Radiology Case Reports 18 (2023) 1058–1064

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Case Report

# Tentorial ectopic schwannoma: A case report and literature review ☆,☆☆

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ARTICLE INFO

*Article history:*
Received 16 November 2022
Revised 4 December 2022
Accepted 6 December 2022

*Keywords:*
Schwannoma
Tentorium
Meningioma
Magnetic resonance imaging

ABSTRACT

Schwannomas arising within the central nervous system unrelated to a major cranial nerve rarely originate from the dura, particularly the tentorium. Hereby, the reported case is a 39-year-old female with blurred vision in her right eye for 1 month who was diagnosed with tentorial ectopic schwannoma. The tumor was almost totally resected through a posterior median incision approach without any neurological deficits. The clinical presentations and radiographic findings of such cases are discussed in the article.



## Introduction

Schwannomas originating from Schwann cells of the neurilemma account for about 8% of all primary intracranial tumors [1]. Approximately 90% of schwannomas are located in the cerebellopontine angle, most closely associated with the eighth cranial nerve. It is reported that intracranial schwannomas rarely originate from atypical sites, such as brain parenchyma, ventricles, sellae, or tuberculum sellae [2–4], especially dura mater (including falx cerebellum, tentorium, etc.). Herein, we presented a rare case of tentorial ectopic schwannoma (TES) that mimicked meningioma and the previously valuable literature was simultaneously reviewed.

## Case report

A 39-year-old woman was admitted with a chief complaint of blurred vision in her right eye for 1 month. No positive signs

☆ Funding: This study has received funding by the National Natural Science Foundation of China (U21A20386, 81971593 to Hui Zhang).
☆☆ Competing Interests: The authors declare that they have no conflict of interest.
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https://doi.org/10.1016/j.radcr.2022.12.012

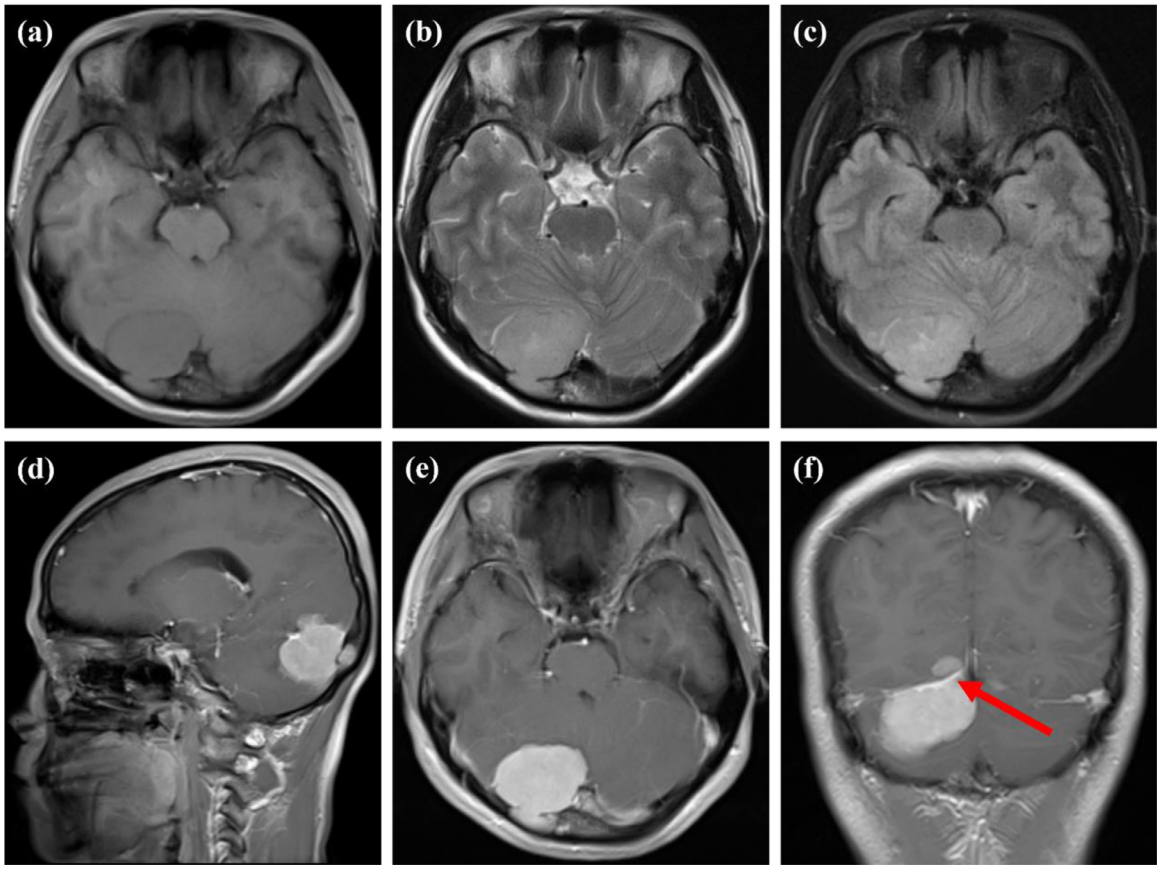


**Fig. 1 – MRI showed that there was a solid mass in the right occipital cerebellar tentorium. The lesion showed isointense on T1WI and slightly hyperintense on T2WI, and relatively homogenous on FLAIR, respectively (a-c). The mass exhibited homogeneous enhancement, with the dural tail sign (arrow) (d-f).**

were found during the neurological examination on admission. There was no history of neurofibromatosis.

Magnetic resonance imaging (MRI) revealed a round solid mass (3.6 × 2.9 × 3.7 cm) with well-defined borders and a well-distributed signal in the occipital and cerebellar regions. The lesion appeared isointense on T1-weighted imaging and slightly hyperintense on T2-weighted imaging, and relatively homogenous on fluid-attenuated inversion recovery (FLAIR) sequences, respectively. On contrast-enhanced T1-weighted imaging, the mass exhibited homogeneous enhancement, accompanied by enhancement and thickening of the neighboring dura (Figs. 1a-f). Computed tomography angiography (CTA) of the head and neck revealed an occipital transtentorial lesion. The lesion is close to the straight sinus, the confluence of sinus, the right transverse sinus, and the sigmoid sinus, which made the sinus cavity compressed and narrower. Moreover, similar findings were observed for magnetic resonance venography (MRV) as CTA (Figs. 2a and b). To sum up, the diagnosis based on preoperative imaging tended to be meningioma.

In preoperative evaluation, tumor involving the cerebral venous sinus represented a challenge for complete resection of the tumor. It may cause intraoperative bleeding and postoperative cerebral infarction as a result of venous sinus occlusion. Tumor resection and venous sinus reconstruction were performed under general anesthesia using a posterior median incision approach. A firm and gray-white mass measuring 4 × 3 cm was exposed after we enlarged the posterior border of the foramen magnum with rongeurs and suspended the dura. The adjacent bone flap and part of the dura mater had been eroded. The tumor base was firmly attached to and eroded the right transverse sinus, which results in excessive intraoperative bleeding. The lesion is partially convex to supratentorial. Counterclockwise cutting was performed along the tumor base until the whole tumor was completely removed. Besides, no definite nerve could be identified in relationship to the tumor. Postoperative MRI revealed no residual tumor (Figs. 3a-c).

The postoperative course was uneventful, and the patient had no neurological deficits at discharge. Histological examination revealed spindled tumor cells arranged in fascicles and diffuse S-100, SOX-10, and Vimentin expression (Figs. 4a-d). These findings were consistent with schwannoma.

## Discussion

Intracranial schwannoma not related to cranial nerves is called "intracranial ectopic schwannomas" (IES) and represents less than 1% of intracranial schwannoma [5]. Among them, dural-based IES, especially TES, is extremely rare. Since

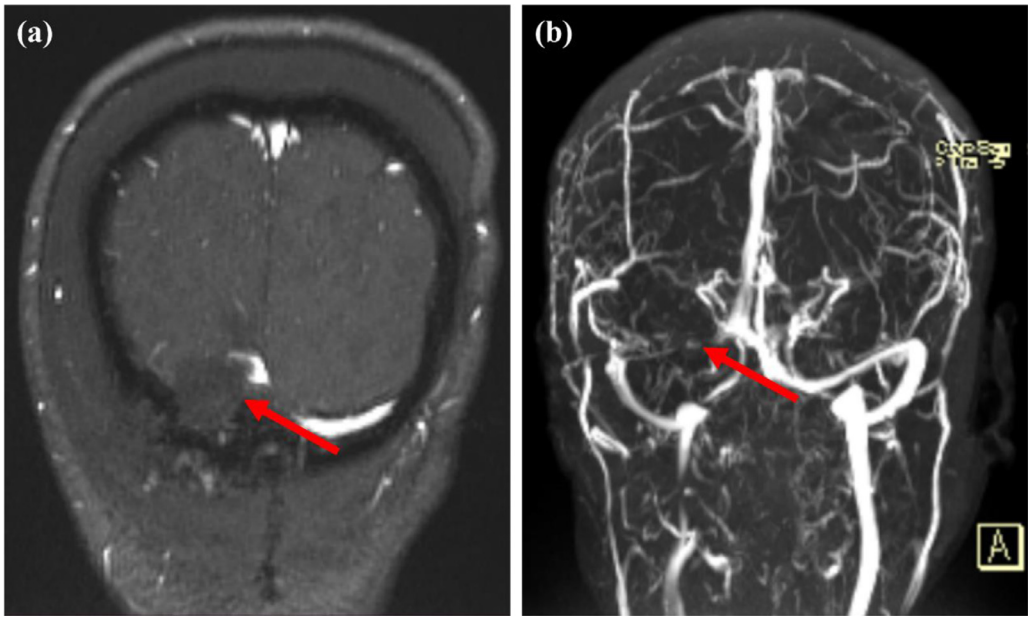


**Fig. 2 – The transverse sinus cavity was compressed and narrower (arrows) (a, b).**

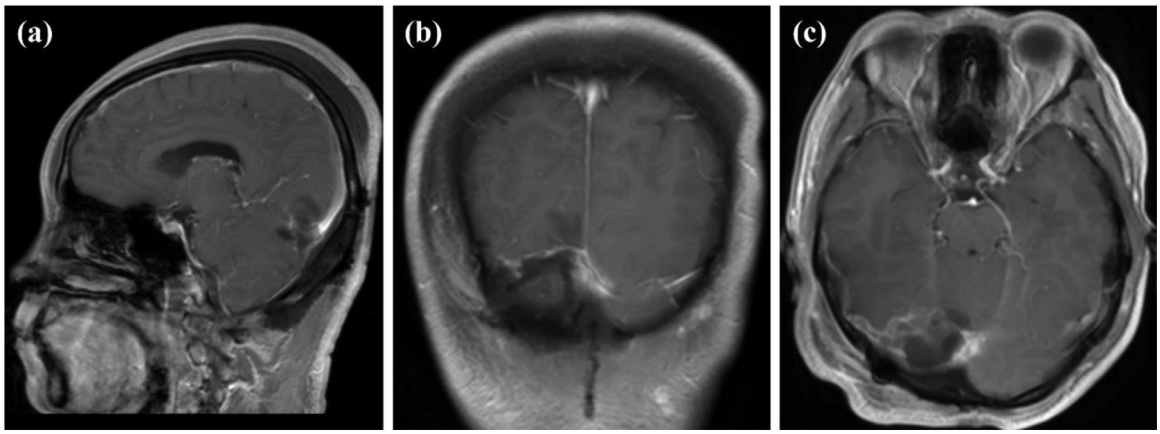


**Fig. 3 – Postoperative MRI revealed no residual tumor (a-c).**

the first case was reported by Flickinger et al. in 1988 [6], we collated only 18 patients with TES all over the world, including this case (Table 1) [1,5–20]. It is popular to occur in children and young adults, and no significant differences are found between genders [5]. The clinical symptoms are nonspecific. Reported symptoms included headaches, focal neurologic deficits, and seizures. Its hereditary mutation appears to associate with neurofibromatosis type 2 (NF2) [9]. In these cases, the mean age of patients was 37.83 years (range, 9-75 years). Eleven patients presented with headaches, and 4 patients presented with dizziness. Less frequent clinical symptoms were as follows: truncal ataxia, physical or facial paresthesia, nystagmus, transient diplopia or a progressive visual impairment (including the present case), vomiting, sixth or seventh cranial nerve paresis, transient global amnesia, and 10 patients had normal neurological conditions. No previous cases were associated with NF2.

All but one of the patients had been investigated with MRI. Typically, IES usually present as a mixed cystic-solid lesion, with heterogeneous enhancement by gadolinium. Retrospective review of previous cases of tentorial schwannoma (Table 1), 13 cases were cystic and solid mass lesions, 4 cases of previous cases and this case were solid homogeneous mass lesions. Intratumoral cysts and calcifications are hallmarks of ectopic schwannomas [12]. We found that cystic degeneration of TES was also common. Calcification was observed in only 2 cases, which might be attributed to the absence of CT examination in some patients. In addition, mild-to-moderate peritumoral edema and bleeding can be observed in some lesions, but the incidence of them is low. Only 3 cases had peritumoral edema, and intratumoral hemorrhage occurred in only 1 patient. The location of the tumor in most of these patients was infratentorial. However, 2 patients had supratentorial growth patterns, and 7 patients (including this case) had

**Table 1 – Summary of the reported cases of TES.**

| Ref. | Age/sex | Clinic | Site and growth pattern | MRI imaging findings | | | | Other imaging findings | Surgery | Results |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | | Appearance | Enhancement | DTS | Peritumoral edema | | | |
| [7] | 64/F | Transient global amnesia | Right tentorium, supratentorial | Cystic | Heterogeneously enhancing mass | - | + | | Subtemporal approach | Complete recovery |
| [8] | 60/F | Long-lasting headaches | Left tentorium, supra- and infratentorial | Solid | Homogeneously enhancing mass | + | - | | Suboccipital approach | Complete recovery |
| [1] | 23/M | Intermittent dizziness and difficulty swallowing | Right tentorium, infratentorial | Cystic and Solid | Heterogeneously enhancing mass | + | - | | Petrosal craniotomy with retrolabyrynthine bony removal | Complete recovery |
| [9] | 42/M | Paresthesia and weakness of the 4 extremities | Left CPA, infratentorial | Cystic and Solid | Homogeneously enhancing mass | + | - | | Retrosigmoid approach | Complete recovery |
| [10] | 17/F | Headaches, intermittent diplopia | Left tentorium, supra-and infratentorial | Cystic | Heterogeneously enhancing mass | - | - | | Orbitozygomatic pterional craniotomy | Complete recovery, MRI: no residual tumor |
| [6] | 22/M | Headaches | Tentorium, N/A | Solid | Homogeneously enhancing mass | N/A | N/A | Calcification | N/A | N/A |
| [11] | 9/F | Dizziness, headaches, vomiting, and gait disturbance | Right tentorium, infratentorial | Solid | Homogeneously enhancing mass | + | - | Supratentorial hydrocephalus | Suboccipital craniotomy | Complete recovery, MRI: no residual tumor |
| [5] | 75/M | Headaches, visual impairment | Left medial occipital, supra-and infratentorial | Cystic | Strong peripheral and intralesional multiseptate contrast enhancement | + | + | Intratumoral hemorrhage | Occipital paramedian left craniotomy | Complete recovery, MRI: no residual tumor (3 months) |
| [12] | 49/F | Headaches, neck pain, vomiting, and unsteadiness of gait | Right CPA, infratentorial | Cystic and Solid | Heterogeneously enhancing mass | + | - | | Retrosigmoid suboccipital craniotomy | Progressing well, residual right mandible paraesthesia (6 weeks) |
| [13] | 20/M | Headaches | Right tentorium, infratentorial | Solid | Homogeneously enhancing mass | + | - | | Extreme lateral supracerebellar infratentorial approach | Without any neuro-logical deficit (6 months) |

(*continued on next page*)

**Table 1 (*continued*)**

| Ref. | Age/sex | Clinic | Site and growth pattern | MRI imaging findings | | | | Other imaging findings | Surgery | Results |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | | Appearance | Enhancement | DTS | Peritumoral edema | | | |
| [14] | 21/M | Generalized headaches and dizziness | Right tentorium, supra- and infratentorial | Cystic | Heterogeneously enhancing mass with internal septan | + | - | Multiple fluid levels | Combined occipital and paramedian suboccipital | Complete recovery |
| [15] | 37/M | Memory and gait disturbance | Right tentorium, supra- and infratentorial | Cystic | Heterogeneously enhancing mass with internal septa | + | - | | Occipital transtentorial approach | Complete recovery |
| [16] | 29/M | Headaches, transient diplopia | Right tentorium, supra- and infratentorial | Cystic and Solid | Heterogeneously enhancing mass | + | - | | Transpetrosal approach | Right trochlear nerve palsy (2 months) |
| [17] | 41/F | Headaches, positional vertigo, and truncal ataxia | Left anteromedial cerebellar regi-on,infratentorial | Cystic and Solid | Heterogeneously enhancing mass | + | - | | Retrosigmoid craniotomy | Complete recovery, MRI: no residual tumor(4 months) |
| [18] | 58/F | Headaches | Left CPA, infratentorial | Cystic and Solid | Heterogeneously enhancing mass | + | + | | Lateral suboccipital retrosigmoid craniotomy | Complete recovery MRI: no residual tumor (2 years) |
| [19] | 48/M | Gait disturbance and papilledema | Left tentorium, infratentorial | Cystic and Solid | Heterogeneously enhancing mass | - | - | Hydrocephalus, calcification | Lateral suboccipital approach | Complete recovery, MRI: no residual tumor (12 months) |
| [20] | 27/M | N/A | Occiput, supratentorial | N/A | N/A | N/A | N/A | CT: Heterogeneously enhancing cystic and solid mass, erosion of skull bone | N/A | N/A |
| Present case | 39/F | Blurred vision in the right eye | Right tentorium, supra- and infratentorial | Solid | Homogeneously enhancing mass | + | - | Erosion of skull bone and venous sinus | Posterior median incision approach | Without any neurological deficit |

Ref., Reference; M, male; F, female; DTS, Dural tail Sign; N/A, not available; +/-: presence/absence of dural tail on MRI; CPA, cerebellopontine angle.

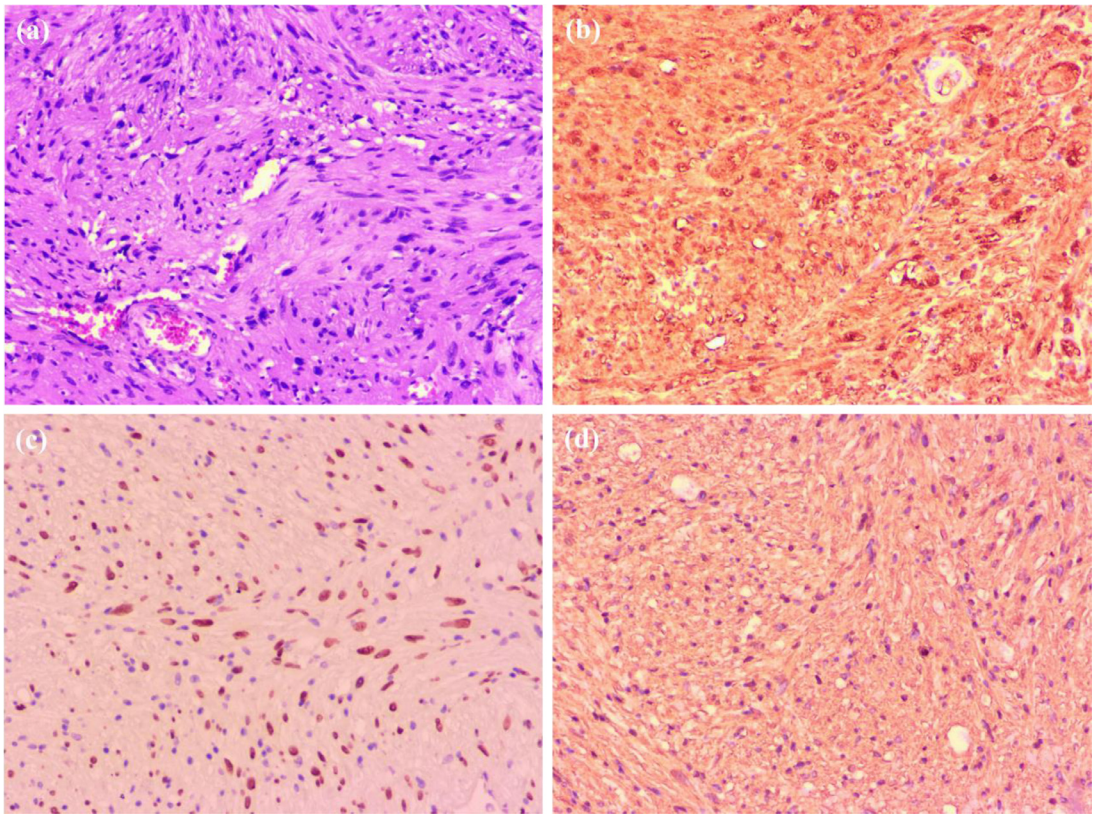


**Fig. 4 – Photomicrographs of surgical specimens. (a) Photomicrographs showing tissue formed by compact neoplastic proliferation of spindle-shaped cells (H&E; original magnification ×100). (b) Photomicrograph showing the tumor with strong positivity on immunostaining against S100 protein (original magnification ×100). (c) Tumor cells were positive for SOX 10 (original magnification ×100). (d) Tumor cells were positive for Vimentin (Vim, original magnification ×100).**

supra-and infratentorial growth patterns. Like our case, the dural tail sign was another important feature of these tumors (13/18), somewhat mimicking meningioma. The existence of this sign is extremely misleading. It is not specific and can be seen in meningiomas, hemangiopericytomas, schwannomas, aneurysms, and even intracranial infections [21]. Therefore, ESC should be considered in the differential diagnosis of tumors based on the tentorium of the cerebellum. It is more difficult to distinguish ectopic tentorial schwannoma with uniform enhancement from meningioma. The differentiating feature of this case may be a slightly higher signal than meningioma on T2-weighted sequences and invasion of the venous sinus and skull bone, whereas bony hyperostosis is associated with meningioma.

Several surgical approaches have been proposed to resect this kind of tumor, depending on the size and site of the tumor (Table 1). All these tumors were treated by total or subtotal excision. Furthermore, the results from long-term follow-up confirmed that good prognosis was maintained among patients.

To facilitate understanding and recognition of these rare tumors, Haga et al. [22] proposed a location-based classification system for schwannomas in the brain, which is divided into intra-axial, periventricular, dural-attached, and other types. Different subtypes may have different origins. Intra-axial schwannoma might be derived from multipotential mesenchymal cells of the neural crest [7]. Periventricular schwannomas arise from the autonomic nerves of the choroid plexus. There are several hypotheses on the origin of dural-attached schwannomas. The Arnold tentorial nerve is a meningeal branch of the ophthalmic branch of the trigeminal nerve (V1), which innervates the dura mater of the tentorial and parieto-occipital regions, the posterior third of the cerebral falx, the superior sagittal sinus and the transverse sinus, as reported previously [23]. The branches of the first 3 cervical nerves provide most of the innervation to the dura mater of the posterior fossa [24]. Besides, the same is true of the hypoglossal nerve, recurrent branch of the vagus nerve, facial nerve, and glossopharyngeal nerve [23]. Therefore, they may originate from the meningeal branches of the above nerves. The second possibility is that they derive from the perivascular nerve plexuses surrounding arteries within the subarachnoid space. Furthermore, given the histological similarity between neuroectodermal Schwann cells and mesodermal leptomeningeal cells [25], dural-based schwannomas may also arise from the transformation of Schwann cells from the pia mater. We surmised that the most reasonable explanation is Schwann cells originating from meningeal branch nerves in the present case, although its true origin is still unclear.

## Conclusion

TES is an extremely rare condition, although the surgical indications may not be modified from the surgical indications for other benign tumors such as meningioma, it should be regarded as the differential diagnosis of a mixed cystic and solid or even solid mass with a tentorial dural tail sign near the mass.

## Research involving human and/or animal participants

This article does not contain any studies with human participants or animals performed by any of the authors.

## Author contributions

All authors contributed to the article. The first draft of the manuscript was written by Hui Yan and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

## Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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