Recurrent painful ophthalmoplegic neuropathy revealing oculomotor nerve schwannoma

Neuropathie ophtalmoplégique douloureuse récurrente révélant un schwannome du nerf oculomoteur commun

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RÉSUMÉ

L'ophtalmoplégie douloureuse est un motif fréquent de consultation aux urgences de Neuro-ophtalmologie. Nous rapportons une observation d'une adolescente de 18 ans qui a présenté quatre épisodes d'ophtalmoplégie douloureuse, mimant une «migraine ophtalmoplégique», révélant un schwannome du nerf oculomoteur commun gauche. Un an après le dernier
épisode, l'examen neurologique a été normal. L'IRM cérébrale avec des coupes centrées sur le nerf oculomoteur commun a montré une prise de constraste persistante de la troisième paire crânienne gauche évoquant un schwannome. Ce mode de révélation de la lésion tumorale, comme chez notre patiente, est exceptionnel et mime une «migraine ophtalmoplégique».

Typiquement, le schwannome du nerf oculomoteur se manifeste par un tableau progressif. Ainsi, une ophtalmoplégie douloureuse avec des céphalées traînantes et une prise de contraste persistante à l'IRM cérébrale doit faire évoquer une lésion tumorale.

Mot clés : migraine ophtalmoplégique, Schwanomme, nerf oculomoteur commun, Neuropathie ophtalmoplégique douloureuse récurrente

SUMMARY

Painful ophthalmoplegia is a common presenting symptom in neuro-ophthalmology emergencies. We report an unusual case of a recurrent painful ophthalmoplegia due to a third nerve schwannoma mimicking « ophthalmoplegic migraine ». A 18 year-old girl had presented 4 episodes of left eye painful ophthalmoplegia respectively in 8, 13, 16 and 17 years old. One year after the last episode, neurological examination was normal. Brain MRI focused on the oculomotor nerve showed an enhancing nodular lesion suggesting a third nerve schwannoma. Thus, recurrent painful ophthalmoplegia revealing oculomotor nerve schwannoma, as described in our case, is exceptional. To our knowledge, only thirteen cases have been reported in the literature. Third nerve schwannoma is a rare cranial nerve tumor, typically revealed by progressive palsy of the oculomotor nerve. Recurrent painful ophthalmoplegia with persistent headache and enhancement in brain imaging should suggest tumoral lesions.

Key words: recurrent painful ophthalmoplegia, schwannoma, ophthalmoplegic migraine, oculomotor nerve

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INTRODUCTION

Recurrent painful ophthalmoplegia is a common clinical situation. Vascular causes and recurrent painful ophthalmoplegic neuropathy (RPON) previously known as « ophthalmoplegic migraine » are the main causes. Schwannomas of the cranial nerves could be a cause of recurrent painful ophthalmoplegia. They are infrequent tumors, comprising only 8% of primary intracranial tumors and most commonly affecting the vestibular and trigeminal nerves [1]. Schwannomas of the third nerve are rare and typically present with gradually progressive painful nerve palsy. Fluctuating symptoms have been rarely reported and are suggestive of RPON leading to frequent misdiagnosis [1]. We report a newly diagnosed patient with oculomotor nerve schwannoma who presented as RPON. We also review the literature on oculomotor cranial nerve schwannomas with recurrent clinical presentation and discuss their clinical features and management.

CASE PRESENTATION

A 18-year-old girl, born to first consanguineous parents, presented to our department with ten-year history of left recurrent migrainous headaches associated with ipsilateral intermittent oculomotor nerve palsy. She had a family history of dementia and hypertension in her grandfather and no personal medical history. At the age 8, she presented with continuous left orbito frontal throbbing headache with nausea, vomiting and photophobia. Within 10 days, when headache improved, ptosis and left monocular horizontal diplopia appeared for 45 days. This episode relapsed three times at 13, 16 and 17 years old. Ophthalmoplegia always resolved after roughly one month without particular treatments. However, she kept an increasingly frequent and disabling headache without ophthalmoplegia lasting about 12 hours and occurring twice a week. At 18 years old, neurological examination was normal. Brain MRI, performed one year from the last attack, revealed an enhancing nodule on the cisternal segment of the left oculomotor nerve suggestive of schwannoma (Fig.1). The patient was kept on symptomatic treatment based on non-steroidal anti-inflammatory drugs and anlagesic if headache. No surgical intervention was attempted. On follow-up, she no longer presented additional episodes.

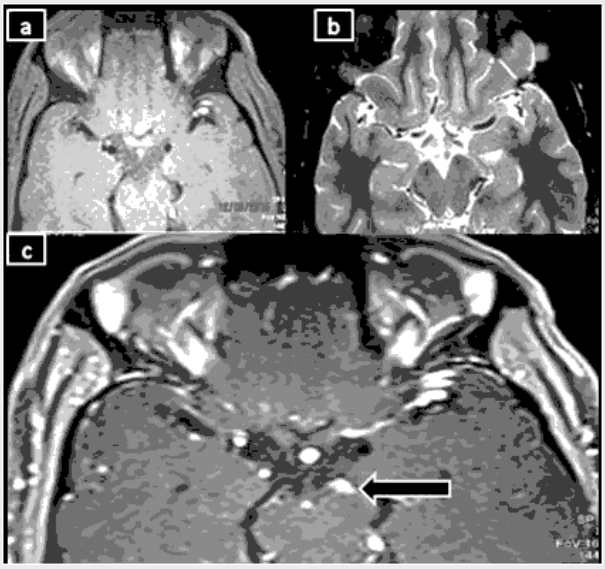

Figure 1. Axial non-enhanced T1-weighted (a), T2-weighted (b) and Gadolinium-enhanced T1-weighted (c) images corresponding to a schwannoma of cisternal segment of third nerve

DISCUSSION

In this article, we report a new case of oculomotor nerve schwannoma that mimicked RPON. Schwannomas are slowly growing peripheral nerve tumors that arise from the Schwann cell layer of cranial nerves. They affects typically vestibular branch of the eighth nerve and less commonly the fifth nerve, the seventh nerve, the fourth nerve and exceptionally the third nerve [2]. From 1975 to 2017, fewer than 107 cases of these oculomotor tumours have been reported across 70 published studies [2]. Moreover, isolated schwannoma of the oculomotor nerve without an associated type II neurofibromatosis, as reported in our case, are rare [2,3]. Intracranial schwannomas typically present with gradually progressive symptoms. Relapsing clinical course had been exceptionally reported and shows recurrent cranial nerve symptoms with migrainous headache [4]. Ptosis and diplopia were associated or preceded by headache, often with migrainous features leading to a first diagnosis of RPON [5]. To our knowledge, only thirteen cases have been reported in the literature [2,4-12]. Data of previously described observations are summarized in table 1. Cranial nerve schwannomas

Table 1: Clinical findings in the patients with oculomotor nerve schwannoma mimicking recurrent painful ophthalmoplegic neuropathy (RPON) [2,4-12]

Case	Authors	Age (years)	Gender	Location of schwannoma	Headache	Number of episodes	Persistent enhancement in MRI
1	Kawasaki et al (1999) [7]	23	F	Cisternal	Yes	6 (7-23 years)	NM
2	Murakami et al (2005) [8]	11	F	Cisternal	Yes	2 (4-6 years)	60 months
3	Bisdorff et al (2006) [9]	14	F	Cisternal	Yes	>2 (4-14 years)	2 months
4	Riahi A et al 2014 [10]	12	F	Cisternal	Yes	3	NA
5	Kim R et al 2015 [6]	52	M	Cisternal	Yes	NM*	12 months
6	Kim R et al 2015 [6]	31	F	Cisternal	Yes	10 (26-31 years)	8 months
7	Shin RK et al. (2015) [4]	41	F	Cisternal	Yes	3 (41-52 years)	13 months
8	Shin RK et al. (2015) [4]	23	F	Cisternal	Yes	7 (7-30 years)	6 months
10	Shin RK et al. (2015) [4]	43	M	Inferior division of III	No	2	10 months
9	Jibia A et al. (2015) [5]	13	F	Cisternal	Yes	NM* (7-13 years)	6 years
11	Abo-Shasha R et al 2018 [2]	46	M	level of the anterior clinoid process	Yes	NM * (26-46 years)	13 years
12	Lee D et al 2018 [11]	10	F	Cisternal	Yes	2 (2-10 years)	12 months
13	Petruzzelli MG et al 2019 [12]	16	M	Fork of the right basilar artery near to perimesencephalic and interpeduncular cisterns	Yes	5 (6 -16)	7 years
Our case	**Mrabet et al**	18	F	Cisternal	Yes	4 (8-17years)	12 months

*NM= Not Mentioned. NA= Not Available

would appear iso-intense to brainstem on both T1- and T2-weighted images on magnetic resonance imaging (MRI). They are characterized by nodular cranial nerve enhancement after Gadolinium injection [3,4]. In our patient, the location of the lesion in the cisternal segment of the third nerve and persistent nodular enhancement, even during asymptomatic period are highly suggestive of schwannoma. Differential diagnosis is mainly discussed with RPON which is a rare syndrome with episodic headaches and ipsilateral ophthalmoplegia. RPON's onset is typically before age 10 years. The headache resolves quickly, but the oculomotor palsy recovers gradually over days to weeks [1]. In RPON, the oculomotor nerve is more commonly involved than the trochlear or abducens nerve [7]. Incomplete recovery of cranial nerve palsy is possible during migrainous attacks. However, persistent ophthalmoplegia rather suggests other organic lesions involving the ocular motor cranial nerve. Gadolinium enhancement and nerve thickening can be shown on MRI in RPON in the acute phase but disappeared among the

12 first weeks after recovery from the symptoms [7]. In our patient, migrainous headache preceded ophthalmoplegia up to 15 days and resolved when nerve palsy appeared in the four described episodes. In table 2, we summarized different aspects of differential diagnosis and consecutive pitfalls.

Cranial nerve schwannomas may be asymptomatic or present clinically with dysfunction of the nerve from which they arise or may cause dysfunction of neighbouring structures. Recurrent painful ophthalmoplegia may also occur with other tumors such as meningiomas, chordomas, and optic nerve gliomas [8]. The mechanism of intermittent unilateral headaches with ipsilateral ophthalmoplegia remains hypothetical. Intermittent release of a chemical substance from schwannoma and resultant stimulation of the trigeminal nerve receptors could explain migrainous headache [9].

Treatment for cranial nerves schwannomas aims to relieve patients from neuro-ophtalmological symptoms caused by the tumour. However, surgical excision which represents the classical approach may cause permanent oculomotor nerve deficit. Stereotactic radiotherapy constitutes a safer alternative with promising results [2].

For small asymptomatic tumors with minor or recurrent symptoms, as in our patient, a conservative strategy called “wait and see” is recommended. A clinical follow up with brain MRI monitoring is thereby necessary [9,10].

CONCLUSION

Oculomotor nerve schwannoma with relapsing clinical course is exceptional. It may manifest as RPON. A clinical and MRI follow up is needed to identify the tumor and to avoid inappropriate diagnosis of primary headaches. Treatment must be individualized for each case.

Conflicts of interest. None.

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Table 2. Recurrent painful ophthalmoplegic neuropathy (RPON) and schwannoma of oculomotor nerve: differences and similarities

	RPON	Oculomotor nerve schwannoma
Age at onset	First decade	Variable
Clinical manifestations	headache Ophthalmoplegia (mainly oculomotor nerve)	Headache Ophtalmoplegia
Outcome	• Recurrent episodes • **Complete regression**	• Progressive course: typical • Recurrent episodes: **rare** • **Incomplete regression**
Brain MRI	-**Diffuse** enhancement of cranial nerve **only** during acute exacerbations -Regression of this enhancement after **3 months** from symptoms onset	- Nodular enhancement in cranial nerve - Location: **cisternal** segment of the nerve - **Persistent** enhancement
Treatment	• Steroids	• Surgery • Stereotactic radiotherapy • « Wait and See »

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