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TIẾP CẬN MỘT TRƯỜNG HỢP ĐAU ĐẦU, SỤP MI, HỘI CHỨNG TOLOSA-HUNT?

BS Hoàng Gia Vinh - MEDIC Kiên Giang

Hành chánh:

Họ và tên: Nguyễn Thị T , nữ, NS: 1952, ID: 2400063222

Địa chỉ: Ấp Hiệp Trung, Xã Mỹ Hiệp Sơn, Hòn đất

Ngày khám 18/03/2024

Lý do khám: Đau 1/2 đầu phải, sụp mi.

Bệnh sử-khám LS:

- Bệnh khởi phát khoảng 10 ngày với đau 1/2 đầu phải, đau trong hốc mắt phải và sụp mi mắt phải ➔ BV Bình An
- TS: tăng huyết áp uống thuốc thường xuyên.
- Khám:

-Đau nhiều trong hốc mắt phải, sụp mi mắt phải, liệt dây III phải, đồng tử mắt phải dãn, PXAS kém so với mắt trái.

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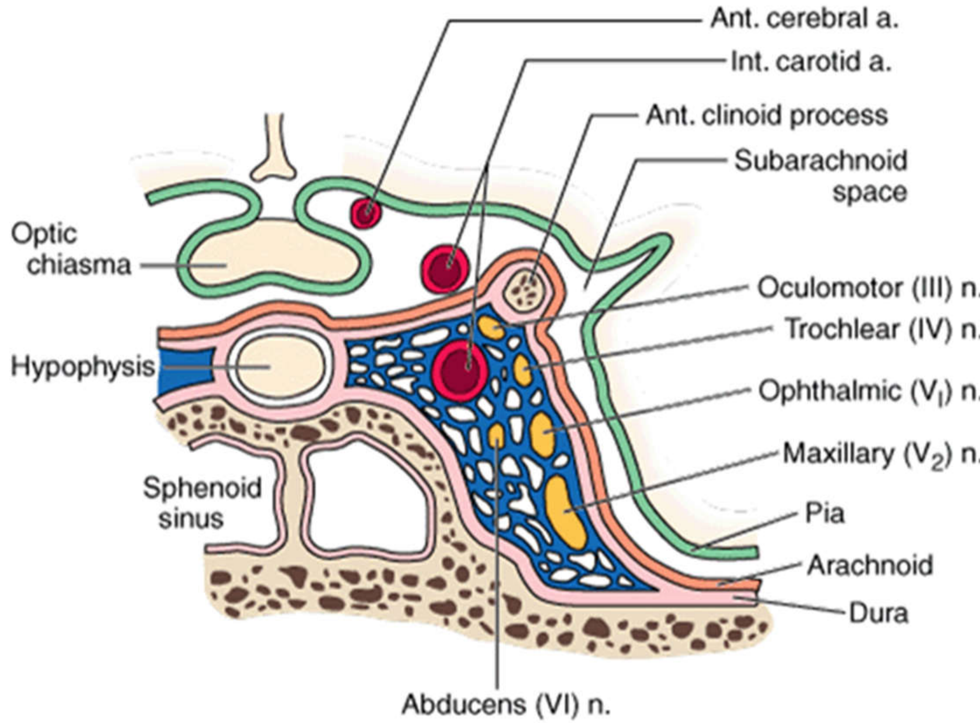
Ant. cerebral a.
Int. carotid a.
Ant. clinoid process
Subarachnoid space
Optic chiasma
Oculomotor (III) n.
Trochlear (IV) n.
Hypophysis
Ophthalmic (V_1) n.
Maxillary (V_2) n.
Sphenoid sinus
Pia
Arachnoid
Dura
Abducens (VI) n.

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Tolosa-Hunt Syndrome

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Assigned editor:

Summary

Tolosa-Hunt syndrome describes episodic orbital pain associated with paralysis of one or more of the third, fourth and/or sixth cranial nerves, which usually resolves spontaneously but can relapse and remit.

History

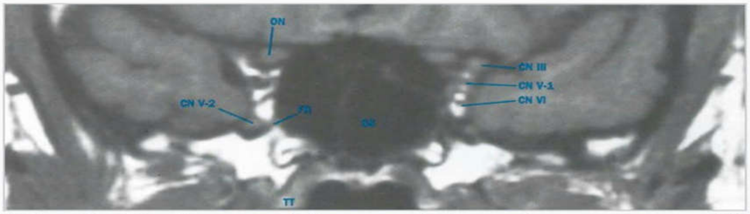
It was first described in 1954 by Tolosa, who found granulomatous inflammation in the cavernous sinus during autopsy of a patient with severe left-sided trigeminal pain and total ophthalmoplegia[1]. In 1961, Hunt reported 6 cases of unilateral painful ophthalmoplegia that tested negative with angiography and lumbar puncture and rapidly resolved with steroids.[1]

Disease Entity

Tolosa-Hunt syndrome was first classified by the International Headache Society in 2004 and now is a part of Classification ICHD-3. ICD-10 for Tolosa Hunt Syndrome is H49.40.

Disease

Unilateral orbital or periorbital pain with paresis of third, fourth and/or sixth cranial nerves secondary to idiopathic inflammation of the cavernous sinus, superior orbital fissure or orbit.



Tolosa Hunt Syndrome would be seen as inflammation, enhancentment or thickening localized in this area (labels). *MRI brain, coronal, TI-weighted, showing the Cavernous Sinus labeled. Courtesy of AAO multimedia.* CN= Cranial Nerve, CN III = Oculomotor Nerve, CN V•I = Trigeminal Nerve-1st Division, CN V•2 = Trigeminal Nerve-2nd Division

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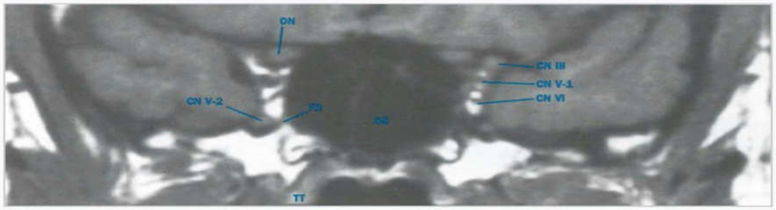

Tolosa Hunt Syndrome would be seen as inflammation, enhancentment or thickening localized in this area (labels). *MRI brain, coronal, TI-weighted, showing the Cavernous Sinus labeled. Courtesy of AAO multimedia.* CN= Cranial Nerve. CN III = Oculomotor Nerve, CN V•I = Trigeminal Nerve-1st Division, CN V•2 = Trigeminal Nerve-2nd Division, CN VI = Abducens Nerve, FR = Foramen Rotundum, ON = Optic Nerve, SS = Sphenoid Sinus, TT = Torus Tubarius. The nerves cannot be labeled with certainty because of their shifting relationships within the cavernous sinus. Note the superomedial course of ON as it passes from the orbit into the brain. Posterior to the point where the inferior border of ON exits the optic foramen, the superior border is covered by a layer of dura, the falciform fold. On TI-weighted scans, the sphenoid bone, air in SS, and dura forming the falciform fold overlying the nerve all have a similar appearance. TT (or tubal elevation) is comprised of cartilage arising from the medial pterygoid process; it marks the nasopharyngeal opening of the auditory (eustachian) tube.

The ICHD-3 Diagnostic criteria for Tolosa-Hunt syndrome are as follows[2]:

A. Unilateral orbital or periorbital headache fulfilling criterion C

B. Both of the following:

a. Granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit, demonstrated by MRI or biopsy

b. Paresis of one or more of the ipsilateral third, fourth, and/or sixth cranial nerves.

C. Evidence of causation demonstrated by both of the following:

a. Headache is ipsilateral to the granulomatous inflammation

b. Headache has preceded paresis of the third, fourth, and/or sixth nerves by ≤2 weeks, or developed with it.

D. Not better accounted for by another ICHD-3 diagnosis (see

b. Headache has preceded paresis of the third, fourth, and/or sixth nerves by ≤2 weeks, or developed with it.

D. Not better accounted for by another ICHD-3 diagnosis (see differential diagnosis section for more detail)

Of note:

Some reported cases of Tolosa-Hunt syndrome had additional involvement of the trigeminal nerve (commonly the first division) or optic, facial or acoustic nerves. Sympathetic innervation of the pupil is occasionally affected.

The syndrome has been caused by granulomatous material in the cavernous sinus, superior orbital fissure or orbit in some biopsied cases.

Careful follow-up is required to exclude other possible causes of painful ophthalmoplegia.

Etiology

Tolosa-Hunt syndrome is idiopathic.

Risk Factors

A possible risk factor for Tolosa-Hunt syndrome is a recent viral infection.

Pathophysiology

Tolosa-Hunt syndrome is idiopathic, sterile inflammation of the cavernous sinus. Its pathology is described as fibroblastic, lymphocytic, and plasmocytic infiltration of the cavernous sinus. Granulocytic and giant-cell infiltrations have also been described. Pathology may extend to involve the superior orbital fissure (sphenocavernous or parasellar syndrome) or orbital apex and affect the optic nerve. Involvement of cranial nerves III, IV, and VI, the superior division of the fifth cranial nerve, as well as the sympathetic fibers in the cavernous ICA or parasympathetic fibers that surround the oculomotor nerve can occur secondary to granulomatous inflammation.

> Diagnosis

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Diagnosis

Symptoms

Painful ophthalmoparesis or ophthalmoplegia is the hallmark of Tolosa-Hunt syndrome[3]. The patient may complain of double vision worse at distance, headaches, dizziness, nausea, neck stiffness, photophobia, blurred vision, and a "boring" pain may be associated with the headache.

Physical Exam

In addition to the standard ophthalmic examination of the patient including vision, IOP, pupil check for APD and nystagmus, slit-lamp and dilated fundus exam, a complete sensorimotor exam should be done. This includes oculomotor exam (to check for esotropia, exotropia, hypertropia or hypotropia), ductions, vergence, saccades, pursuit, and head tilt/turn. A common finding is abduction deficit associated with esodeviation that increases with gaze to the affected side. Lids should be checked for ptosis or lid retraction or any change in lid aperture during eye movements (to check for aberrant regeneration). Lid strength, fatigue or variability should be noted. Facial sensation should be checked. Stereopsis and color plates should also be evaluated.

Clinical diagnosis

Involvement of multiple contiguous cranial nerves strongly suggest a lesion in the cavernous sinus or subarachnoid space. Only one nerve may be involved, most likely the sixth cranial nerve, which is the only one not protected within the dural wall of the cavernous sinus.

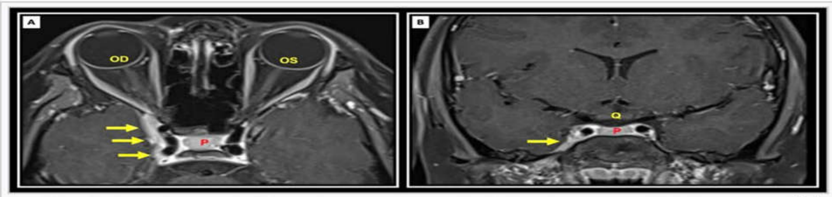
In addition to the complete ophthalmic exam as described, the physician must closely look for Horner syndrome, facial hypoesthesia or engorgement of ocular surface vessels, orbital

Diagnostic Procedures

The most appropriate imaging includes MRI /MRA (DWI series) which provides information about the cavernous sinus and orbital apex in greater detail than a CT. MRI may be able to provide detail of granulomatous inflammation, aiding in formal diagnosis of Tolosa-Hunt syndrome. However, these results may be unreliable. Biopsy can also be used to demonstrate granulomatous inflammation and may be more reliable, but the procedure may be more difficult[3]

A CTA w/ and w/o contrast can also be obtained if an MRI/MRA is not available. A lumbar puncture may be done to check for opening pressure and CSF should be evaluated for infection/oligoclonal bands.

Recent evidence supports the use of High resolution 3D skull base MRI with isotropic constructive interference in steady state (CISS) and 0.6-mm cut images with and without contrast as effective way to visualize cranial nerves and cavernous sinus lesions that were not previously visualized.[4]



MRI orbits with and without contrast, axial(A) and coronal (B) sections, showing enhancement of the right cavernous sinus (arrows) in a patient with right third and sixth abducens painful palsy. P- Pituitary gland. Q- Quiasm. OD- Right and OS- Left eye. Direct contribution of Claudia Maria Prospero Ponce MD.

Laboratory Tests

Work up should include tests that can rule out the various diseases listed above given the history and context of the patient. This can include

Serum

- CBC w diff
- ESR

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- Glucose
- Protein
- Cell count and differential (WBC, RBC)
- Cytology
- Culture
- Gram stain
- Oligoclonal bands

In few recent reports, associations with Pathogenic Nucleotide-Binding Oligomerization Domain 2 Polymorphism[6] and COVID-19 vaccination [7] have been reported.

Differential Diagnosis

Tolosa-Hunt syndrome is considered a diagnosis of exclusion. Thus, the following entities must be considered and ruled out before a diagnosis of Tolosa-Hunt syndrome is made:

- Ischemic disease: Hemorrhage, ischemic mononeuropathy
- Infectious process: post viral syndrome, chronic inflammation of petrous bone (recurrent ear infections), syphilis, basal meningitis
- Anatomical malformation: aneurysm, AVM, carotid-cavernous fistula, cavernous sinus thrombosis, pseudotumor cerebrii, Duane syndrome/Moebius syndrome, Chiari malformation
- Inflammatory disease: Sarcoidosis, granulomatosis with polyangiitis (formerly Wegener's), Behcet's disease, IgG4 Disease
- Autoimmune condition: myasthenia gravis, thyroid disease, lupus
- Neoplastic disease: meningioma, neurogenic tumor, hemangioma, lymphoma/leukemia, schwannoma, pituitary adenoma, metastasis, CPA lesion, nasopharyngeal carcinoma, chordoma, chondrosarcoma, brain stem glioma in children
- Demyelinating disease: MS
- Others: Diabetes mellitus, Head trauma, BBPV, Meniere's, ophthalmoplegic migraine

Management

Oral steroids are the mainstay of treatment. Both symptoms and

Management

Oral steroids are the mainstay of treatment. Both symptoms and physical exam findings (headache, ptosis, ophthalmoplegia, etc) can be expected to resolve rapidly with an oral steroid taper regimen over 3-4 months. The patient can be co-managed with the Neurology service to rule out other entities listed in the differential diagnosis of Tolosa-Hunt syndrome.

Before starting steroid, fungal infection of the orbit with fungal sinusitis (mucormycosis in diabetic/immunocompromised) is a differential diagnosis that must be considered, because in that case starting steroid will worsen the disease.

Several therapies have been investigated for instances of steroid-resistant Tolosa-Hunt syndrome. A case report by Lee, et al. has been published in which gamma-knife radiation therapy is used to provide marked short-term relief without relapse of symptoms[8]. Antimetabolic agents such as methotrexate, infliximab, and mycophenolate mofetil have also been shown to cause dramatic improvement in patients whom are deemed to be steroid-resistant[9][10][11]

Complications

A rare complication is aberrant regeneration of cranial nerves.

Prognosis

Prognosis of Tolosa-Hunt syndrome is excellent. Full recovery is expected with steroid treatment. The disease may have a relapsing-remitting course.

References

1. ↑ 1.0 1.1 Tolosa E. Periarteritic lesions of the carotid siphon

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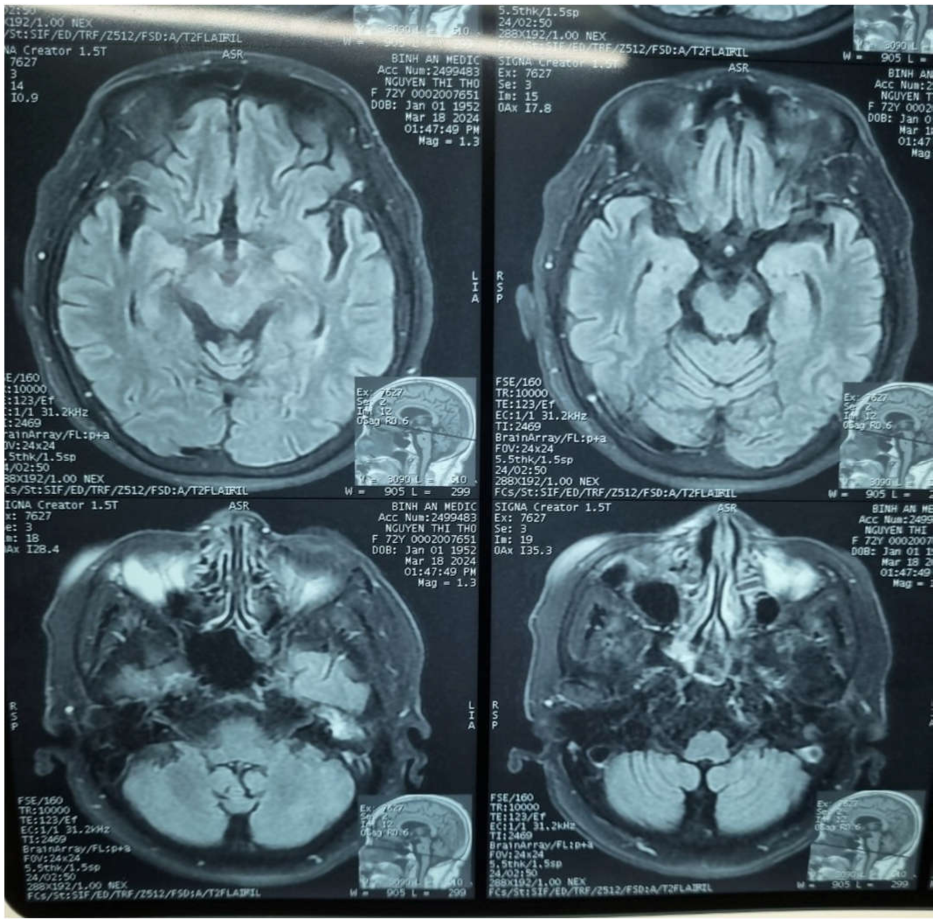

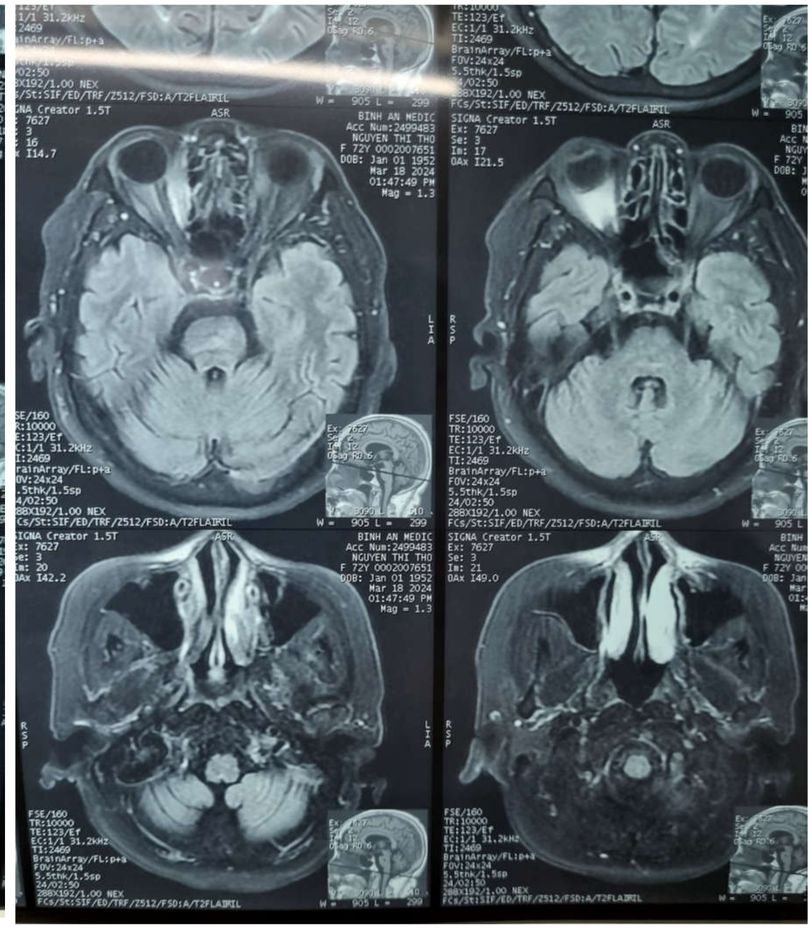

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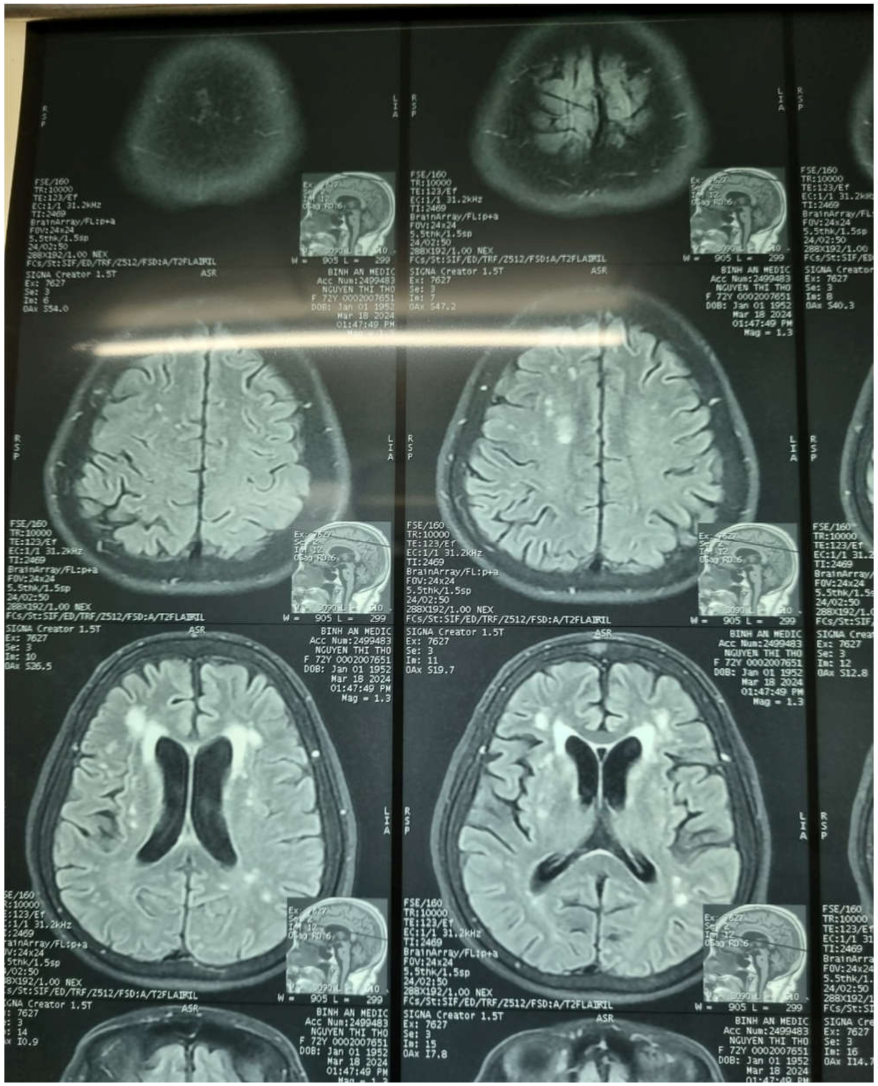

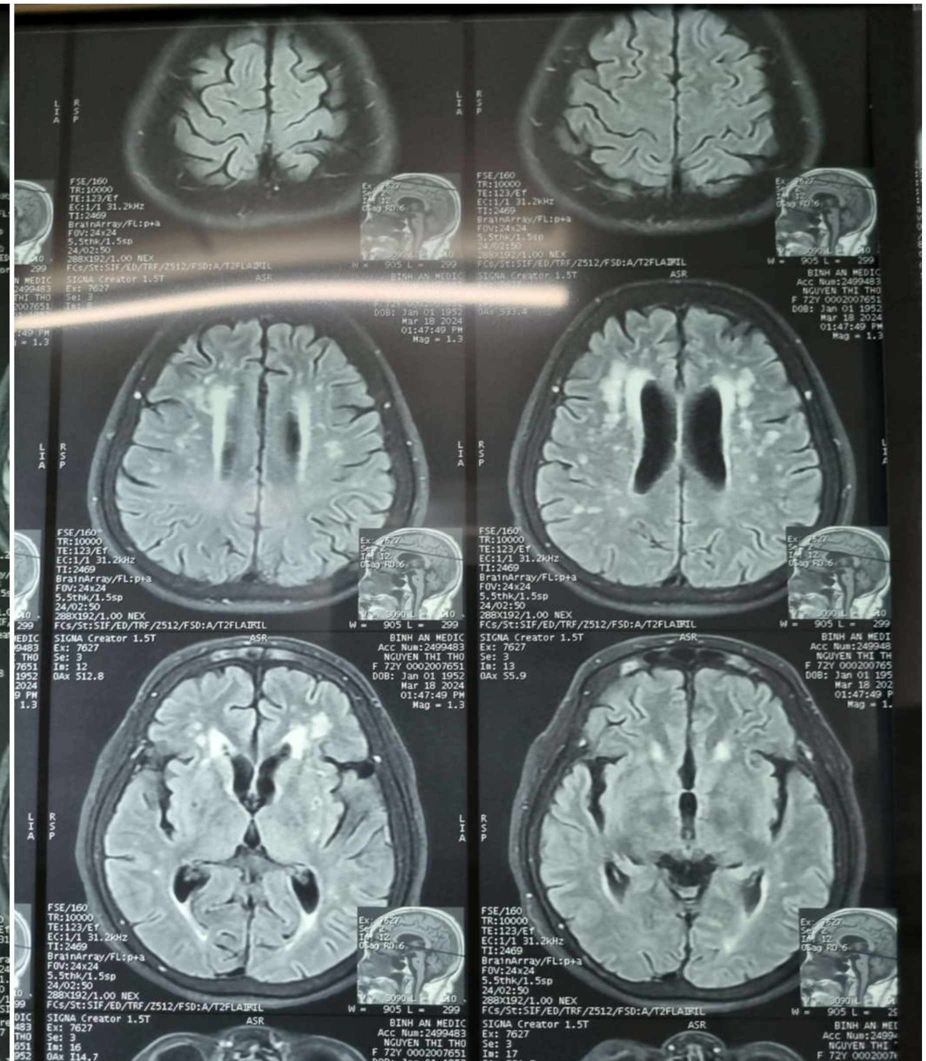

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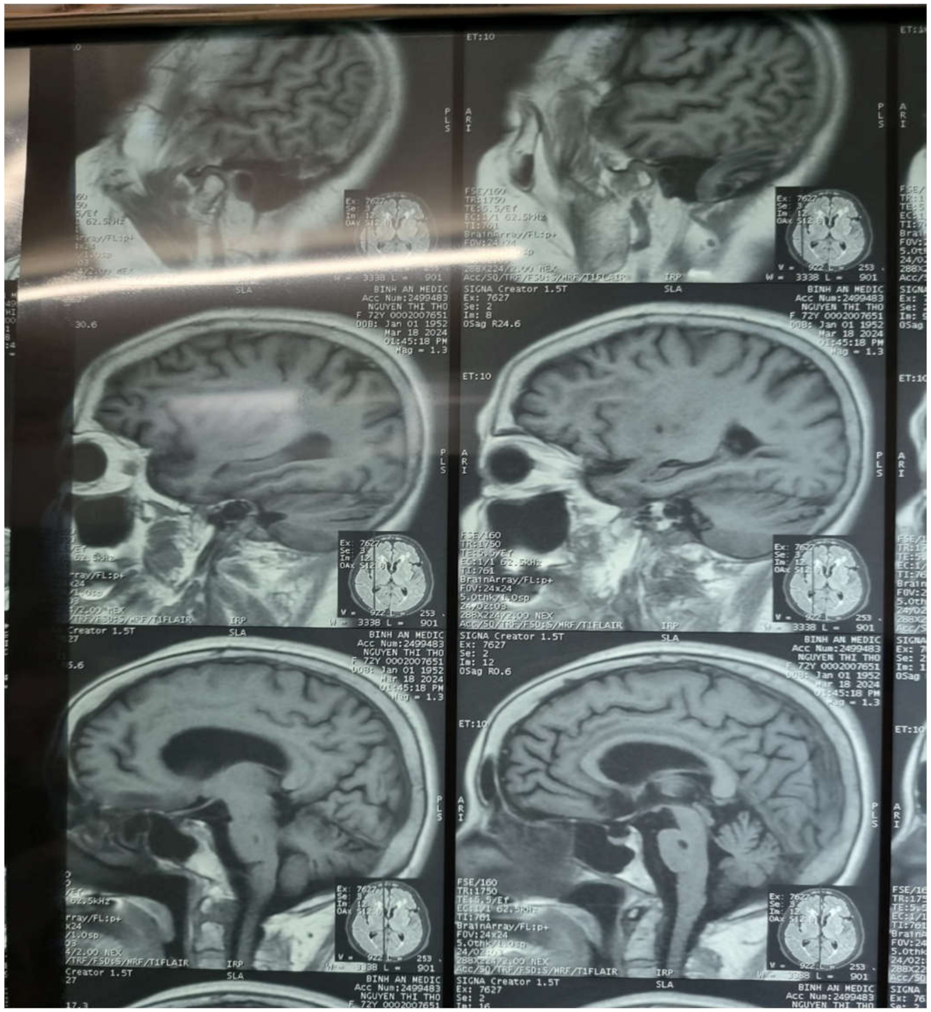

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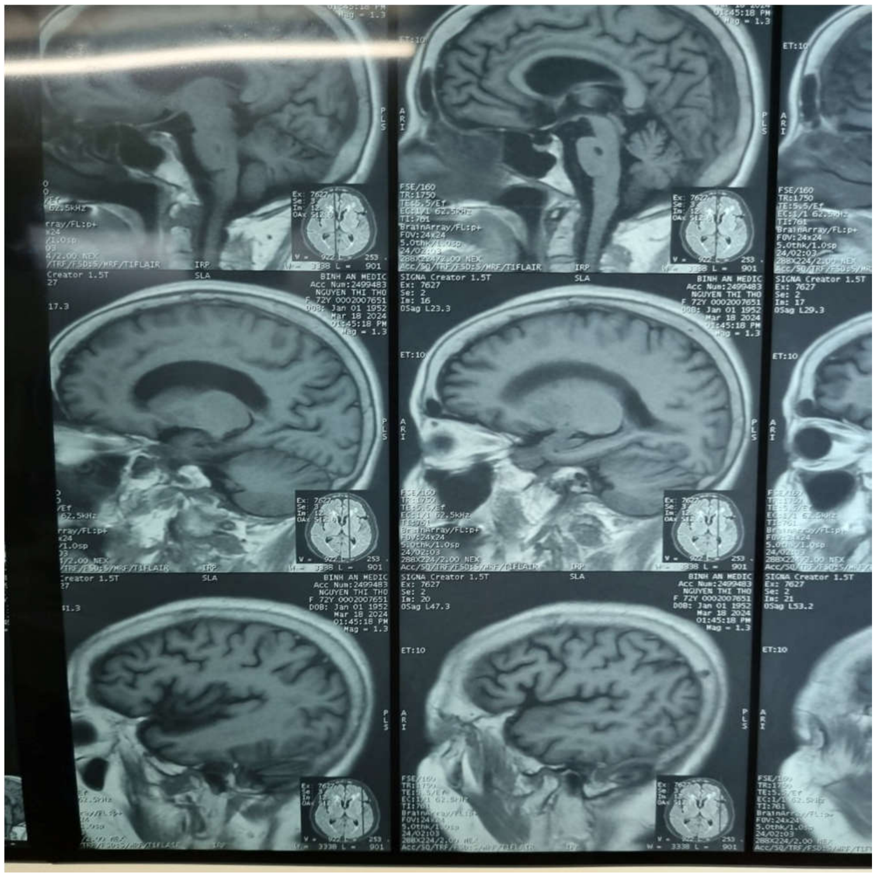

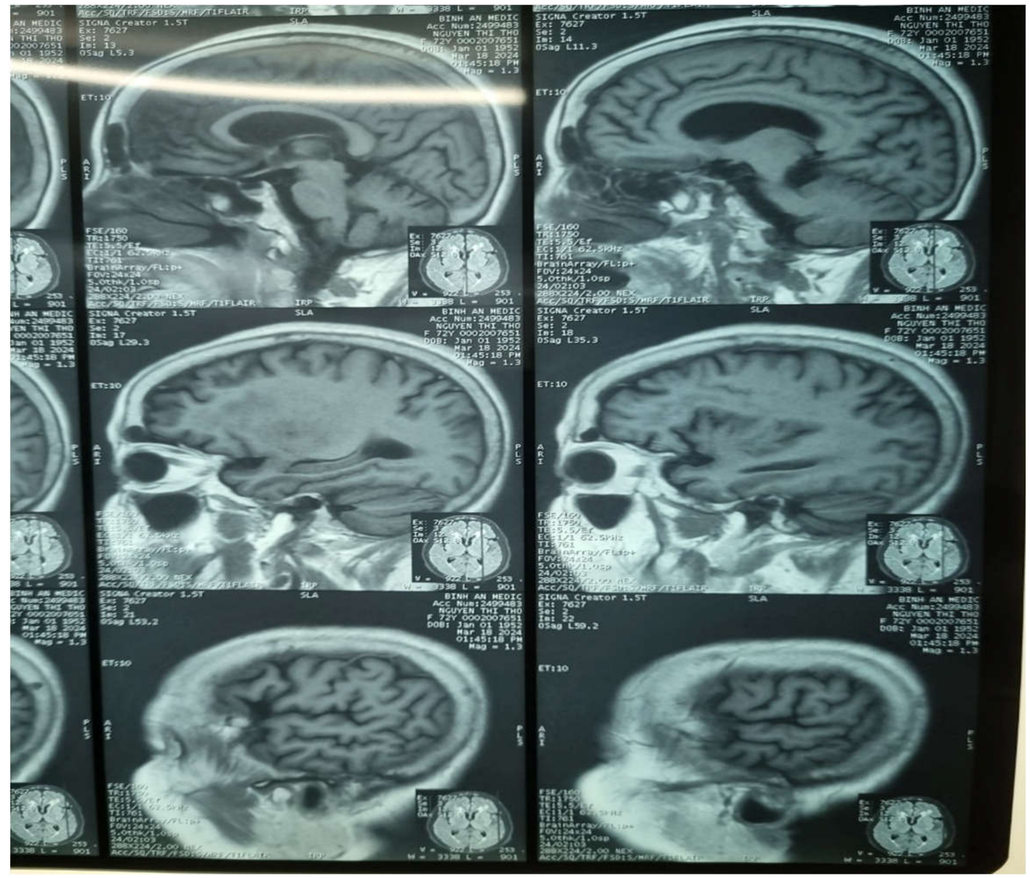

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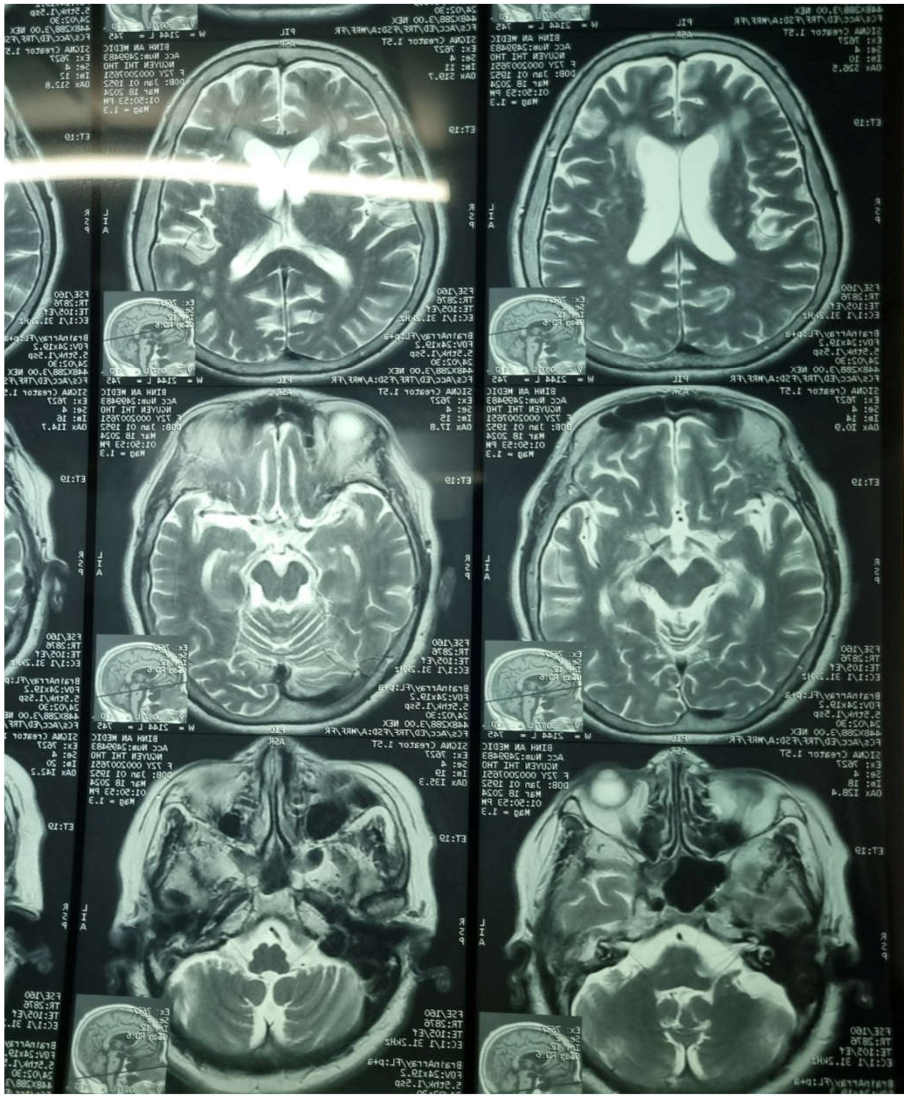

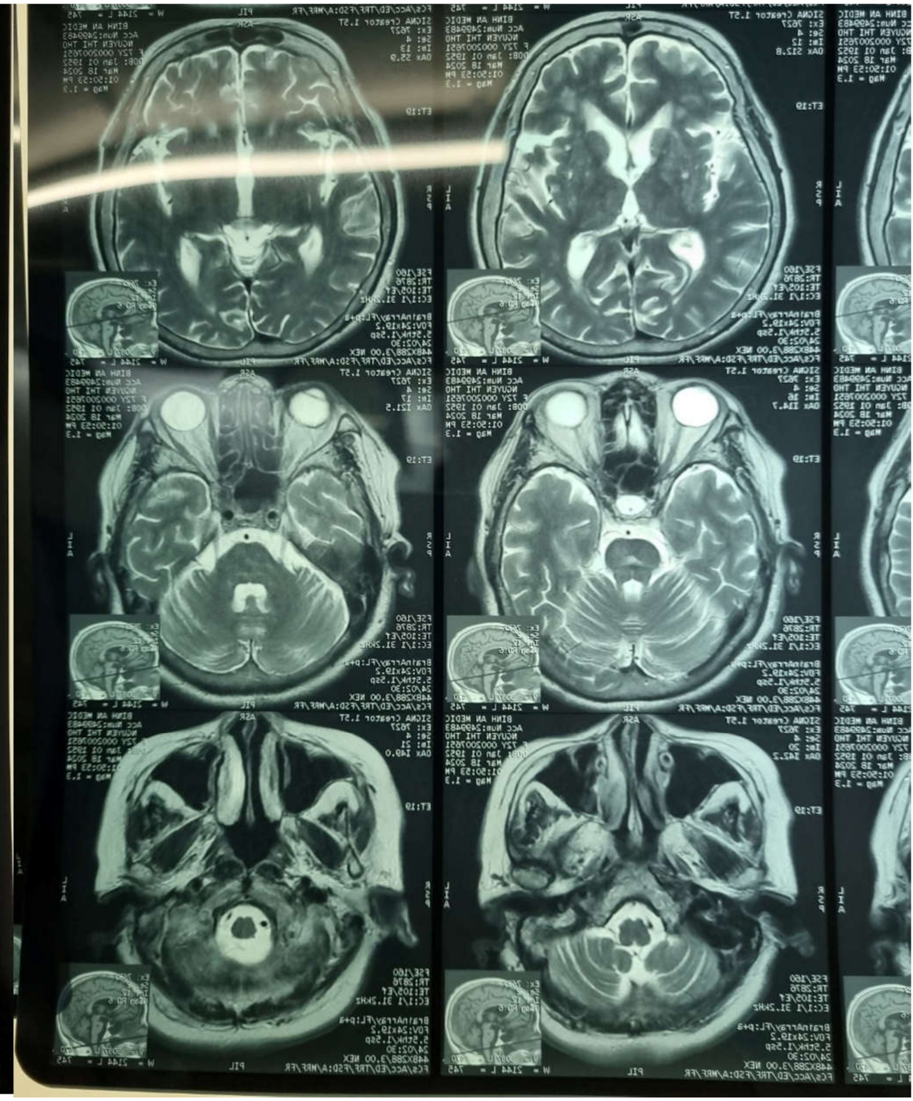

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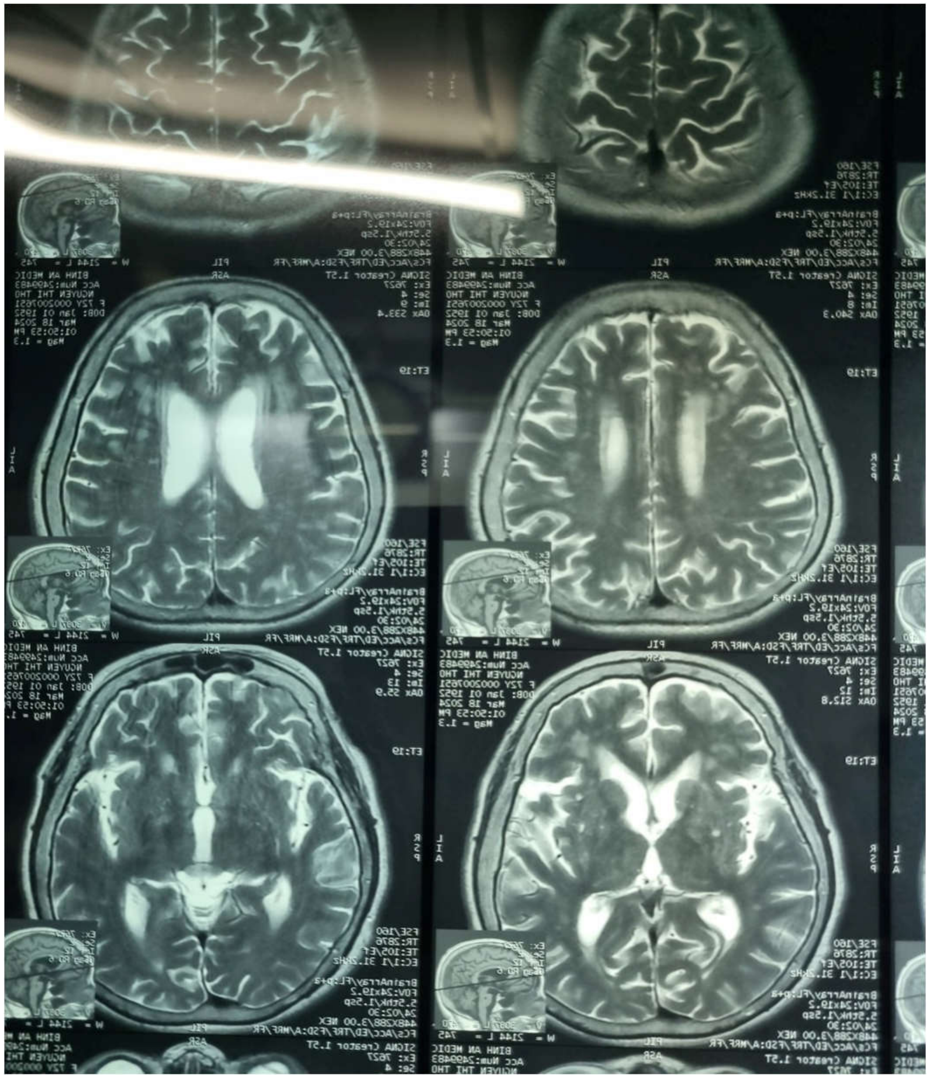

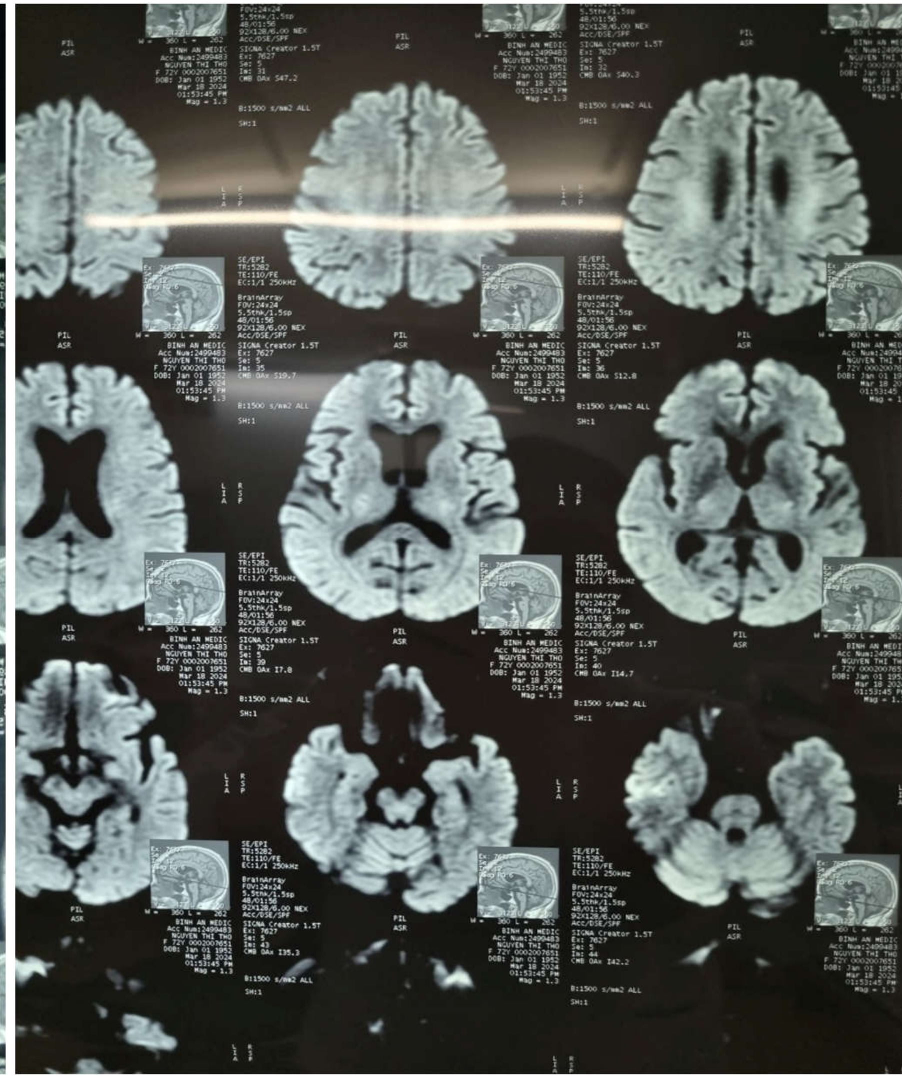

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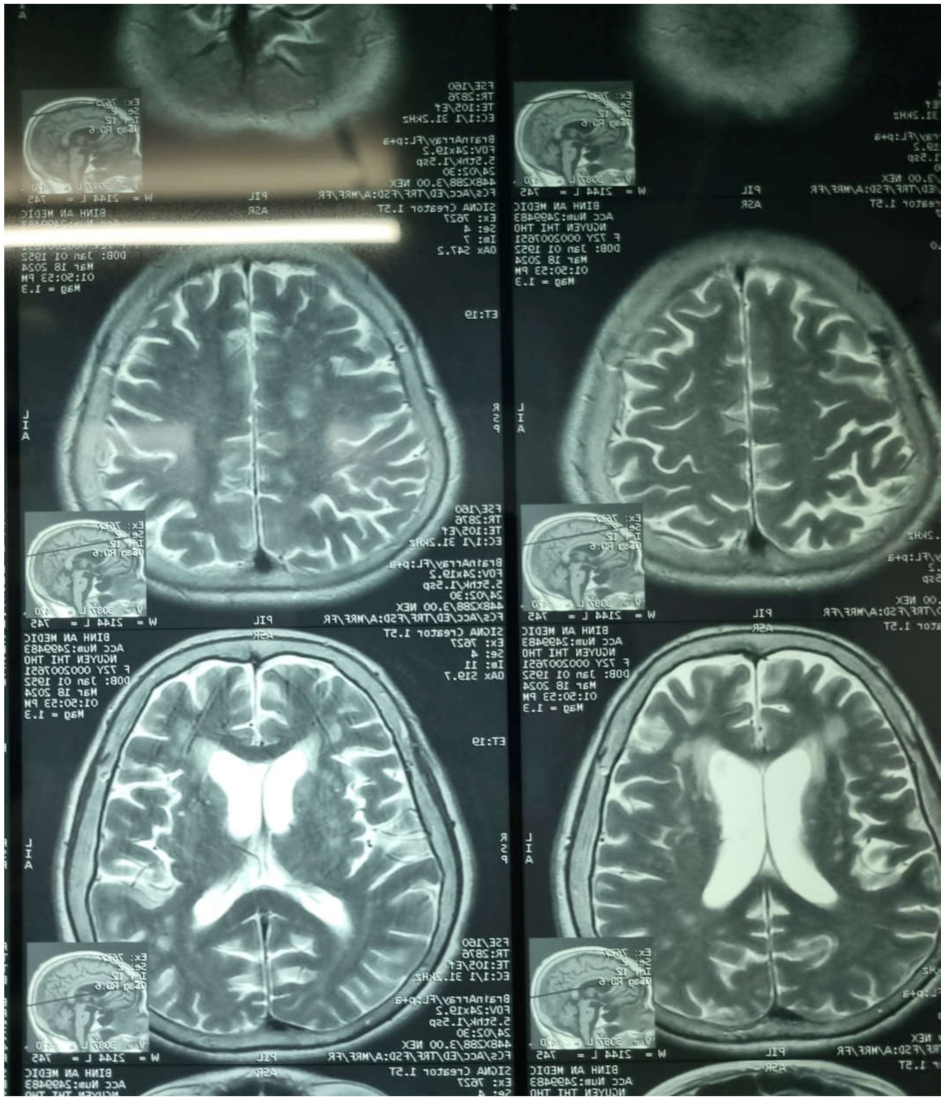

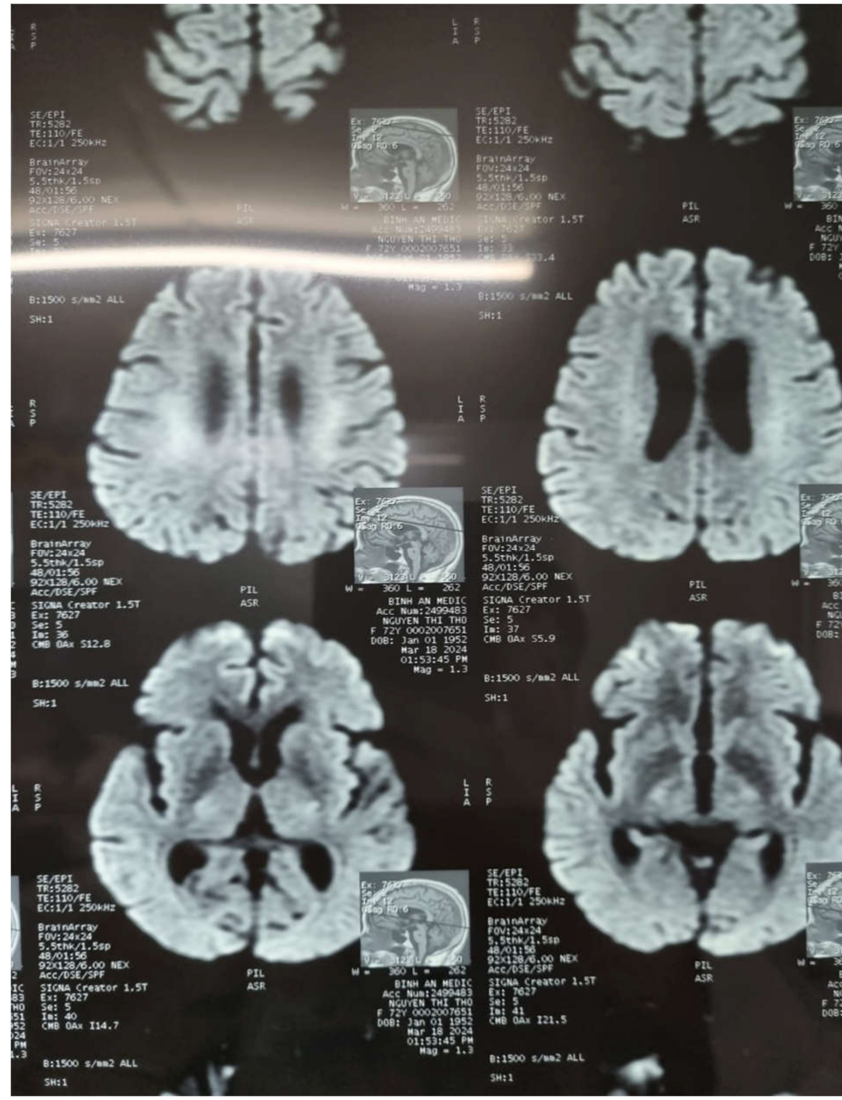

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BỆNH VIỆN BÌNH AN MEDIC KIÊN GIANG

80-82 Nguyễn Trung Trực - Phường Rạch Giá - An Giang

PHIẾU KẾT QUẢ CHỤP MRI

Mã KCB: 2400063222 **Họ tên:** **ĐT:** **Năm sinh:** 1952 **Phái:** Nữ
Địa chỉ: **Ngày chụp:** 18/03/2024
Lâm sàng: liệt dây III, VI , sụp mi mắt phải, đau đầu kéo dài, theo dõi tai biến mạch máu não, khối choáng chỗ nội sọ
Chẩn đoán: H49.0-Liệt dây thần kinh [vận nhãn] III(liệt dây III, IV phải theo dõi HC tolosa -hunt);I69.3-Di chứng nhồi máu não
BS chỉ định: BS Hoàng Gia Vinh **Khoa/Phòng:** Nội Thần Kinh & EEG **Đối tượng:** BHYT
Nơi làm việc:

KỸ THUẬT: Chụp cộng hưởng từ sọ não (0.2-1.5T) [không có chất tương phản]

Não bệnh nhân được khảo sát qua các mặt cắt theo những chuỗi xung: Sagittal T1WI; Axial T2WI, Axial T2 FLAIR, DWI. Không tiêm thuốc tương phản.
XOANG:
- Các xoang hàm, sàng, trán, bướm hai bên không viêm.
- Vòm hầu và các khoang cạnh hầu hai bên trống.
- Viêm tai xương chũm trái.

HỐ SAU:
- Tổn thương não cũ cầu não trái.
- Não thất 4 ở giữa, không dãn.
- Các bể dịch não tủy quanh thân não bình thường.

TẦNG TRÊN LỀU:
- Tổn thương dạng nốt chất trắng dưới vỏ vùng trán, đỉnh hai bên, tăng tín hiệu trê T2WI, T2FLAIR.
- Hệ thống não thất bình thường về hình dạng, tín hiệu, kích thước.
- Cấu trúc đường giữa không lệch. Rãnh vỏ não bình thường.

Kết luận:

Không thấy khối choán chỗ nội sọ.
Tổn thương não cũ cầu não trái.
Tổn thương dạng nốt chất trắng dưới vỏ vùng trán, đỉnh hai bên. Bệnh lý chất trắn do bệnh lý mạch máu nhỏ.
Viêm tai xương chũm trái.

Đề nghị:

Kết hợp lâm sàng.

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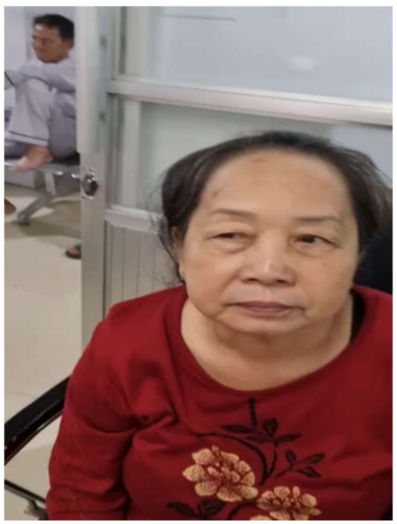

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