Tortuous optic nerve

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Aims and objectives

Tortuous optic nerves (TON) are abnormally curved optic nerves. These are easily detectable changes even on brain MRI scans. Based on the literature optic nerve (ON) becomes tortuous in case of intracranial hypertension [1]. TON is an often reported MRI finding in benign intracranial hypertension (BIH) [1-4], but only a few studies mentioned TON associated with sinus thrombosis or Chiari malformation [5,6]. However, tortuous optic nerve can be associated with diseases without clinical signs of IC hypertension, such as neurofibromatosis type 1 (NF1) [7-10].

Optic nerve has intracranial, intracanalicular, intraorbital and intraocular segments. Intraocular ON fibers passes through the lamina cribrosa and forms the intraorbital part of the nerve, which part has free space for eyemovements. The nerve's intracanalicular part is tightly fixed in the optic canal. The development of optic nerve tortuosity associated with elevated intracranial pressure is attributed to the fixation of proximal and distal part of the nerve [2]. The elevation of ICP can be the result of increased intracranial CSF, blood or brain volume based on Monroe-Kellie principle [11,12]. Mechanism of raised ICP is not clear in a group of BIH patients (idiopathic BIH). In other group of BIH patients with transverse sinus stenosis and/or sinus thrombosis ICP elevation can be a consequence of reduced CSF outflow [5],[13].

Optic nerve sheath dural ectasia is a saccular dilatation of the meninges surrounding the optic nerve in the orbita [14]. This disease is most common associated with NF1 patients [14].

In this study our goal was to examine which groups of pathologies are associated with TON. We speculated on the possible explanation of the mechanism of TON in the different pathological groups as well.
Methods and materials

50 consecutive TON patients’ MR studies were examined retrospectively. There were 23 female and 27 male, ranging in age from 1 to 83 years (mean: 33 years).

We established the existence and degree of optic nerve tortuosity on 3-plain MR images based on Armstrong’s recommendations (Fig. 1 on page 4) [15]. MR images were acquired with a 1.5 T GE Signa Excite HDxt MR Scanner (GE Healthcare, Chalfont St. Giles, UK). In axial plane we observed T2 FSE sequences (TR:<2500 msec, TE: 85 msec, ST: 5 mm). In sagittal and coronal plane acquired T1W images were reviewed (TR:<600 msec, TE: 5-15 msec, ST: 5 mm). Based on pathological findings in some cases axial and coronal STIR, coronal IR sequences and sagittal T2WI were made.

In axial fast spin-echo T2-weighted images we rated if there is an interruption of the optic nerve out of the axial plane without (factor 1) or with (factor 2) return into the axial plane or there is a deviation of the ON within the axial plane (factor 3). Sagittal T1-weighted images showed the vertical tortuosity of optic nerves (factor 4). On coronal images we searched for lack of congruity of the optic nerves (factor 5) and dilation of the anterior portion of the ON sheath (factor 6) (Fig. 1 on page 4). [15]

The existence of all the 6 factors were checked in each optic nerves (100 cases) by two observers independently. Kruskal-Wallis one-way analysis of variance on ranks test was performed to compare the results of all of the groups with each other, with p<0.05 indicating significance.

Patients were divided into five groups based on the probable mechanism of optic nerve tortuosity. In the first three groups there were signs of intracranial hypertension. In the first group, cases of intracranial CSF volume growth (CSF), in the second, increased intracranial blood volume (BV), in the third, brain space occupying lesions (SOL) were presented. Patients with type 1 neurofibromatosis (NF1) belonged to the fourth group. Patients with other (O) presumed mechanism of TON composed the last group.
Fig. 1: a, b, c: axial fast spin-echo T2-weighted images of our patients. Interruption of the ON out of the axial plane without return (a) and with return (b) into the axial plane. Deviation of the optic nerve within the axial plane (c). d: sagittal T1-weighted image of vertical tortuosity of the optic nerve. e: on coronal STIR image the lack of congruity of the optic nerves and dilation of the anterior portion of the ON sheath can be detected. [15]

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Results

24% (n=12) of TON patients belonged to the CSF group (Fig. 2 on page 7). In the patients of this group, hydrocephalus developed due to different origins, eg. meningitis or subarachnoid haemorrhage. In the BV group, 16% (n=8) of the TON patients were diagnosed with sinus thrombosis or BIH with sinus stenosis. Space occupying lesion (SOL) was detected in 36% (n=18) of the cases. 12% (n=6) of TON patients was diagnosed with type 1 neurofibromatosis (NF1) (Fig. 3 on page 7). Other developmental mechanism was suspected in staphyloma, orbital trauma, Chiari malformation (Fig. 4 on page 8) and pansinusitis, this group was 12% (n=6) of the cases.

The existence of the 6 tortuosity factors were examined in every patients. In addition, the degree of optic nerve tortuosity was estimated in all of the 100 optic nerves (presence of six tortuosity factors). There was no significant difference among the groups (Kruskal-Wallis rank sum test, p-value = 0.1325) (Fig. 5 on page 9).

In the first three groups ICP elevation from different origins explains TON (Fig. 6 on page 9). The globes are tethered in the orbit by the rectus muscles and their ligaments. In case of increased ICP followed by perioptic nerve sheath distension this tethering provides resistance to the distal portion of the ONS, which results in kinking of the inflated optic nerve [1,4].

In the fourth, NF1 group dural ectasia can be a possible mechanism of TON formation.

In group 5 the reason of TON were local intraorbital lesions in two cases (staphyloma, orbital trauma). Posterior staphyloma is an abnormal protrusion of the uveal tissue through an acquired defect of the posterior scleral layer. It leads to elongation of the globe in the anteroposterior axis, affecting the posterior pole [16]. Increased size and deformity of the eyeball cause that the two points fixed optic nerve become tortuous in cases of posterior staphyloma (Fig. 7 on page 10).

Trauma followed by edema can develop TON due to its space occupying nature (Fig. 8 on page 11).

In cases of Chiari I malformation (n=2) the displacement of cerebellar tonsils below the level of the foramen magnum were observed. The ectopic tonsils can lead to the interruption of CSF flow and this may result increased ICP. However, a recent study reported significant tonsillar ectopia in BIH patients [17]. From this aspect cerebellar tonsillar ectopia can also be a consequence of increased ICP.

In cases of sinusitis (n=2) the mucosal swelling can result lymphatic disturbance, which might cause IC hypertension by blocking CSF absorption (Fig. 9 on page 11). Recent
studies reported that the lymphatic route of cerebrospinal fluid absorption mainly drains to the nasal mucosa through the cribriform area [18-20]. In animal studies, biological stain injected into the subarachnoideal space appeared in nasal submucosa [18]. Blocking this route of CSF absorption also caused intracranial pressure elevation [19]. Pansinusitis associated cases raise the possibility that lymphatic route dysfunction of CSF absorption can play an important role in the development of IC hypertension.
Fig. 2: Proportion of TON patients among the five groups.

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Fig. 3: 9-year-old boy with multiple cafe au lait spots and neurofibromas on his body. a: on coronal STIR sequence multiple hyperintense lesions in the basal ganglia. b: vertical tortuosity of ONs on T1WI. Axial T2WI shows hyperintense lesions in the cerebellum (c), and deviating ONs from the axial plane (d)

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**Fig. 4:** 5-year-old boy with 2-3 times epileptic episodes. a: sagittal T2 FSE shows the cerebellar tonsils below the plane of foramen magnum. No significant hydrocephalus or other intracerebral lesion. b: on axial T2 FSE image optic nerves deviation from the axial plane without return. c: on coronal IR image incongruity and deviation of the ONs and dilation of the anterior portion of the ON sheath presented.

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**Fig. 5:** Presence of the six tortuosity factors in different groups. There was no statistical difference between the groups (Kruskal-Wallis rank sum test, p-value = 0.1325). CSF (n=12), SOL (n=18), BV (n=8), NF1 (n=6) and other (O) (n=6) groups were created based on the suspected developmental mechanism of TON.

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Fig. 6: 3-year-old girl with strabismus, ptosis and ataxia. a: on axial T2 FSE images marked internal hydrocephalus was detected. Cause of obstruction was a non-enhancing tumour in the dorsal part of the mesencephalon. b: vertical tortuosity of the right ON on T1 weighted image.

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Fig. 7: Posterior staphyloma of both eyes in a 67-year-old male patient. On axial T2WI (a) both optic nerves are kinking in the axial plane. On sagittal T1WI (b) vertical tortuosity of the right ON can be seen.

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Fig. 8: A wooden foreign body reached the 30-year-old male patient’s left orbit while sawing. After surgical remove of the foreign body the deviation of the left optic nerve is still visible on axial (a) and coronal (b) STIR image. In the sagittal plane (c) mild tortuosity can be detected.

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**Fig. 9:** On axial T2WI image (a) marked mucosal thickening can be detected in all of the paranasal sinuses in a 73-year-old patient. On sagittal image the tortuous right ON and right maxillary sinusitis can be seen (b). In another case on the coronal T2WI (c) the incongruity of the optic nerves and pansinusitis are visible.

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Conclusion

Optic nerve tortuosity develops not only in intracranial hypertension of different causes and in NF-1 patients, but also in cases without obvious intracranial pressure elevation. It can not be considered as a specific sign, but may draw attention - if the well-known associations can be excluded - to some rarer pathologies such as local orbital lesions. It might also raise the possibility of the causative relationship between the Chiari malformation and increased ICP. It might highlight the significance of lymphatic drainage in the CSF absorption.
Personal information

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