Case 14605

Eurorad

Radiological findings in Dyke-Davidoff-Mason Syndrome (DDMS)

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Section: Neuroradiology
Area of Interest: Neuroradiology brain Neuroradiology spine Head and neck
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: Digital radiography
Special Focus: Congenital Infection Ischaemia / Infarction Case Type: Clinical Cases
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Patient: 82 years, female

Clinical History:

An 82-year-old lady presented in an acute confused state. She had history of epilepsy, right hemiperesis and cognitive impairment.

On examination she was drowsy, disorientated with right upper and lower limb weakness, brisk reflex and hypertonia. The rest of the physical examination was normal.

Imaging Findings:

CT brain (Fig. 1a and1b) revealed severe left cerebral hemiatrophy with skull hypertrophy and enlargement of the left frontal sinus (Fig. 1C). CT AP of the skull (Fig. 2) demonstrated skull hypertrophy and hyper-pneumatisation of the left frontal sinus.

Discussion:

We report a case of an acutely confused 82 year old patient with known history of epilepsy, cognitive impairment and right hemiparesis. Imagine findings briefly show left cerebral atrophy, ipsilateral calvarian thickening and hyper-pneumatisation of the left frontal sinus. Our differential diagnosis includes: Dyke-Davidoff-Mason Syndrome (DDMS), Rasmussen Syndrome and Sturg-Webers Syndrome.

In 1933, DDMS was first described in a case series by Dyke, Davidoff and Mason. The condition was described as skull radiographic changes resulting from cerebral hemiatrophy [1]. Cerebral hemiatrophy in early life can result in compensatory ipsilateral changes characterised by calvarian thickening, hyper-pneumatisation of the sinuses and elevation of the orbit. Clinically, affected patients present in early childhood with contralateral hemiparesis, facial asymmetry, seizure and mental retardation.

DDMS can be divided into congenital or acquired. Congenital cerebral hemiatrophy can be distinguished from acquired cerebral hemiatrophy in early life by these radiological findings - ipsilateral midline shift and absence of the sulcal prominence that replaces the gliotic tissue [2].

Likely aetiologies for congenital DDMS can be due to vascular occlusion or circulatory anomalies in the middle

cerebral artery territory. Reports have suggested DDMS has a predilection for the left cerebral hemisphere [3]. Aetiologies for acquired DDMS are infection, ischaemia and trauma.

Rasmussen syndrome (also known as Rasmussen encephalitis) is a rare inflammatory disease seen in children. Seizure and cognitive impairments are known clinical presentations. Cerebral hemiatrophy is a well-recognised radiological finding. However, it differs from DDMS because there is usually no skull hypertrophy [4].

Sturge-Webers Syndrome (also known as encephalotrigeminal angiomatosis) can also present with seizure, mental retardation and a distinct port-wine stain on the forehead (ophthalmic division of the trigeminal nerve -V1). Radiologically, there is no midline shift [5], there can be cortical and subcortical calcifications with tram-teak sign.

In this case report, our patient did not have signs to suggest Sturge-Weber syndrome or Rasmussen syndrome. In addition, the clinical and radiological features were consistent with DDMS.
Differential Diagnosis List: Dyke-Davidoff-Mason Syndrome (DDMS), Rasmussen syndrome, Sturge-Weber syndrome

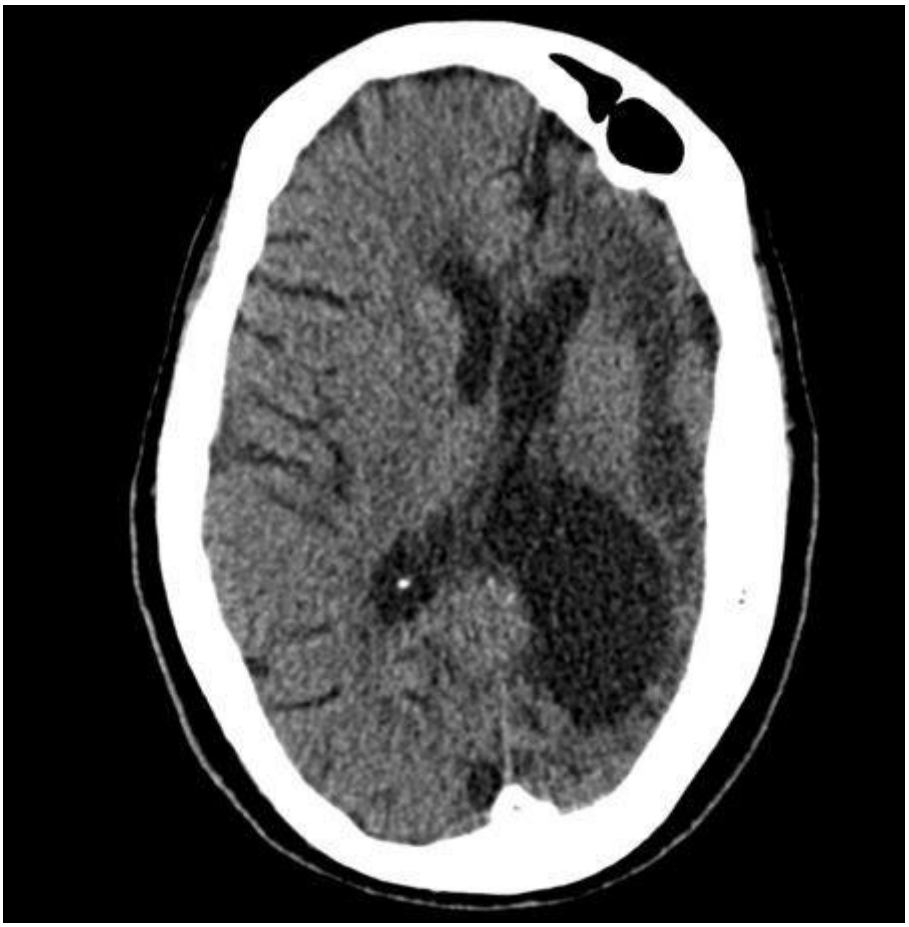
Final Diagnosis: Dyke-Davidoff-Mason Syndrome (DDMS)

References:

Dyke CG, Davidoff LM, Masson LB (1933) Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinus. Surg Gynecol Obstet 57:588–600
Sener RN, Jinkins JR. MR of craniocerebral hemiatrophy (1992) MR of craniocerebral hemiatrophy. Clin Imaging 16:93–97 (PMID: 1547482)
Kumar S, Lakshmaiah V, Naidu K. (2011) Case report: A rare cause for seizures and mental retardation – Dyke Davidoff Masson Syndrome. Int J Biol Med Res 2(4):1186–8
R D Tien, B C Ashdown, D V Lewis, Jr, M R Atkins and P C Burger (1992) Rasmussen's encephalitis: neuroimaging findings in four patients. AJR Am J Roentgenol 158 (6): 1329-32 (PMID: 1590136)
Pascual-castroviejo I, Viaño J, Moreno F et-al (1976) Hemangiomas of the head, neck, and chest with associated vascular and brain anomalies: a complex neurocutaneous syndrome. AJNR Am J Neuroradiol 17 (3): 461-71 (PMID: 8881240)

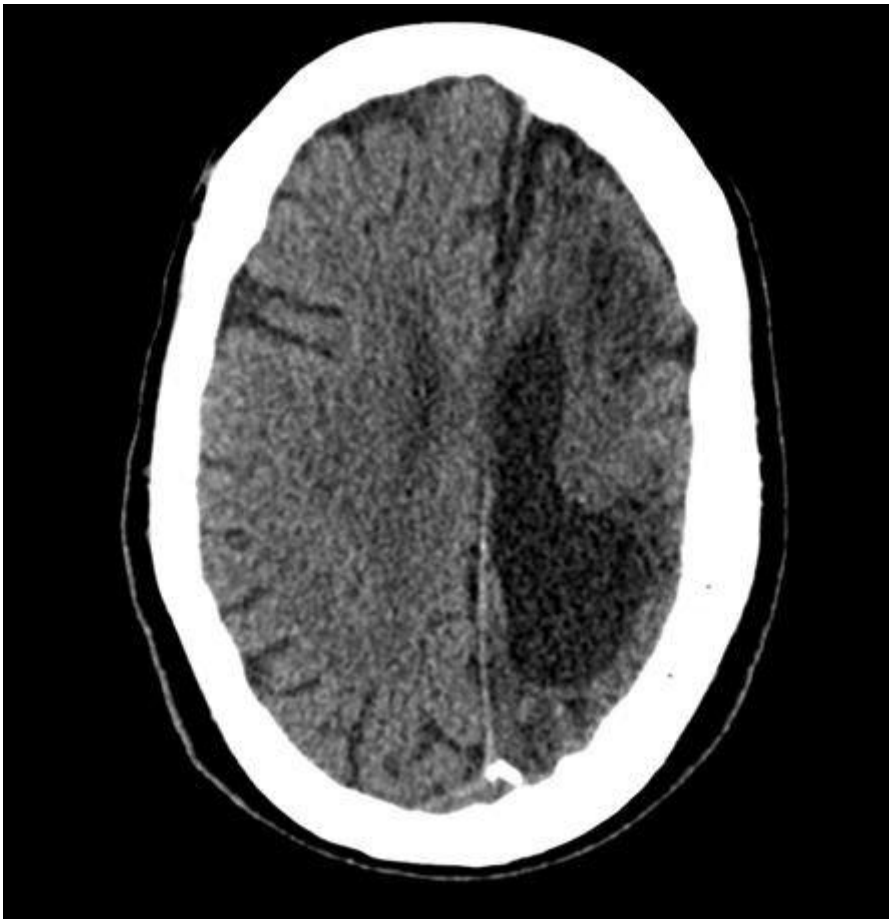
Figure 1

a



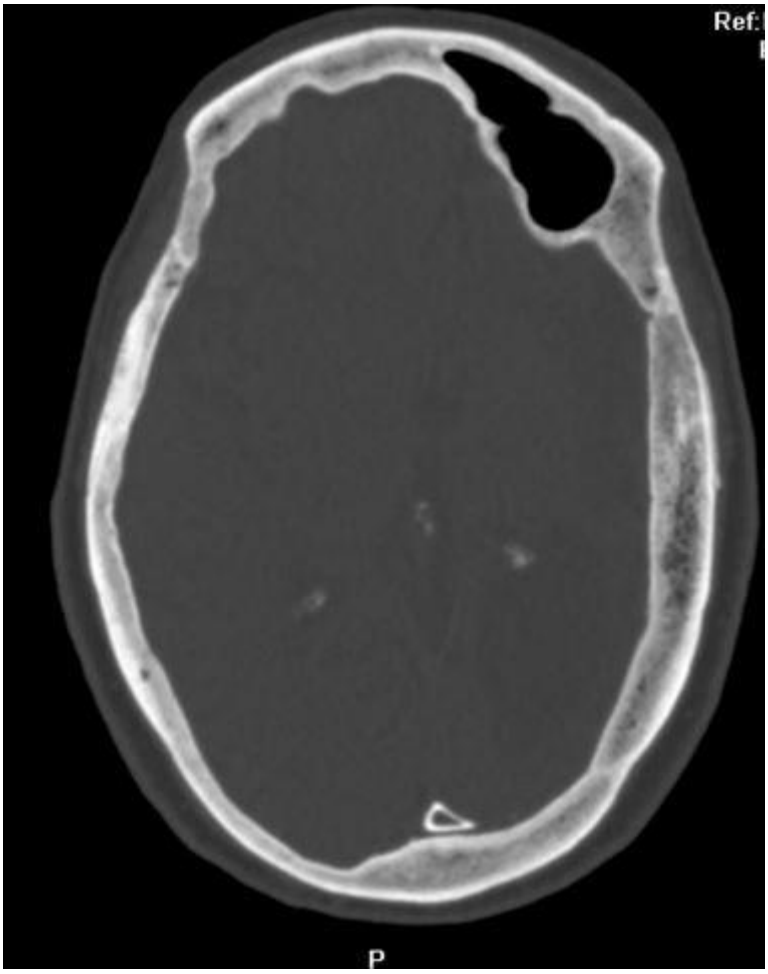
Description: CT brain, demonstrating severe left cerebral hemiatrophy with ipsilateral compensatory skull hypertrophy, enlarged frontal sinus consistent with DDMS. There are other findings of bilateral cerebellar atrophy. **Origin:** Department of Radiology, Midland Regional Hospital Mullingar, Co. Westmeath.

b



Description: CT brain, demonstrating severe left cerebral hemiatrophy with ipsilateral compensatory skull hypertrophy, enlarged frontal sinus consistent with DDMS. There are other findings of bilateral cerebellar atrophy. **Origin:** Department of Radiology, Midland Regional Hospital Mullingar, Co. Westmeath.

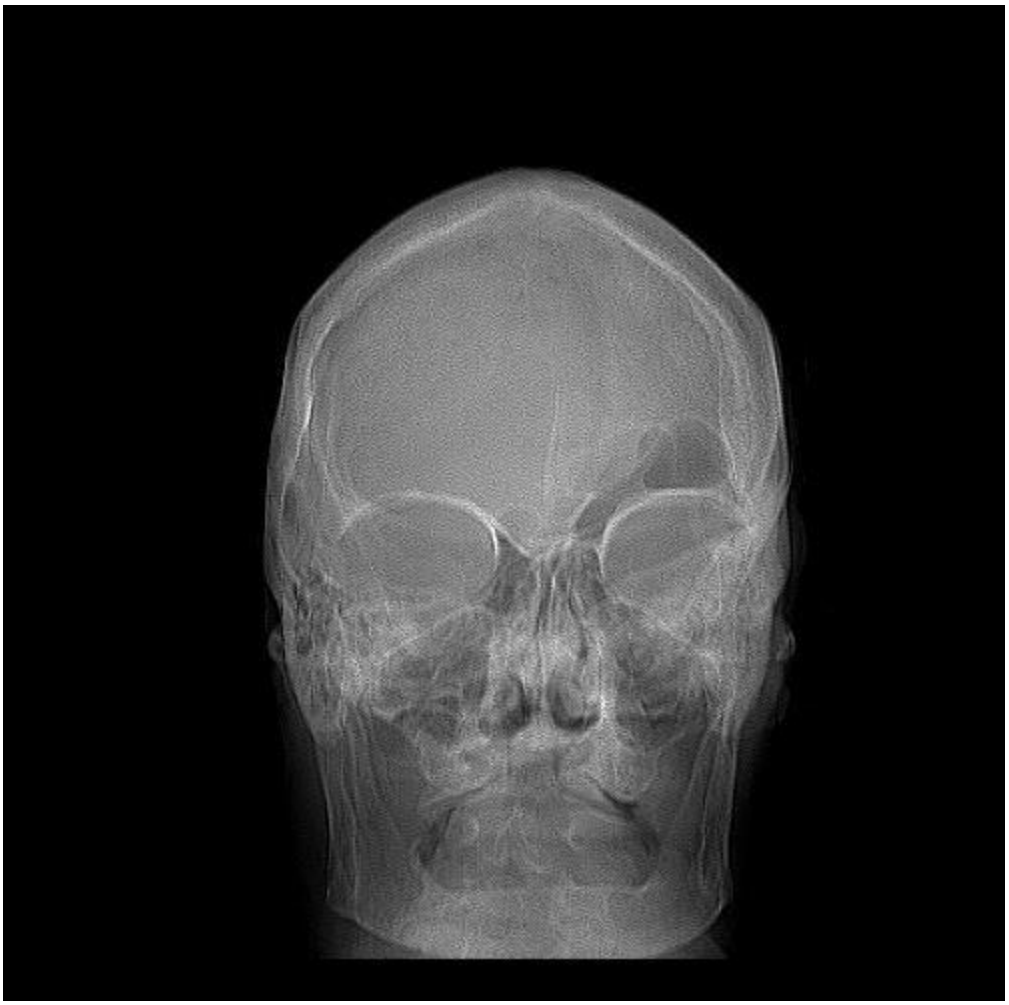
c



Description: CT brain bone window, the left frontal sinus hyper-pneumatisation is better illustrated.
Origin: Department of Radiology, Midland Regional Hospital Mullingar, Co. Westmeath.

Figure 2

a



Description: There is hyper-pneumatisation of the left frontal sinus, elevated left petrous ridge, left skull thickening and falx displaced to the left. **Origin:** Department of Radiology, Midland Regional Hospital Mullingar, Co. Westmeath.