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RESEARCH ARTICLE

PNEUMATOCYST COMPLICATING A GIANT NECROTIC MACROPROLACTINOMA: ABOUT A VERY RARE CASE.

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Abstract

Treatment of patients with giant and invasive macroprolactinomas consists of dopamine agonists (cabergoline or bromocriptine). When they reduce the size of invasive prolactinomas, the intra and extra cranial spaces may communicate causing cerebrospinal fluid (CSF) leakage and / or pneumocephalus which are very scarce complications after medical treatment. We report the case of an 18-year-old girl who was admitted in our department for hormonal evaluation of a pituitary tumor, she had been treated with cabergoline after a giant invasive prolactinoma was diagnosed. After 4 weeks, she presented with severe headaches and liquid nasal discharge, her brain CT showed a skull base lysis with erosion of the sellar floor creating a communication with the sphenoidal sinus resulting in a pneumatocyst. A depletive lumbar puncture was performed, with diamox and prophylactic antibiotics with a good evolution marked by the drying up of the CSF rhinorrhea. Clinicians should be aware of these complications symptoms to ensure proper diagnosis and treatment.

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Introduction:-

Prolactinomas are the most frequent pituitary tumors, they occur more commonly in females in young population [1]. Most of the prolactinomas can be medically treated by bromocriptine or cabergoline, which are largely used as the treatment of first choice for prolactinomas, including giant, large and invasive ones.

Cabergoline is generally rapidly efficient in normalizing the prolactin level, while the tumor reduction can be modest or delayed. Intratumoral hemorrhagic necrosis may possibly occur in giant prolactinomas under cabergoline with cerebrospinal fluid fistula in high dosage of cabergoline. Exceptionally, air from the sphenoid sinus can accumulate within the capsule of the necrosed macro adenoma through a one-way valve mechanism. If under pressure, this trapped air is called a “pneumatocyst” or “pneumocephalus”, and exerts a significant mass effect on the adjacent cerebral structures [2].

Case report:-

An 18-year-old female patient with a history of primary amenorrhea, admitted in the endocrinology department for hormonal evaluation of a pituitary tumor, whose onset dates back to one year by the installation of intermittent right hemi temporal headaches with ipsilateral retro-eye pain and double vision during up and down eye movements, the

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patient also reported asthenia which is increasing during the day, a recent undocumented weight gain estimated moderate, and a hair loss. On examination she had an asymmetrical look with exophthalmia of the right eye, her pulse rate was at 76/min, her blood pressure was at 120/90, she has a statural delay at -2 DS, and her BMI was at 24.5 kg / m², otherwise she had no galactorrhea or other associated signs.

On blood examination, she had an hyperprolactinemia at 10,000 mIU / l, TSH at 1.78 mIU / l, T4 at 9.84 pmol / l, cortisol at 44 ug / dl, FSH at 4.4 IU / l and LH at 0.9 IU / l, her pituitary MRI showed a large intra and supra-sellar lesion of 7 * 4.2 * 3.8 cm infiltrating the cavernous sinuses, carotid arteries and the right endo-orbital cavity with exophthalmia grade II (Fig 1 and 2).

The diagnosis of macro prolactinoma was retained and the patient was put on cabergoline 2 mg per week and levothyroxine 50 ug per day.

Evolution was marked by installation of severe headaches, decreased visual acuity of the right eye and a liquid nasal discharge 1 month after starting the treatment, her brain CT showed a lysis of the skull base with erosion of the sellar floor creating a communication with the sphenoidal sinus resulting in a pneumocephalus (Fig 3). The management consisted on a depletive lumbar puncture, with rehydration, and the patient was put under hydrocortisone, diamox and prophylactic antibiotics, 3 weeks later, she was discharged home, currently, she presents a regression of headaches and visual disturbances with absence of rhinorrhea, the control MRI showed a sellar and supra sellar hydroaeric cavity which measures 50 * 48 * 40mm (Fig4).

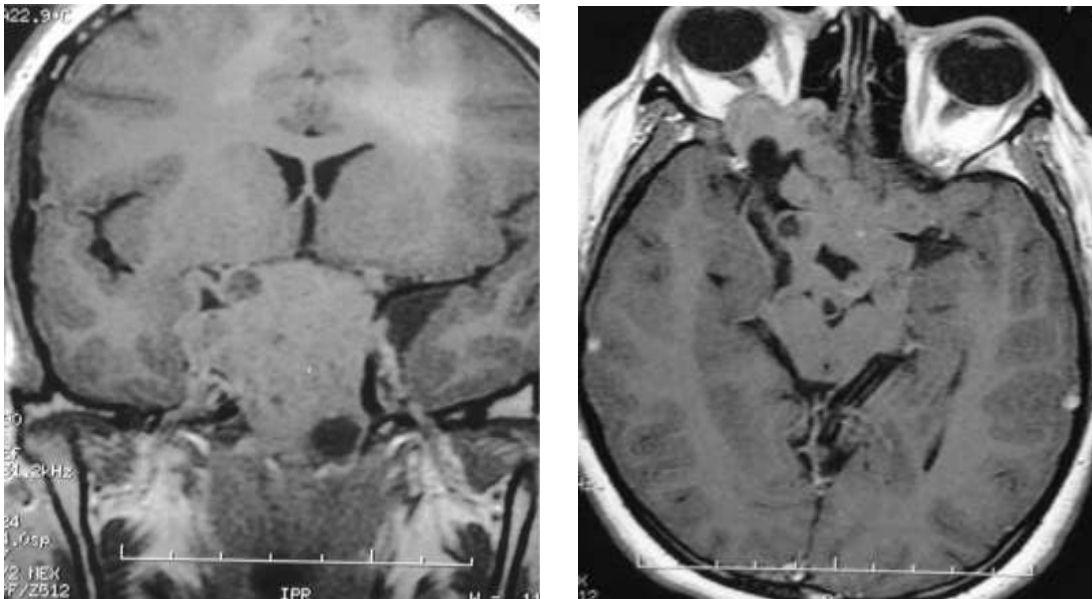


Fig 1 and 2: pituitary MRI at initial evaluation before the introduction of oral therapy. Giant invasive macro adenoma.



Fig 3:-Brain CT 1 month after cabergoline showing a pneumatocyst

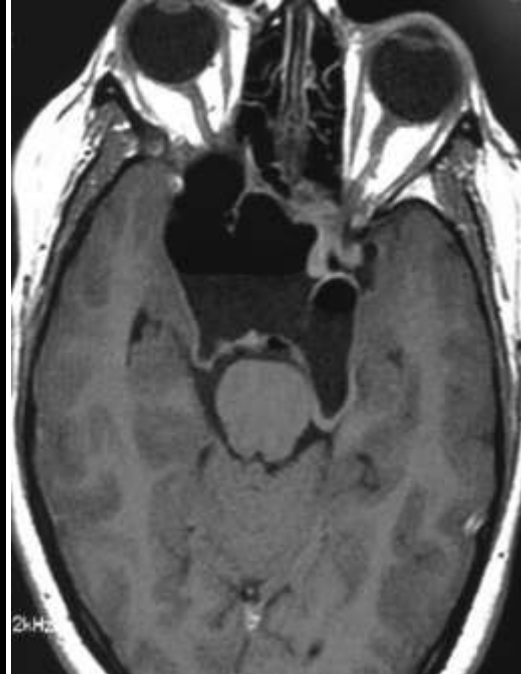


Fig 4: pituitary MRI 2 months after oral therapy showing the hydroaeric cavity

Discussion:-

Giant invasive macroprolactinomas are pituitary adenomas larger than 4 cm with significant extrasellar extension, very high prolactin levels exceeding 1000 MIU / L and no concomitant growth hormone (GH) or adrenocorticotrophin (ACTH) secretion [3]. They represent rare pituitary tumors which account for about 4% of all pituitary adenomas. They are frequently observed in men, in contrast to women who develop mainly micro prolactinomas, which is the first particularity of our case, this gender difference can be explained by the longer delay in diagnosis in men, because of the poorly understood symptoms, it is also due to a high frequency of rapidly growing tumors in men comparatively to women, which are often invasive and frequently bromocriptine resistant [4].

Cerebrospinal fluid (CSF) rhinorrhea refers to the drainage of cerebrospinal fluid through the nose, it is a life-threatening entity that occurs as a result of a basal skull bone defect with interruption of arachnoid and dural membrane. It is commonly classified as traumatic and non traumatic CSF including spontaneous and cabergoline induced ones [5]. CSF rhinorrhea as a complication of dopamine agonists treatment has been described previously as the result of rapid tumor shrinkage with a delay from 3 days to 17 months [6-9]. It can be misdiagnosed in patients with allergic rhinitis or other nasal discharge condition, which can lead to catastrophic complications of ascending meningitis, and can be easily differentiated by analysis of fluid for sugar and beta-2 transferrin, which is present in CSF [10].

However, the usual treatment is neurosurgery with trans-sphenoidal adenoma deblinking and repair of CSF leak by skull base reconstruction. In our patient we have managed to avoid surgery by depletive lumbar puncture, diamox and prophylactic antibiotics, which represent the second particularity of our case.

The other particularity of our case, is the occurring of a pneumocephalus, also known as pneumatocele, pneumatocele or intra cranial aerocele, it refers to the presence of an intracranial gas collection. It was described for the first time by Thomas in 1866 during the autopsy of a patient with trauma [11]. Pneumocephalus can have several other causes such as neurosurgical and oto-rhino-laryngological procedures, infection by gas-forming organisms, parasellar or paranasal sinus malignancies, and other less frequent causes [12].

Only very few cases of pneumocephalus, as a complication of a giant invasive macroprolactinoma few weeks following the initiation of medical treatment with cabergoline, have been described in the literature [13-16].

Pneumocephalus is due to a fistula created by the invasion of a giant macroprolactinoma, which destroys the basal skull bone, this fistula allows the communication between extra and intracranial spaces, additionally cabergoline induced tumor shrinkage enables the ambient air to enter the intracerebral compartments creating an air cavity.

Two hypotheses may explain its development. The first is the one-way valve which requires positive pressure events, such as sneezing and coughing, this event allows the air to enter causing tension pneumocephalus [17]. The second one is the inverted bottle theory, according to which CSF leak creates a negative intracranial pressure gradient, with influx of air [18].

Conclusion:-

Prolactinomas are the only pituitary adenoma which can erode the bone. Cabergoline induced tumor shrinkage will unmask the skull base defects, leading to CSF leak and / or pneumatocyst. Non-recognition can lead to severe complications such as meningitis. Both clinicians and patients should be aware of symptoms allowing early diagnosis and treatment.

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