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Dermoid Cyst in the Posterior Cerebral Fossa Associed to the Basilar Impression, Case Report

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Abstract :
BACKGROUND : *Basilar impression is a rare clinical condition characterized by upward protrusion of the odontoid process into the intracranial space, leading to bulbomedullary compression. decompression is indicated with or without posterior fixation.*
Klippel-Feil syndrome (KFS) is characterized by specific congenital anomalies of segmentation of the cervical spine.
On the other hand, dermoid cyst is a rare entity accounting for 0.04–0.7% ***(1-2-3)*** *of all intracranial tumours and the most common location is in the posterior fossa, at or near the midline.*
We present a rare case that associate a basilar impression, Klippel Feil syndrome and dermoid cyst in the posterior cerebral fossa .
CASE DESCRIPTION*: A 16 -year-old woman presented with HIC syndrome*
Findings on magnetic resonance imaging revealed hydrocephalus and cranial settling with the odontoid indenting the ventral medulla with posterior compression.
Computed tomography demonstrated the presence of basilar invagination with Kippel -Fiel syndrome.
The patiente underwent a surgery with the setting of DVP in the first place than decompression with posterior fixation and dermoid cyst excision in the second place.
CONCLUSION : *In this case, dermoid cyst of the posterior cerebral fossa is associated with dermal sinus and hydrocephalus, Klippel Feil syndrome and impression basilaire.*
The therapeutic management include a ventriculopéritoneal shun, decompression with posterior fixation and dermoid cyst excision.
Key words: *a case report, dermoïde cyst, posterior fossa, dermal sinus. Klippel–Feil syndrome, basilar impre*

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I. Introduction:

Dermoid tumour is a rare entity, accounting for 0.04–0.7% of all primary intracranial tumours **(1-2-3*)*** **.**Embryologically, it is derived from ectopic inclusion of epithelial cells during neural tube closure and it may be associated with various dysraphic disorders such as dermal sinus, myelomeningocele, and Klippel–Feil syndrome (KFS) as a consequence of a somite segmentation failure **(4-5-6-7*)***. The coexistence of dermoid tumour located within the posterior fossa and KFS is extremely **(4-6-8-9-10).**In this report, a review of cases with KFS in association with posterior cranial fossa dermoid tumour, illustrated by a recently admitted patient is reported, with special attention to the clinical and anatomical features of this rare combination of anomalies.

II. Case Report :

This 16-year-old woman presented with a sudden headache followed by nausea and vomiting. On examination at admission her Glasgow Scale was 15 points with cerebellar syndrome was found. The patiente had a short neck with a low hairline implantation and restricted neck motion. Fourth weeks before admission started with a mild headache and neck pain.
Magnetic resonance imaging (MRI) showed an hydrocephalus due to a tumor located in the midline of the posterior fossa and placed dorsally to the fourth ventricle***(Figure 1A-1B).***
Emergent computed tomographic (CT) scan showed an absence of fusion of the anterior arch of C1 and agenesis of the posterior arch of C1.(Figure 2).

Cervical MRI disclosed no abnormalities in the spinal cord or in the soft tissues ***(Figure 3).***
A DVP was inserted just after admission.
Surgical management of basilar impression was done by decompression with posterior fixation (Occipital-C2-C3) with excision of the dermaoid cyst ***(Figure 4)*** . A very rudimentary dermal sinus was discovered in the skin

reaching the duramater through a small midline hole in the occipital bone. A well encapsulated intradural lesion was found the tumor was totally removed. Pathological examination confirrmed the diagnosis of dermoid cyst and dermoid tract. The postoperative course of the patient was uneventful and 2 months after surgery she remains free of symptoms.

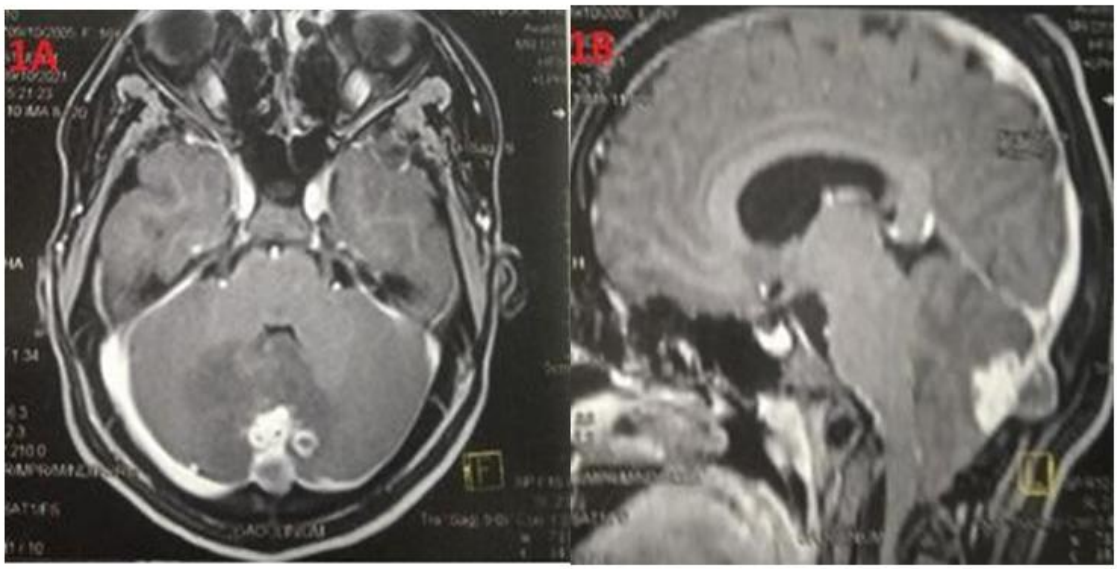

Fig1A : T1-weighted axial MR images ,*Figure 1B :* T1-weighted sagittal MR image showing an intra axial expansive process of the inferior vermis, arnold chiari malformation type 1 , medial cerebrospinal dermoid cyst and median occipital bone opening without fluid sac or nerve herniation.

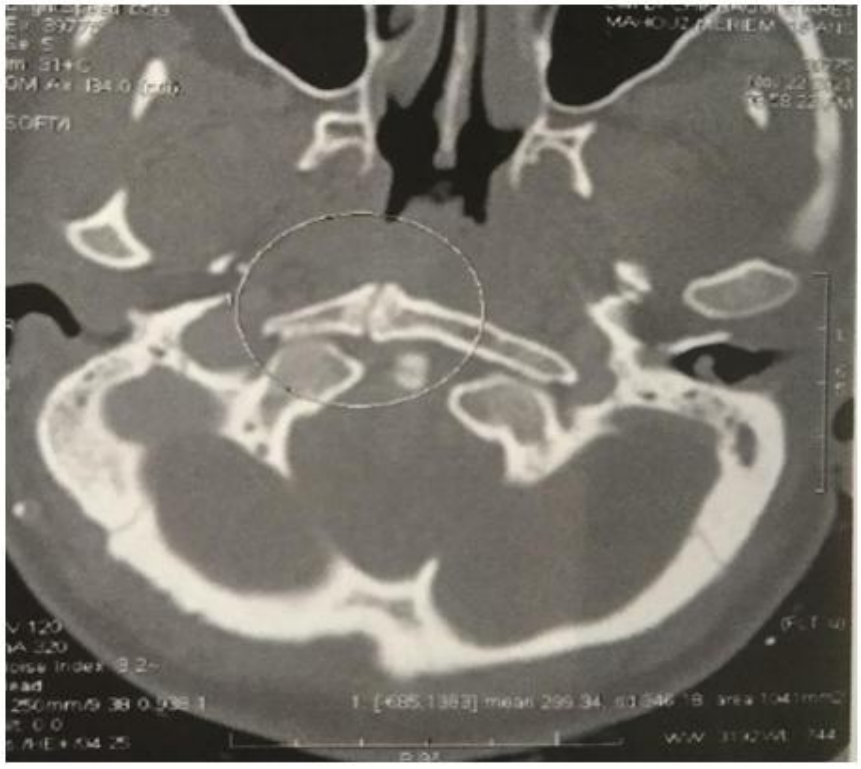

Fig 2 : computed tomographic (CT) scan showed an absence of fusion of the anterior arch of C1.

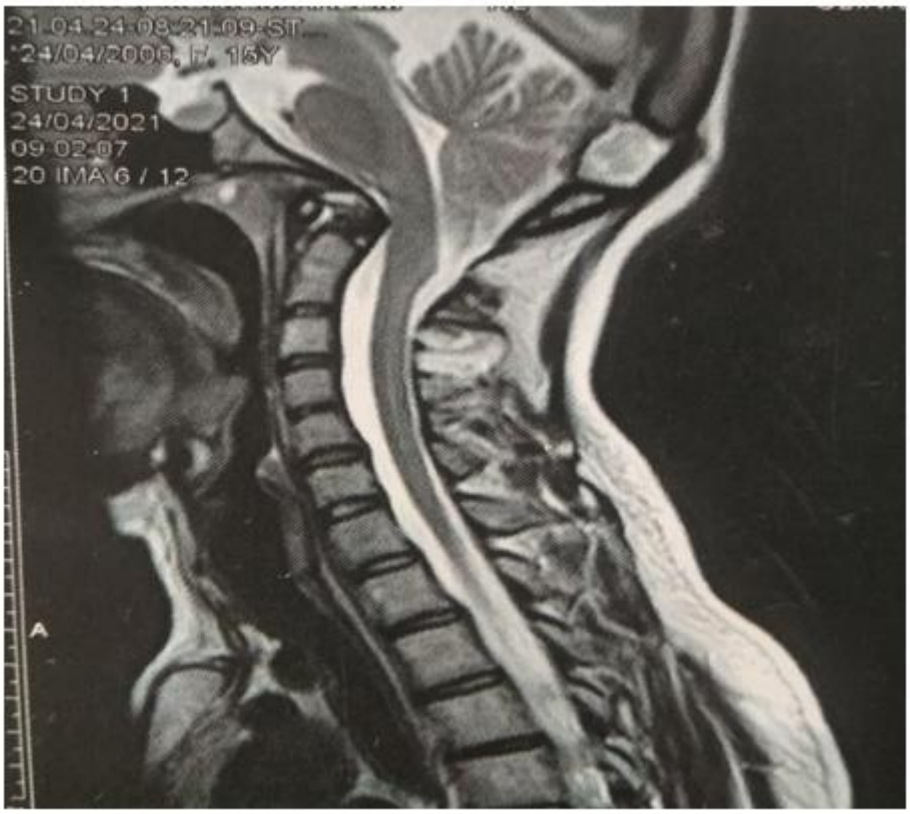

Fig3 : T2-weighted sagittal MR images showed a fusion of C2-C3, dermoid cyst, dermal sinus ans arnold chiari malformation type 1 witn no abnormalities in the spinal cord .

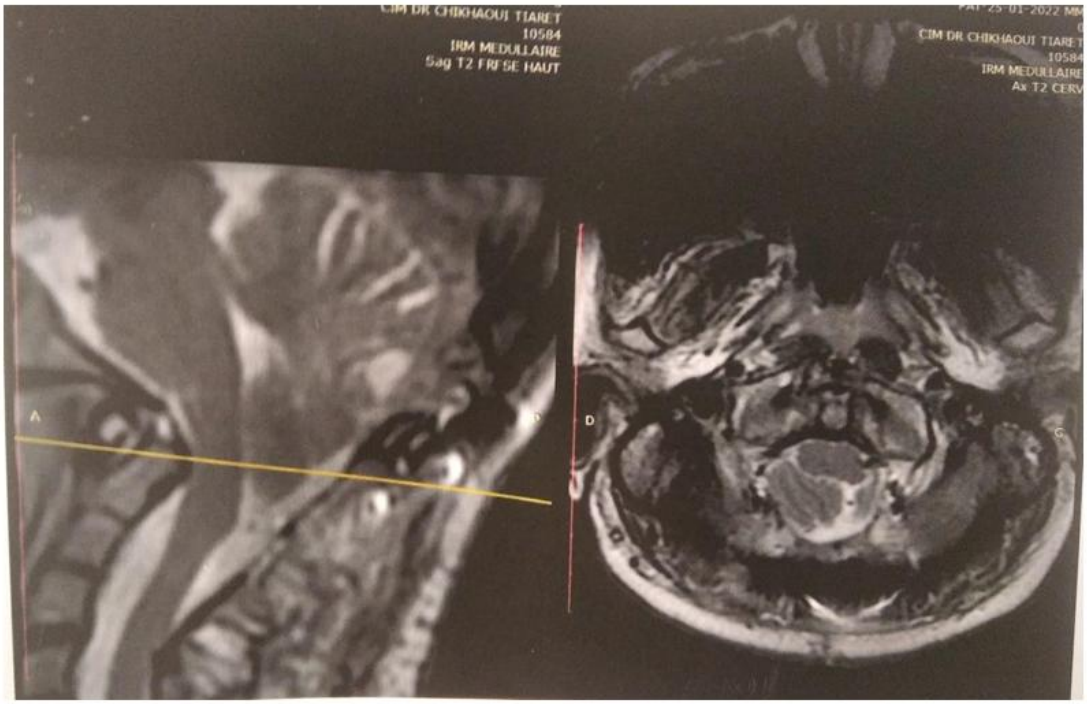

Fig 4 : post operative T2-weighted axial MR images *and* T2-weighted sagittal MR image of the same patient.

III. Discussion:

The intracranial dermoid cyst arises from ectoderm, probably derived from cell nest included during the closure of neural tube. The cyst wall is lined with stratified squamus epithelium and mixed appendages of ectodemal origin including sebaceous gland, sweat gland, and hair follicles (11-12). The cyst content is mixture

of decomposed epithelial cells containing keratin, glandular secretion, lipid metabolites, and cholesterol. Rarely dental enamels are also observed [6]. Dermoid cysts are found in supratentorial and infratentorial compartment and within the spinal canal (13). The commonest location of intracranial dermoid cyst is posterior fossa.

Intracranial dermoid cyst can be associated with complete or incomplete sinus tract, usually ending in an intracranial location of dermoid (12).

Kippel- Feil syndrome may be associated with craniovertebral junction anomaly, 52 % cases of Kippel-Fiel syndrome had classical triad of short neck, low hairline, and limitation of neck movements. However, the association with posterior fossa dermoid is rare. (14-15)

Many hypotheses are postulated for association of Kippel-Feil syndrome and cranio- verteberal junction anomaly with posterior fossa dermoid.

Gardner postulated Kippel-Feil deformity production is associated with neural and skeletal abnormalities, are produced due to distortion of somites by over distension of the neural tube. The failure of segmentation of cervical sclerotome can produce altered tissue tension at the cranio-vertebral junction region. These promote entrapment of dermal tissue, which give rise to posterior fossa dermoid, which may have associated dermal sinus (16) . Castillo et al. (18) postulated abnormal migration of neural crest might play a role in constellation of associated pathology.

A cranial computed tomography may show associated bone defect, intracranial dermoid or other associated defects. The posterior fossa dermoid cyst should be differentiated from another cystic lesion of posterior cranial fossa including cerebellar abscess. An early defect with few differentiated cells leads to development of dermoid(17).

These are hypodense due to cholesterol and keratin of epithelial cells, having well defined hypodense margin with a slight post contrast peripheral enhancement produced bycapsule nad inflamed pericapsular area.

IV. Conclusion:

Here, we report a case of a patient with complex craniocervical malformation accompanied by posterior cranial fossa dermoid cyst.

Full evaluation is needed before surgery as well as careful management .

An early diagnosis and an appropriate surgery to prevent complications such as neural compression should be the goal.

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