

Demyelinating Dz. of CNS (Dr. Eyad)

* Case A chief complaint and history :-

- A 16 years old female patient previously healthy presented initially with rapid progressive 3-5 hours bilateral loss of vision.
- History of URTI two weeks ago.
- she was admitted in Medical Department - M. Hospital.
- Initial Diagnosis: Acute CVA in Basilar artery.
- ophthalmological Examination: no signs of optic neuritis.
- Brain MRI → normal
- Brain MRI angiography → normal
- Follow up ophthalmological Examination → signs of acute bilateral optic neuritis with total bilateral loss of vision.

acute bilateral optic neuritis with total bilateral loss of vision
Demyelinating Dz

two days after admission, she developed:

1. paresthesia in both lower limbs, Rt & Lt
2. Decrease deep sensation in both LLs Rt & Lt
3. Decrease pin & temperature sensation in both LLs Lt & Rt
4. Urine retention and incontinence.

→ Neurological examination :-

- 1) Total bilateral loss of vision
- 2) ↓ lower limbs deep tendon reflexes Rt & Lt

2) Lower flaccid paraparesis "shock stage" Rt & Lt

4) A sensory level "hyposthesia" was noted below C7-T1 Lt & Rt.

Investigation :-

- CBC, ESR
- Full blood chemistry
- TFTs
- PTT, PT, INR
- CRP, RF
- ANA, DNA (Ds) antibodies
- ANCA
- Anti-cardiolipin IgM and IgG → antiphospholipid
- Lupus anticoagulant
- HBsAg, HCV
- vita B12
- Urine analysis.
- Normal ← Investigation 11/1

CSF analysis :-

CSF cell count → WBC - 12 (L: 70%, N: 30%)

RBC : 0

CSF protein : 116

CSF sugar : 89

CSF culture : negative

chest X-ray → Normal.

Sarcoidosis : not

Cervical spine MRI →

↑ ↓ ↓ ↓ ↓ Demyelination 2' supratentorial & also in spinal cord & in right side

(Final Diagnosis) → Neuromyelitis Optica
(Devic's Dz)

2/11

Treatment of Acute D₂ attack in

- Treatment of Acute D₂ attack :-
- 1) steroid : • pulse steroid therapy : 1g Methylprednisolone daily for Five days.
 - 2) IVIG : 400 mg/kg daily for 5 days.
 - 3) A proton pump inhibitor.
 - 4) Prophylactic Anticoagulant → Heparin
 - 5) Symptomatic therapy.
 - 6) Physiotherapy.
1. University Hospital for therapeutic

↳ symptomatic therapy.
↳ physiotherapy.
→ she was transferred to Al Najah national university hospital for therapeutic plasma exchange.

- Five cycles was done.
- long-term Treatment → Immunos "Azathioprine" 50mg 1*2 cicles
→ Hyporic "Mycophenolate" 360 2*2 ✓ 110 days

(Classification of the Acquired Demyelinating Dz of CNS)

- 1) Multiple sclerosis "Disseminated Sclerosis".
- 2) Transverse myelitis.
- 3) Acute Disseminated Encephalomyelitis "ADEM".
- 4) Neuromyelitis optica "NMO" (Devic's Del).
- 5) Acute & subacute necrotizing hemorrhagic leukoencephalitis.

- ★ Transverse myelitis

↳ segmental spinal cord injury caused by acute inflammation.

- Uncommon Dz → 1-5 cases per million.
- Most common cause is idiopathic result from an autoimmune process.
viral illness, upper RTI, history of infection, etc.
- The inflammation of the spinal cord is generally restricted to one or two segments usually in the thoracic cord.
- symptoms typically developed rapidly over several hours.
weakness & sensory disturbance below the level of inflammation.
- direct damage of spinal cord is rare.

damage of spinal cord 1. 2. 3. 4. 5. 6. 7. 8. 9. 10. 11. 12. 13. 14. 15. 16. 17. 18. 19. 20. 21. 22. 23. 24. 25. 26. 27. 28. 29. 30. 31. 32. 33. 34. 35. 36. 37. 38. 39. 40. 41. 42. 43. 44. 45. 46. 47. 48. 49. 50. 51. 52. 53. 54. 55. 56. 57. 58. 59. 60. 61. 62. 63. 64. 65. 66. 67. 68. 69. 70. 71. 72. 73. 74. 75. 76. 77. 78. 79. 80. 81. 82. 83. 84. 85. 86. 87. 88. 89. 90. 91. 92. 93. 94. 95. 96. 97. 98. 99. 100. 101. 102. 103. 104. 105. 106. 107. 108. 109. 110. 111. 112. 113. 114. 115. 116. 117. 118. 119. 120. 121. 122. 123. 124. 125. 126. 127. 128. 129. 130. 131. 132. 133. 134. 135. 136. 137. 138. 139. 140. 141. 142. 143. 144. 145. 146. 147. 148. 149. 150. 151. 152. 153. 154. 155. 156. 157. 158. 159. 160. 161. 162. 163. 164. 165. 166. 167. 168. 169. 170. 171. 172. 173. 174. 175. 176. 177. 178. 179. 180. 181. 182. 183. 184. 185. 186. 187. 188. 189. 190. 191. 192. 193. 194. 195. 196. 197. 198. 199. 200. 201. 202. 203. 204. 205. 206. 207. 208. 209. 210. 211. 212. 213. 214. 215. 216. 217. 218. 219. 220. 221. 222. 223. 224. 225. 226. 227. 228. 229. 230. 231. 232. 233. 234. 235. 236. 237. 238. 239. 240. 241. 242. 243. 244. 245. 246. 247. 248. 249. 250. 251. 252. 253. 254. 255. 256. 257. 258. 259. 260. 261. 262. 263. 264. 265. 266. 267. 268. 269. 270. 271. 272. 273. 274. 275. 276. 277. 278. 279. 280. 281. 282. 283. 284. 285. 286. 287. 288. 289. 290. 291. 292. 293. 294. 295. 296. 297. 298. 299. 300. 301. 302. 303. 304. 305. 306. 307. 308. 309. 310. 311. 312. 313. 314. 315. 316. 317. 318. 319. 320. 321. 322. 323. 324. 325. 326. 327. 328. 329. 330. 331. 332. 333. 334. 335. 336. 337. 338. 339. 340. 341. 342. 343. 344. 345. 346. 347. 348. 349. 350. 351. 352. 353. 354. 355. 356. 357. 358. 359. 360. 361. 362. 363. 364. 365. 366. 367. 368. 369. 370. 371. 372. 373. 374. 375. 376. 377. 378. 379. 380. 381. 382. 383. 384. 385. 386. 387. 388. 389. 390. 391. 392. 393. 394. 395. 396. 397. 398. 399. 400. 401. 402. 403. 404. 405. 406. 407. 408. 409. 410. 411. 412. 413. 414. 415. 416. 417. 418. 419. 420. 421. 422. 423. 424. 425. 426. 427. 428. 429. 430. 431. 432. 433. 434. 435. 436. 437. 438. 439. 440. 441. 442. 443. 444. 445. 446. 447. 448. 449. 450. 451. 452. 453. 454. 455. 456. 457. 458. 459. 460. 461. 462. 463. 464. 465. 466. 467. 468. 469. 470. 471. 472. 473. 474. 475. 476. 477. 478. 479. 480. 481. 482. 483. 484. 485. 486. 487. 488. 489. 490. 491. 492. 493. 494. 495. 496. 497. 498. 499. 500. 501. 502. 503. 504. 505. 506. 507. 508. 509. 510. 511. 512. 513. 514. 515. 516. 517. 518. 519. 520. 521. 522. 523. 524. 525. 526. 527. 528. 529. 530. 531. 532. 533. 534. 535. 536. 537. 538. 539. 540. 541. 542. 543. 544. 545. 546. 547. 548. 549. 550. 551. 552. 553. 554. 555. 556. 557. 558. 559. 560. 561. 562. 563. 564. 565. 566. 567. 568. 569. 570. 571. 572. 573. 574. 575. 576. 577. 578. 579. 580. 581. 582. 583. 584. 585. 586. 587. 588. 589. 590. 591. 592. 593. 594. 595. 596. 597. 598. 599. 600. 601. 602. 603. 604. 605. 606. 607. 608. 609. 610. 611. 612. 613. 614. 615. 616. 617. 618. 619. 620. 621. 622. 623. 624. 625. 626. 627. 628. 629. 630. 631. 632. 633. 634. 635. 636. 637. 638. 639. 640. 641. 642. 643. 644. 645. 646. 647. 648. 649. 650. 651. 652. 653. 654. 655. 656. 657. 658. 659. 660. 661. 662. 663. 664. 665. 666. 667. 668. 669. 670. 671. 672. 673. 674. 675. 676. 677. 678. 679. 680. 681. 682. 683. 684. 685. 686. 687. 688. 689. 690. 691. 692. 693. 694. 695. 696. 697. 698. 699. 700. 701. 702. 703. 704. 705. 706. 707. 708. 709. 710. 711. 712. 713. 714. 715. 716. 717. 718. 719. 720. 721. 722. 723. 724. 725. 726. 727. 728. 729. 730. 731. 732. 733. 734. 735. 736. 737. 738. 739. 740. 741. 742. 743. 744. 745. 746. 747. 748. 749. 750. 751. 752. 753. 754. 755. 756. 757. 758. 759. 760. 761. 762. 763. 764. 765. 766. 767. 768. 769. 770. 771. 772. 773. 774. 775. 776. 777. 778. 779. 780. 781. 782. 783. 784. 785. 786. 787. 788. 789. 790. 791. 792. 793. 794. 795. 796. 797. 798. 799. 800. 801. 802. 803. 804. 805. 806. 807. 808. 809. 810. 811. 812. 813. 814. 815. 816. 817. 818. 819. 820. 821. 822. 823. 824. 825. 826. 827. 828. 829. 830. 831. 832. 833. 834. 835. 836. 837. 838. 839. 84

complete TM

Incomplete TM

- Incomplete TM $\leftarrow$ spinal cord $\leftarrow$ partial segmental inflammation.
 - (5-10%) risk of MS. Complete TM $\leftarrow$ inflammation in spinal cord.
 - (much more common relevance to MS) $\leftarrow$ Incomplete TM $\leftarrow$ inflammation in spinal cord.
 - Follow up $\leftarrow$ MS.
- MS $\leftarrow$ incomplete TM $\leftarrow$ inflammation in spinal cord.
- MS $\leftarrow$ brain & spinal cord $\leftarrow$ MRI.
- MS $\leftarrow$ CSF $\leftarrow$ oligoclonal bands.

Etiology of Transverse Myelitis or

$\rightarrow$ Most causes of TM is idiopathic results from autoimmune process.

- occur as the first manifestation of MS (Incomplete TM).
- viruses (measles, EBV, HZV, SV, HIV)
- Mycoplasma pneumoniae
- SLE & other vasculitis
- RA
- Antiphospholipid
- scleroderma

(Investigation of TM)

- CSF
 - $\rightarrow$ $\uparrow$ WBC
 - $\rightarrow$ $\uparrow$ Total protein
 - $\rightarrow$ positivity of oligoclonal bands.

(methylprednisolone) $\rightarrow$ acute stage of MS & TM. IVIG.

Beta Interferon $\rightarrow$ future attack & prevention. MS.

• MRI $\rightarrow$ For brain & spinal cord

$\rightarrow$ Hypertense & swollen at the level of inflammation in spinal cord.

MS $\leftarrow$ Brain $\leftarrow$ MRI.

(Treatment of TM)

① Medical therapy:

1) pulse therapy methylprednisolone : 1000mg IV daily for 5 days

2) IVIG : 400 mg/kg W daily for 5 days.

③ plasmapheresis

④ Anticoagulant for prevention of DVT & PE

② Medical rehabilitation and physiotherapy.

↳ Post infectious encephalomyelitis.

(viral) ← primary

← encephalitis : التهاب دماغ

لا علينا

(ADEM)

Secondary

(ADEM) ← secondary →
 ↘ Immune mediated process → autoimmune D.C.

- Acute immune-mediated demyelinating disorder of the brain & spinal cord.
- Occur often after vaccination/virus infection (Measles, influenza, EBV, HIV).
- Multifocal neurological signs & symptoms.

• كيف نرفع ADEM عن ال MS ١٩

diagnose for pertussis: viral infection & URTI risk history & ADEM 11 Oct 10 & -
• Vaccination

• Fever is constitutional symptom + is in ADEM " .

mood status changes
and seizures

في حال الإصابة cortical lesion

• کوئی نہ ADEM ملے گا

• اد ADEM بلون اكرزيه الوضاله ۱۱-۱۲ اي MS مي مي ۲۵-۴۵

(ADEM. Investigation of ADEM)

TM مقف

- CSF $\rightarrow$
 - $\uparrow$ WBC
 - $\uparrow$ total protein
 - Positive oligoclonal bands $\rightarrow$ (MS)

- Brain & spinal cord MRI

(Treatment of ADEN)

(Treatment of ADEM)

→ 1) Medical therapy: • pulse therapy methylprednisolone 1000 mg IV daily for 5 days.

- IV 16 : 400 mg/kg IV daily for 5 days or plasmapheresis

- Anticoagulant for protection.

2) Medical rehabilitation and physiotherapy.

— Neuromyelitis Optica "Devic's Dz" —

→ subacute autoimmune disorder characterized by demyelination & inflammation of both optic nerve & spinal cord "TM"

- bilateral optic neuritis

لا تفتك لازم يكون معنا ٢٢١ مع

- SLE, Behcet's Dz and Sarcoidosis produce a similar picture.
- NMO may be closely related to MS.

- * typically lesions extending over two-three spinal segment.
- * optic neuritis & TM can occur at the same time, or one may follow the other.
- classic NMO → Known as Devic's D2.
- up to 20-30% of cases, NMO attacks are preceded by infection or vaccination.

(Clinical features of neuromyelitis optica)

- Total, rapid bilateral visual loss.
- spinal cord symptoms "sensory & motor".
- Pain, paraesthesia & hypoaesthesia in limbs.
- Autoimmune sphincters dysfunction
- ^{have} poorer prognosis for recovery of vision than those with MS
- Disability following attacks of relapse.

Diagnosis of NMO (Wingerchuk criteria)

- | | |
|--|-----|
| • Bilateral optic neuritis | (1) |
| • TM | (2) |
| • Spinal cord MRI lesion extending over 2-3 spinal segment | (1) |
| • No brain MRI finding | (2) |
| • NMO-IgG seropositive | (2) |

CSF examⁿ →

- WBC ↑
- cytology
- protein, sugar
- oligoclonal bands oCB
- PCR.

Cell count is normal in 40% of NMO
oCB are positive in 20% of NMO.

(Main treatment of NMO)

- Remission & improvement of relapse
- long term stabilization of D2
- symptomatic therapy of residual symptoms.

Treatment of acute Dz attacks :-

• جی ڈی سی

Long term treatment of NMO :-

1. → Azathioprine . First
2. Mycophenolate → second
3. combination therapy → third

↑
↓
↑
↓
liver

(Acute & Subacute -) →

↳ Rare

- It is regarded as a very acute form of postinfectious disseminated encephalomyelitis
- clinical picture → decrease of conscious level & multifocal signs & symptoms
- pathology → perivascular infiltration, hemorrhage / Demyelination & necrotizing changes in vessels

Diagnosis

↳ CSF → ↑ CSF pressure, lymphocyte & erythrocyte

↳ Brain MRI →

Brain biopsy →

Treatment → High dose methylprednisolone.