Case report

Clin Ter 2023; 174 (3):??-?? doi: 10.7417/CT.2023.????

Papilledema in childhood for diagnosis of pseudotumor cerebri or late hydrocephalus: a case report

E.M. Vingolo[1,2], M. Malvasi[2*], S. Ciarlo[3], A. Pompucci[3], F. Pacella[2], E. Pacella[2]

[1]Fiorini Hospital Terracina AUSL, 04019 Terracina, Latina, Italy; [2]Department of Sense Organs, Faculty of Medicine and Dentistry, Sapienza University of Rome, Rome, Italy; [3]UOC of Neurosurgery, Santa Maria Goretti Hospital, Latina, Italy

Abstract

A case of hydrocephalus is presented in a 13-year-old female with transient loss of vision from 1 week and papilledema, previous ophtalmological history was negative. Visual field was performed, and neurogical examination proved to be hydrocephalus. In literature few cases of Papilledema in adolescent children with hydrocephalus has been reported. The aim of this case report is to decode the signs, symptoms and factors associated with papilledema in children with hydrocephalus at an early stage in order to prevent a poor visual-functional residual (permanent low vision). ***Clin Ter 2023; 174 (3):??-?? doi: 10.7417/CT.2023.????***

Key words: *Papilledema, hydrocephalus, pseudotumor cerebri, childhood, loss vision*

Introduction

Hydrocephalus most frequently results from excessive cerebrospinal fluid (CSF) volume, with increased size of the cerebral ventricles and endocranial pressure, or from reduced brain mass due to degenerative diseases or congenital abnormalities. The etiopathogenesis is not yet fully known, probably resulting from an imbalance between production and absorption, most frequently with intraventricular accumulation of CSF.

The case we describe is a form of tetra-ventricular late hydrocephalus, which, as reported in the literature, is rare and causes severe morbidity and death in children, which necessitates an early diagnosis, since although it presents a slow progression the prognosis is generally inauspicious.

It came to our attention in a 13-year-old girl, with general subjective symptoms: headache, stupor, disorientation, diplopia, and altered state of consciousness that had been present for a week, and ophthalmological disorders transient loss of visual acuity (AV) lasting a few seconds of recent onset to be attributed to the progressive papilledema. The diagnosis was made after performing all relevant clinical examinations, including comprehensive ophthalmic examination, neurological and neurosurgical evaluation and instrumental examinations such as visual field (C.V.), bulbar ultrasound, optical coherence tomography (OCT), brain RM.

The purpose of this rare case report is to emphasise how early diagnosis and treatment are indispensable to avert the complications associated with the onset of papilledema with consequent irreversible loss of visus, as well as to describe the clinical presentation, which is of considerable neuro-ophthalmological and neurosurgical difficulty, and which represents a diagnostic challenge that requires teamwork to enable diagnosis to be made as soon as possible.

Case report

A 13-year-old white woman, overweight (BMI=25), came to our observation, at the Fiorini Hospital in Terracina, Latina, Italy, Department of Ophthalmological Sciences, "La Sapienza" University of Rome, for ocular pain, photophobia, transient loss of AV in both eyes since 1 week, asthenia, drowsiness and headache.

At the general anamnesis the patient reported, in particular, an increase in body weight and dysmenorrhoea, the ophthalmological anamnesis no refractive defects was reported, nor previous ocular pathologies worthy of note, while for about 6 months divergent strabismus corrected with the use of prismatic lenses (prism 5 with internal base) and a VA of 10/10.

The complete ophthalmological examination of our adolescent with papilledema included the valuation of best correct visual acuty (BCVA) which was 3/10 in right eye, and 4/10 in left eye, and which could not be improved with lenses. Colour vision was generally compromised with the progression of the papilledema. The adnexa were intact, the anterior segment normal. Intraocular pressure (IOP), measured with a Goldmann applanation tonometer was 12 mmHg in both eyes. Fundus examination revealed the presence of marked papilledema with significant changes in its morphology. The oedema partially involved the posterior

Correspondence: Mariaelena Malvasi, email: mariaelena.malvasi@uniroma1.it

ISSN 1972-6007

pole took on the typical star-shaped appearance with sparing of the macular region, the retina in the periphery was uninjured and on the plane.

The C.V. performed with Octopus 900 kinetic vectors perimeter, to assess and monitor any deficits and identify any imminent risk of visual function, showed concentric constriction of the isopters in both eyes, even if the execution of the examination is partially complete due to the symptoms in progress that did not allow a good collaboration of the patient. It can be observed in the right eye, with the aim 1-4 exclusion of the blind spot, and in the left eye with the aim 1-3 an enlargement of the blind spot. At the OCT examination, performed with Heidelberg Spectralis sistem, there was no evidence of oedema in the macular area, while there was an increased thickness of the retinal nerve fibre layer (RNFL) with a value of 633 μm in righe eye and 732 μm in left eye. The clinical picture was compatible with pseudotumor cerebri (PTC).

The bulb ultrasound in B-scan, performed with a 50 MHz probe showed vitreal inhomogeneity. The increased value of the transverse diameter of the bilateral optic nerve head: right eye = 5.97 mm and left eye = 5.97 mm respectively from the retinal plane and marked accumulation of CSF in the subarachnoid periosteal spaces.

In order to reduce the oedema of the posterior pole, the first treatment approach was conservative, with methylprednisolone sodium succinate 1g ev 1ml die for 3 days and Esomeprazole 20 mg 1 ml die. A new ophthalmological assessment was carried out on the second day of therapy, as the patient reported a worsening headache, accompanied by asthenia and chest pain, increased photophobia, persistent pain in both eyes, which confirmed the worsening of AV of 1/10 in the right eye and 2/10 in the left eye. The anterior segment was in the normal range and IOP of 12 mmHg in both eyes. The objectivity of the fundus examination revealed the persistence of oedema of the papilla with slight reduction (margins less blurred) the oedema involved the posterior pole and licked the macular region, while the retina in the periphery was unharmed. Performing a new C.V. showed increased constriction of the isopters in both eyes: the right eye presented a reduced visual field at 5° central with perimetric residual at 5° central, annular scotoma and the left eye presented a perimetric residual at 10° central, annular scotoma. On OCT the RNFL thickness was still above normal, but decreased from the previous finding with a value (583 μm in righ eye, and 680 μm in left eye).

A neurosurgical consultation and a brain RM with gadolinium was therefore immediately requested, which excluded the presence of cerebral neoplasms, stenosis and venous sinus thrombosis; showing instead increased ventricular spaces with accumulation of CSF in the optic sheaths to be referred to endocranial hypertension, empty sella due to compression of the pituitary gland and herniation of the CSF-filled arachnoid diverticulum in the sella turcica, the latter clinical sign was compatible with a pseudotumor cerebri form. Consequently, medical therapy with acetazolamide (Diamox) and Furosemide was carried out.

In view of the worsening visual acuity and clinical symptoms, and the lack of efficacy of medical therapy, after consultation and then the neurosurgical and ophthalmological diagnosis of increased intracranial pressure (IPC), resulting from tetra-ventricular hydrocephalus; patients was candidate to surgery. In order to avert loss of vision, after informed consent of the parents, Helzinchi's statement, the patient underwent ventricle-peritoneal shunt implantation surgery, through a right abdominal incision and tunnelling of the catheter at the right retroauricular level, a Progav 2 valve was connected. 0 (160 mmHg Shuntassisant 25 cmH20), then a right frontal pre-coronary flap was created. then the catheter was introduced through a hole and the CSF leak was assessed, then the valve was connected and function was checked at the abdominal level. Finally, the catheter was introduced into the peritoneum and attached to the superficial abdominal fascia.

One week after surgery AV showed progressive improvement progressing by 2 Snellen lines in both eyes, with progressive improvement in the following months: after 1 month in both eyes BCVA 3/10, with suboptimal vision quality; IOP was 16 mmHG. The C.V afther 1 month post surgery showed improvement of the previously observed scotoma. Ophthalmoscopically the papilla showed gradual resolution of the oedema documented by OCT scans showing reduction in the thickness of the RNFL 476 μm in right eye and 500 μm in left eye. Fig. 1 shows the grey map of the visual field performed before and afther surgery.

Thickness trend of RNFL before and afther surgery acquired by OCT execution is showed in Fig. 3 and the rispective trend of optic disc infrared (IR) images, before and afther surgery, rispective acquired during the OCT execution, wich is rappresentative of papilledema severity.

Discussion

Hydrocephalus is a condition caused by an imbalance between cerebrospinal fluid production and absorption; it can cause severe morbidity and death in children. Subjective symptoms in children in order of frequency are: headache (57-87%), amaurosis fugax, blurred vision, pulsatile tinnitus, diplopia, transient visual loss of < 30 sec (16 - 42.3%). Less frequently occurring are paresthesias, abducens paralysis (CN VI) 10-17%, back and lower limb pain 4-8%, nausea and vomiting 12.7-52%, arthralgias and unsteady gait, irritability, fatigue and increased head circumference volume. (1)

Sometimes these symptoms may be absent, particularly if ICP increases slowly, resulting in unspecificity (2). Papilledema is considered one of the most important signs of hydrocephalus and a determining factor in management strategy (2). The absence of papilledema in younger infants with hydrocephalus can be attributed to the presence of open fontanelles, which in the newborn allow for the expansion of the skull and are protective against an increase in ICP, so that its increase is not sufficient to cause papilledema (3).

In the literature, the incidence of papilledema has been estimated to be 14-50% in children with hydrocephalus caused by ventriculomegaly, and approximately 65% in children with hydrocephalus secondary to primary brain tumours. (2). Papilledema is one of the most important signs of hydrocephalus and if undetected, can lead to irreversible damage up to blindness due to optic atrophy. The absence of papilledema may be a false-negative sign (3). Lee HJ et al. (4) reported absence of papilledema in 41% of children with

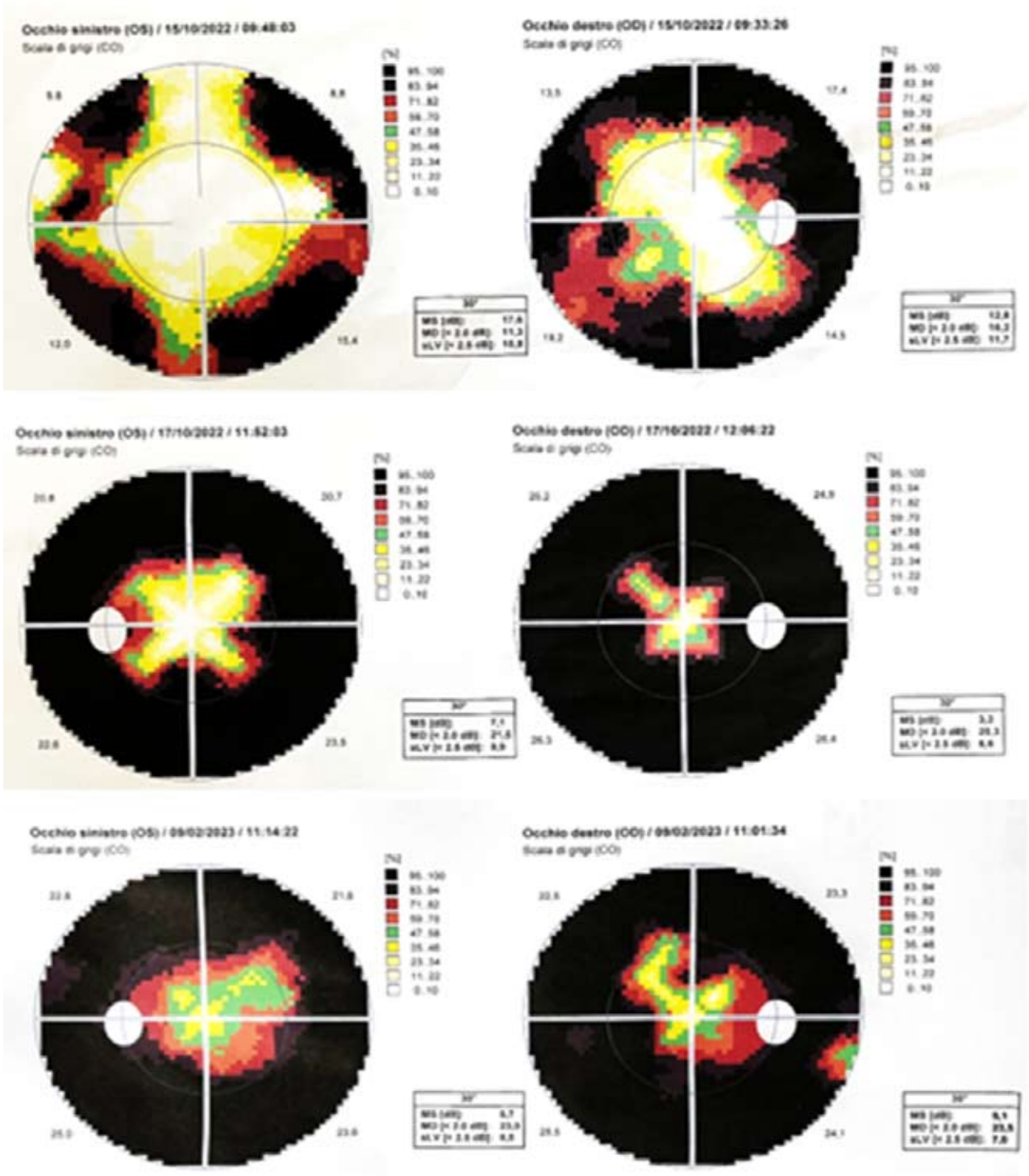

Fig. 1 shows the grey map of the visual field performed before and afther surgery.

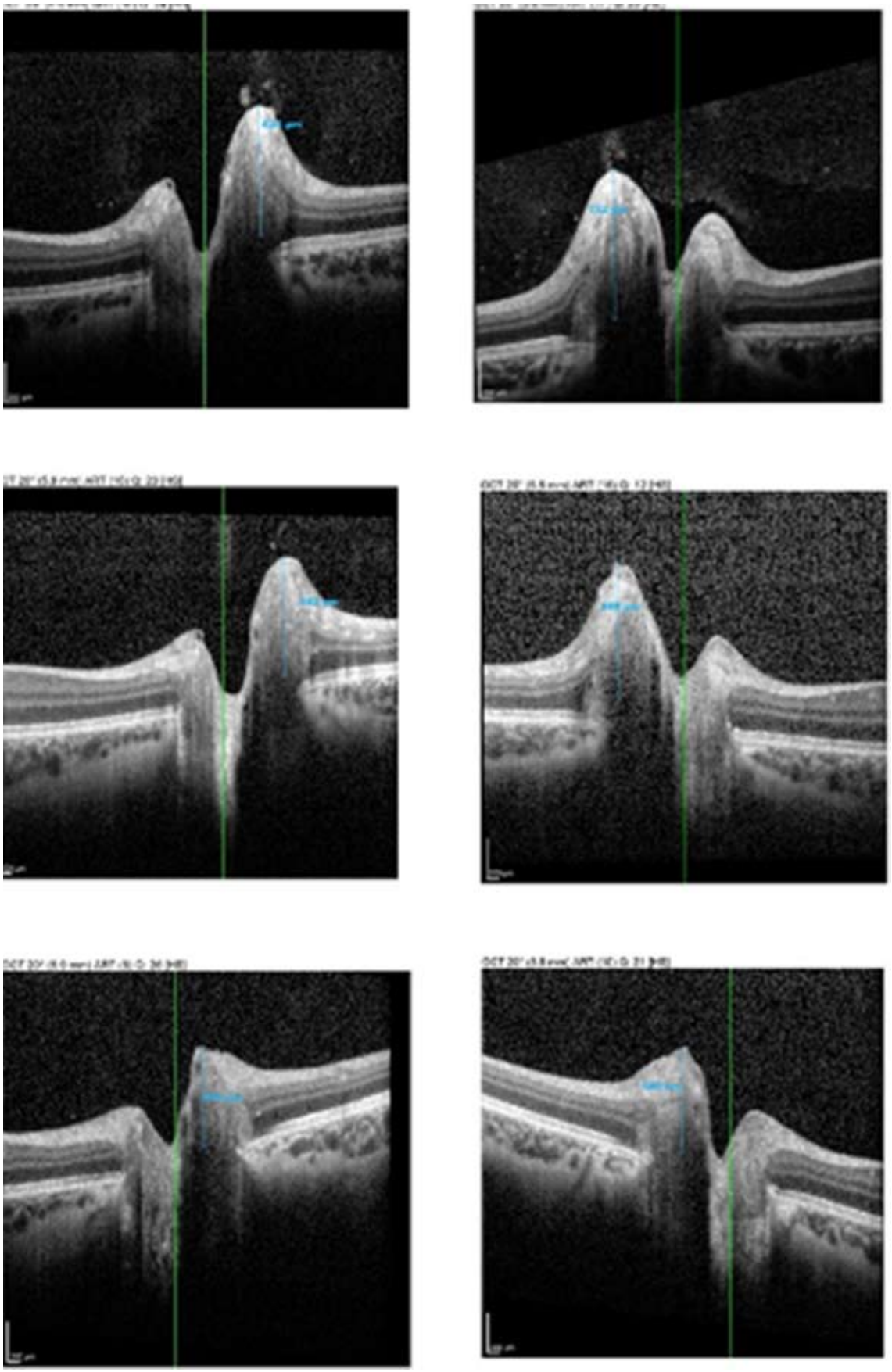

Fig. 2 Thickness trend of RNFL measured before and afther surgery acquired by OCT execution

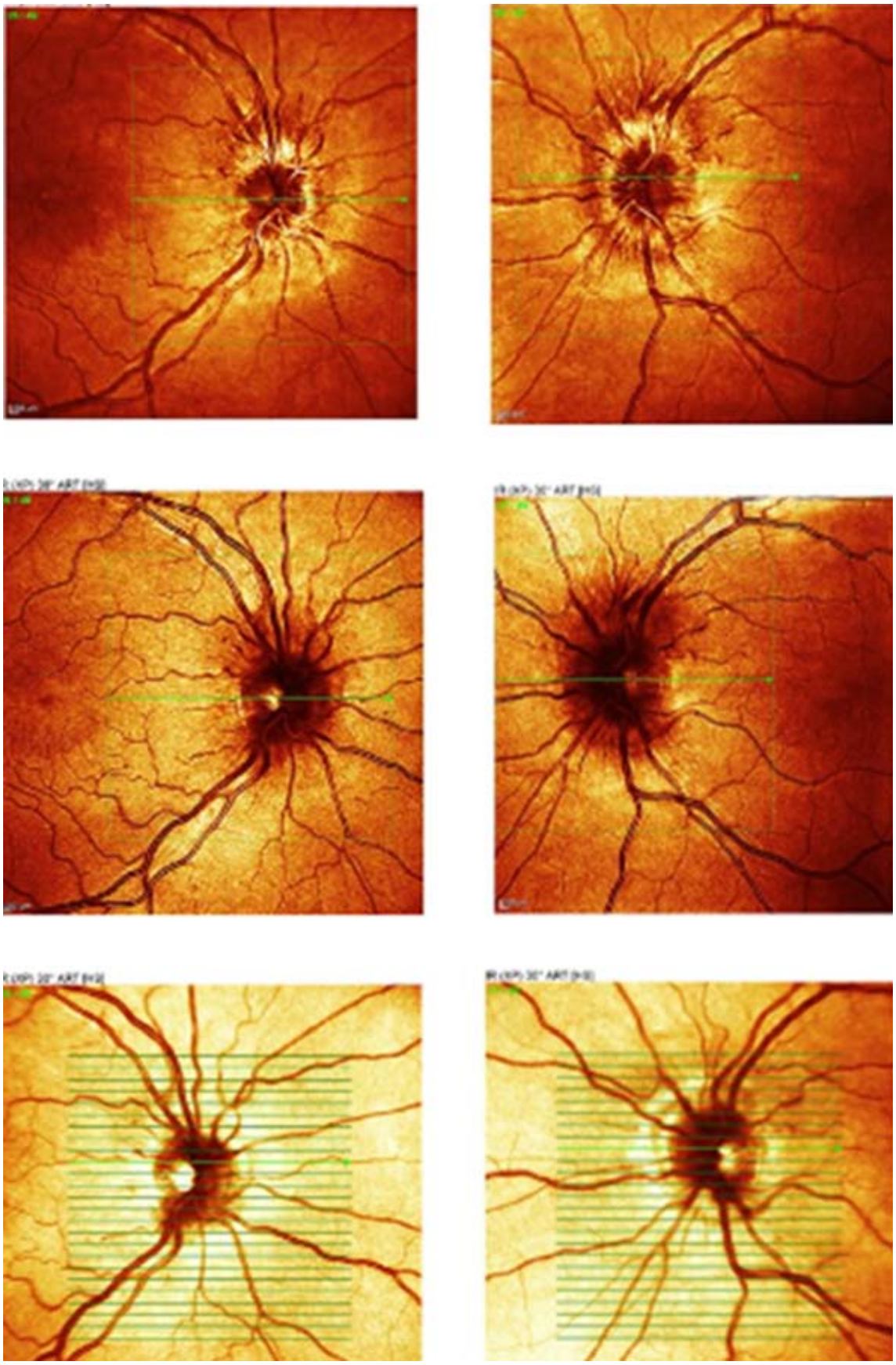

Fig. 3 Trend of optic disc infrared (IR) images before and afther surgery rispective acquired during the OCT execution

hydrocephalus who were, however, younger than those with papilledema and it was also more common in older patients (mean age of 8.8 ± 4.2 years) with higher ICP (of 33.3 ± 9.1 cm H2O) with a mean symptom or sign of 3.4 ± 3.9 months, the causes of which were recognised in order tumour (59%), congenital anomaly (19%), (19%), haemorrhage (13%) and infection (9%).

The duration of ICP elevation is an important factor in the development of papilledema (3). In fact, pathophysiologically, the alteration that occurs in the optic nerve fibres during papilledema is the axonal swelling caused by the elevation of the ICP, with consequent impairment of axoplasmic flow. On the contrary in the case of acute elevation of ICP there is a blockage of both anterograde and retrograde axoplasmic flow that is not associated with swelling of the optic disc, for this reason papilledema is not considered a sensitive sign in the case of acute elevation of ICP. (3,5).

ICP associated with papilledema found in an overweight female adolescent, presented to our observation in the first instance with signs and symptoms typical of increased intracranial pressure, led us to hypothesise a case of pseudotumor cerebri (PTC) a condition in which the brain parenchyma is normal.

In paediatric studies, the annual incidence of PTC is estimated at 0.6 - 0.71/100,000 (6). Obesity is a risk factor that impacts beyond the age of 12 years, as it is closely related to pubertal status in the pathophysiology of primary PTC. In contrast, in children under 12 years of age, weight does not seem to influence the development of PTC (8). The diagnostic criteria for PTCS were revisited by Friedman et al. in 2013, our case did not present any features of a PTC (1) although it is reported in the literature that up to 53-78% of cases in children included obesity (7,8).

For the assessment and correlation of changes in optic nerve head volume and total retinal thickness with the degree of papilledema, instrumental examinations such as orbital ultrasonography and OCT (9) were performed in order to identify signs of optic neuropathy from ICP, and thus identify patients requiring timely treatment in order to preserve vision (9, 10).

In fact, the following report describes the well-documented case of papilledema in an adolescent female patient with hydrocephalus and optic neuropathy from IPC, diagnosed through the appropriate instrumental support that showed dilatation of the ventricular spaces quickly, and excluded through a careful differential diagnosis with a PTC, whose clinical presentation in children varies with age and often takes on sometimes blurred pictures, which make diagnosis difficult.

The presence of papilledema in hydrocephalus is one of the most useful indicators for the preoperative diagnosis of increased intracranial pressure (7).

In fact, the extent of papilledema represented in our study an important criterion for the morpho-functional assessment of the therapeutic efficacy of medical therapy for ventriculoperitoneal shunt surgery, as well as being considered as a criterion for post-operative follow-up.

In accordance with the most recent literature, surgical treatment is currently the optimal therapeutic aid in hydrocephalus. In fact, the patient presented a marked improvement in visual acuity and objective picture following the ventriculoperitoneal shunt surgery.

Indeed, it is essential to emphasise the importance of early diagnosis of hydrocephalus, even when it presents in these atypical forms, which may be suggested by the presence of papilledema. It is advisable in these patients to pay attention to visual symptoms and therefore to seek timely ophthalmic consultation, in order to preserve visual function, as chronic papilledema may lead to irreversible optic atrophy.

References

1. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. The most reently revised diagnostic criteria for PTCS, including criteria for assessing children both with and without the presence of papilledema. Neurology. 2013 September 24; 81(13):1159–65 doi: 10.1212/WNL.0b013e3182a55f17
2. Corbett JJ. Neuro-ophthalmologic complications of hydrocephalus and shunting procedures. Semin Neurol 1986; 6:111–123
3. Nazir S, O'Brien M, Qureshi NH, et al. Sensitivity of papilledema as a sign of shunt failure in children. J AAPOS 2009; 13:63–66
4. Lee HJ, Phi JH, Kim SK, et al. Papilledema in children with hydrocephalus: incidence and associated factors. J Neurosurg Pediatr. 2017 Jun; 19(6):627-631. doi: 10.3171/2017.2.PEDS16561. Epub 2017 Apr 7. PMID: 28387641
5. Steffen H, Eifert B, Aschoff A, et al. The diagnostic value of optic disc evaluation in acute elevated intracranial pressure. Ophthalmology 1996; 103:1229–1232
6. Matthews YY, Dean F, Lim MJ, et al. Pseudotumor cerebri syndrome in childhood: incidence, clinical profile and risk factors in a national prospective population-based cohort study. Arch Dis Child. 2017 August; 102(8):715–721. doi: 10.1136/archdischild-2016-312238
7. Bursztyn LL, Sharan S, Walsh L, et al. Has rising pediatric obesity increased the incidence of idiopathic intracranial hypertension?. Can J Opthalmol. 2014 February; 49(1):87–91. doi: 10.1016/j.jcjo.2013.09.015
8. Paley GL, Sheldon CA, Burrows EK, et al. Overweight and obesity in pediatric secondary pseudotumor cerebri syndrome. Am J Ophthalmol. 2015 February; 159(2):344–52.e1. doi: 10.1016/j.ajo.2014.11.003
9. Chen JJ, Thurntell MJ, Longmuir RA, et al. Causes and prognosis of visual acuity loss at the time of initial presentation in idiopathic intracranial hypertension. Invest Opthalmol Vis Sci. 2015 June; 56(6):3850–9. doi: 10.1167/iovs.15-16450
10. Turchetti P, Pacella F, Pacella E, et al. An immunocompetent migrant presenting with neurosyphilis with an unusual unilateral papillitis: a case report. Eur J Med Res. 2012 Feb 14; 17(1):3. doi: 10.1186/2047-783X-17-3. PMID: 22472320; PMCID: PMC3337542