and no need for radiotherapy or chemotherapy. Moreover, recurrence is rare, and the prognosis is good.

Conclusion

We describe the case of 51-year-old woman with ES of the right lateral ventricle. Based on our results and the literature review, clinical manifestation, pathology, and MRI features of ES of the lateral ventricle are summarized as follows. First, the incidence of ES among adolescents is higher in males than in females, and most tumors are located in the right lateral ventricle. Second, cystic and solid changes are a characteristic manifestation of the disease. Some tumors can also appear calcified; contrast-enhanced scanning reveals obvious enhancement of solid components; however, cystic components are not enhanced. Third, varying degrees of edema are apparent around the tumor. According to these

characteristics, a clinical diagnosis can be improved for reference.

Author contributions JZ contributed to conception and design. XL contributed to the literature search and writing of the initial manuscript. All authors contributed to data analysis and interpretation, and gave suggestions for writing.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This article does not contain studies with animals by the author.

Informed consent Informed consent was obtained from all individual participants included in the study.

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