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Case Report

Association of Craniovertebral Junction Anomalies, Klippel-Feil Syndrome, Ruptured Dermoid Cyst and Mirror Movement in One Patient: A Unique Case and Literature Review

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ABSTRACT

The Klippel-Feil syndrome (KFS) has been reported to be associated with intracranial neoplasms, most frequently epidermoid or dermoid cysts. To our knowledge, however, patients who present with a posterior fossa dermoid cyst (DC) and KFS are extremely rare with only 24 previously reported cases in the English literature worldwide. Therefore, we present the first report of a patient with a craniocervical ruptured DC accompanied by craniovertebral junction (CVJ) anomalies, KFS and mirror movement. Meanwhile, a literature review of KFS accompanying with posterior fossa DC discusses these conditions from the embryological, anatomical, clinical and therapeutic perspectives. Additionally, the combination of CVJ anomalies, KFS and DC may represent a new syndrome that has previously gone unnoticed.

KEYWORDS: Craniovertebral junction, Dermoid cyst, Klippel-Feil syndrome, Mirror movement, Rupture

INTRODUCTION

Dermoid cysts (DCs) are rare neoplasms, accounting for 0.1–0.7% of all intracranial tumors (20, 29). In 1936, Love and Kernohan first reported the association of DCs of the posterior cranial fossa with cervical fusion anomalies, particularly Klippel-Feil syndrome (KFS) (20). Subsequently, only 23 other cases of KFS-associated posterior cranial fossa DC (1, 3, 4, 7-10, 13, 15, 19, 20, 23, 25, 27, 28, 34, 35, 40, 41, 44) have been reported in the English literature, as listed in Table I. Because these diseases are very rare, the association between DC and KFS may be pathophysiologically significant. We present the first report of a patient with a craniocervical ruptured DC accompanied by craniovertebral junction (CVJ) anomalies, KFS and mirror movement.

CASE REPORT

History. A 28-year-old unmarried woman presented with a five-month history of unsteady gait, which had worsened one month before admission. The onset of this symptom occurred without any obvious cause. She did not complain of headache, nausea or vomiting episodes. In addition, since birth, the patient had experienced involuntary movement of the contralateral hand while performing voluntary movements with the other hand, although this symptom had never previously caused distress. There was no family history of medical disorders.

Physical Examination. The patient, whose neck was short with a high neck/height ratio (1:20.5, normal 1:9) (32), had a low posterior hairline, extending to the level of the C7 spinous process (Figure 7A). Upon physical examination, the examiner was able to palpate one apparent pit in the

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nape of the patient's neck (Figure 7A), and flexion-extension motions were maintained to a limited extent (Figures 1B, C). Mirror movement was easily elicited and primarily confined to the hand. The patient had gait ataxia, as well as apparent clumsiness in performing the heel-knee-tibia test and finger-to-nose test; Romberg's sign was positive. The other neurological examinations were negative.

Radiographic Examination. Plain radiography and computed tomography (CT) of the head and neck revealed congenital fusions (classified as Type I of KFS) of C2-C7 and T1-T3 associated with platybasia or basilar invagination (Figures 1A-C, 2A, B) (11). A three-dimensionally (3D) reconstructed CT of CVJ demonstrated spina bifida occulta from C2 to C7 (Figure 3A-C). Most notably, CT showed a low-density, calcified,

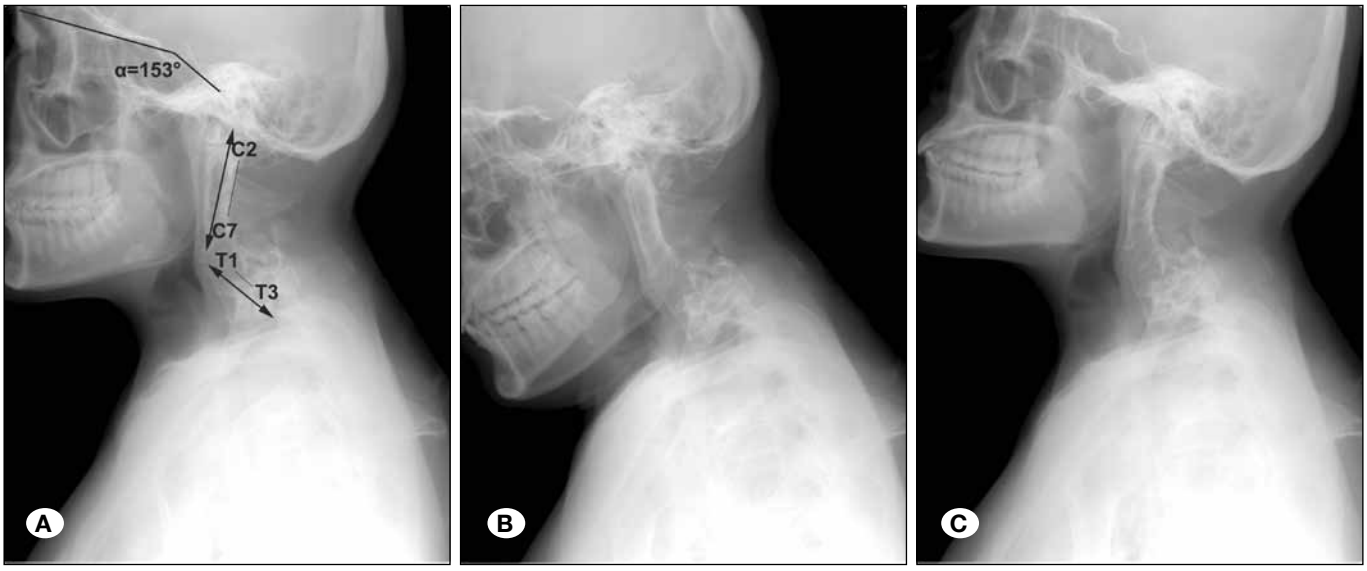

Figure 1: Plain cervical spine radiographs. **A)** Plain radiograph shows congenital fusions (Type I of KFS) (*double arrowheads*) with 153° NTB angle of Welcker joining the nasion (N), tuberculum (T), and basion (B). The normal angle should be less than 130° (29), platybasia with an increased NTB angle. **B, C)** Flexion and extension of cervical spine.

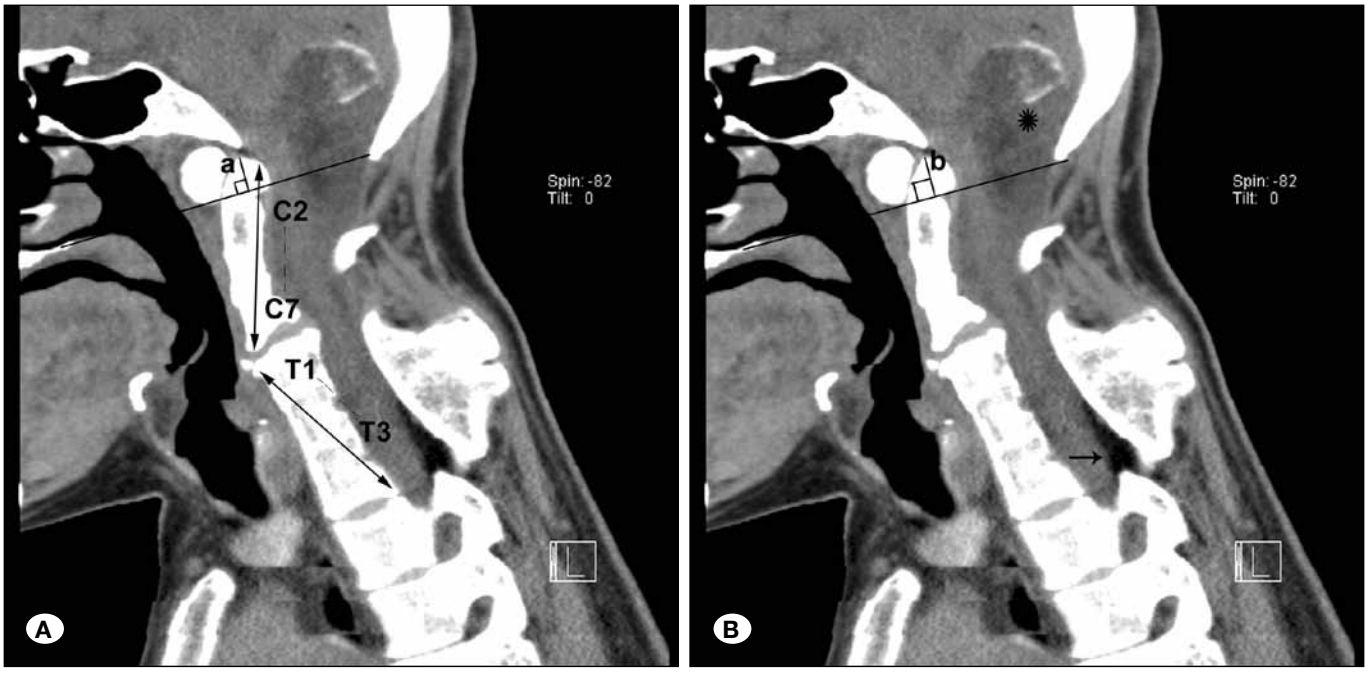

Figure 2: Midsagittal reconstructed images from cervical spine CT. **A)** *Double arrowheads* reveal congenital fusions (Type I of KFS) (11). Radiographic criteria for basilar invagination with dens protrudes >2.5mm (*a*=8.0mm) above Chamberlain's line between hard palate and opisthion (20). **B)** Another criteria with dens protrudes >5.0mm (*b*=9.8mm) above McGregor's line between hard palate and the lowest point of occiput (20). Besides, there are two lesions (*asterisk* and *black arrow*).

circumscribed mass lesion (measured as -70~170 Hu) around the foramen magnum. In addition, another lesion (measured as -120 Hu) was found in the upper thoracic spinal canal. CT indicated that the second lesion was likely to be a lipoma (Figure 2A, B).

Magnetic resonance imaging (MRI) revealed one oval intradural mass located in the craniocervical area, measuring 28.6 mm anteroposteriorly and 50.3 mm rostrocaudally. The lesion was heterogeneous and had areas of hyper-, iso, and hypo-intensity on T1-weighted images and areas of hyperintensity on T2-weighted images (Figure 4A-C). In addition, multiple small, short T1WI and long T2WI signal lesions were scattered in the brain fissures, subarachnoid spaces and ventricles (Figure 4A, Figure 5A-D). These lesions were assumed to be fatty droplets derived from spontaneous rupture of the DC (Figure 5A).

3D CTA of the vertebral artery (VA) (Figure 6A-E) demonstrated that the third segment of the right vertebral artery (RVA) had an abnormal course, leaving the transverse foramen of the axis and passing into the foramen magnum below the occipitalized lateral mass of C1; this was classified as a Type I VA at the CVJ with occipitalization of the atlas (42). The second segment of the left vertebral artery (LVA) was obviously distorted from the osseous fusion of C2-C7. In addition, there was a fenestration of the vertebrobasilar artery.

Surgery. With the use of microsurgical instruments, total resection of the lesion was performed (Figure 7A-C). The anterior portion of the lesion was adherent to the dorsal surface of the brainstem, where an inconspicuous cleft was observed, as shown in the preoperative MRI (Figure 5A). Next, hydrocortisone was used to extensively irrigate the resection bed, in order to prevent postoperative chemical meningitis. No occipitocervical (OC) fixation or fusion was performed during the surgery. After the successful operation, a postoperative neck collar was prescribed for one month.

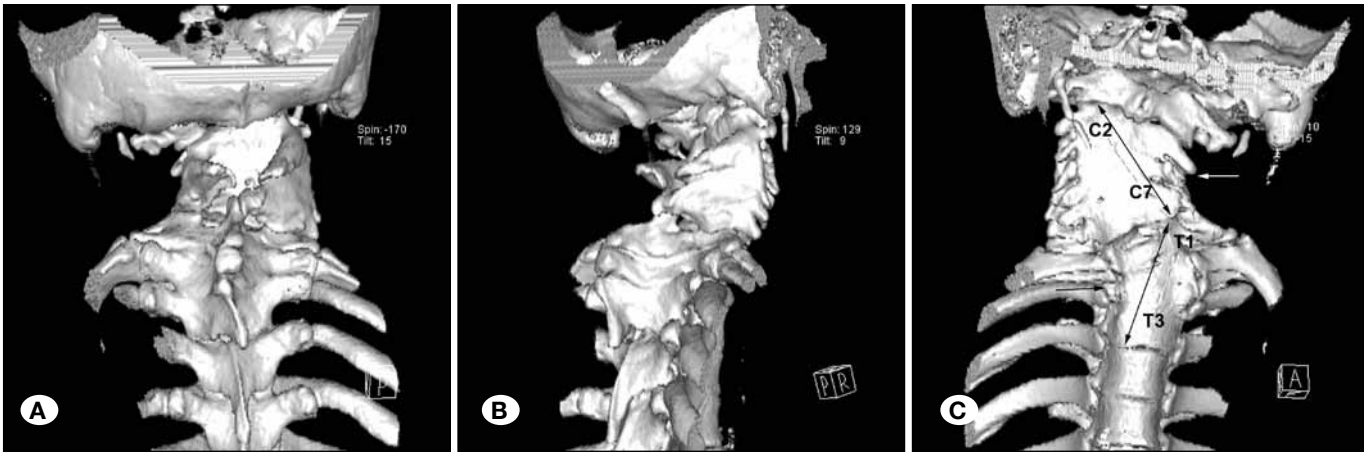

Figure 3: 3D CT reconstruction of craniovertebral region. **A)** Posterior view rendering of spina bifida occulta of C1-C7. **B)** Lateral view showing of defects of posterior arch of atlas. **C.** Anterior view indicating of scoliosis of congenital fusion (*double arrowheads*), bifid anterior arch of atlas, and atlantooccipital joint asymmetry, cervical and upper thoracic spine concave to right (*white arrow*) and left (*black arrow*) respectively.

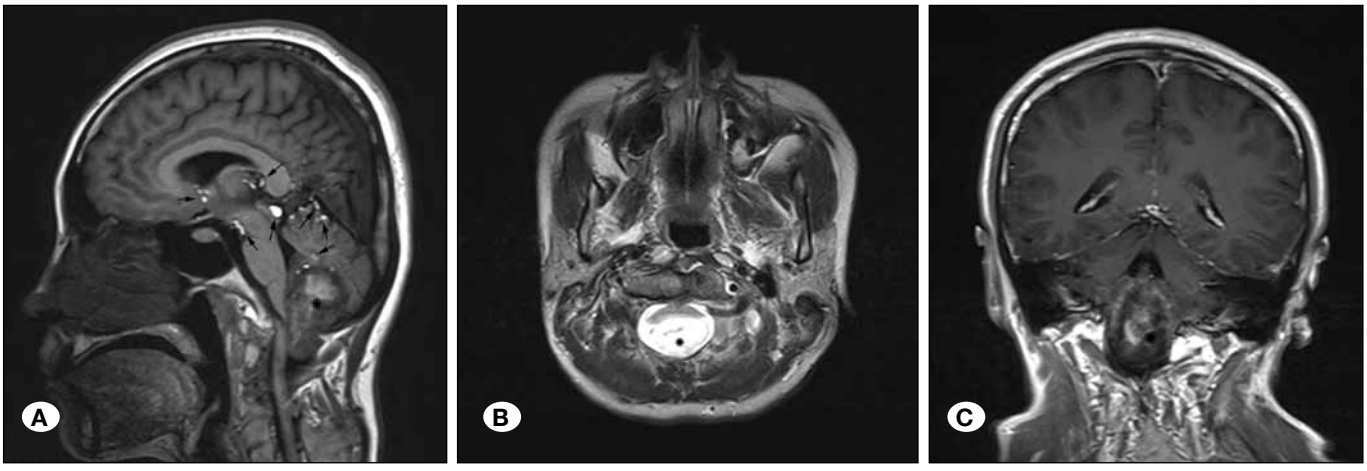

Figure 4: Preoperative MRI of head and neck. **A)** Sagittal T1WI reveals one heterogeneous mass (*asterisk*) and scattered droplets (*arrows*). **B)** Axial T2WI shows a hyper-intense mass (*asterisk*). **C)** Coronal contrast-enhanced T1WI of the lesion (*asterisk*).

Pathological Findings. Microscopic examination of the excised mass revealed the presence of a classic DC with calcification (Figure 8).

Postoperative Course. The course was uneventful, and the patient was discharged with no further neurological deficits on the 10th day after the surgery.

Follow-up. After six months of follow-up, the patient's preoperative symptoms had resolved, with the exception of mirror movement. Compared with the preoperative MRI findings (Figures 4A-C, 5A-D), postoperative MRI of the head showed no recurrence of the lesion; the fat droplets were stable without enlargement, reduction or apparent displacement (Figure 9).

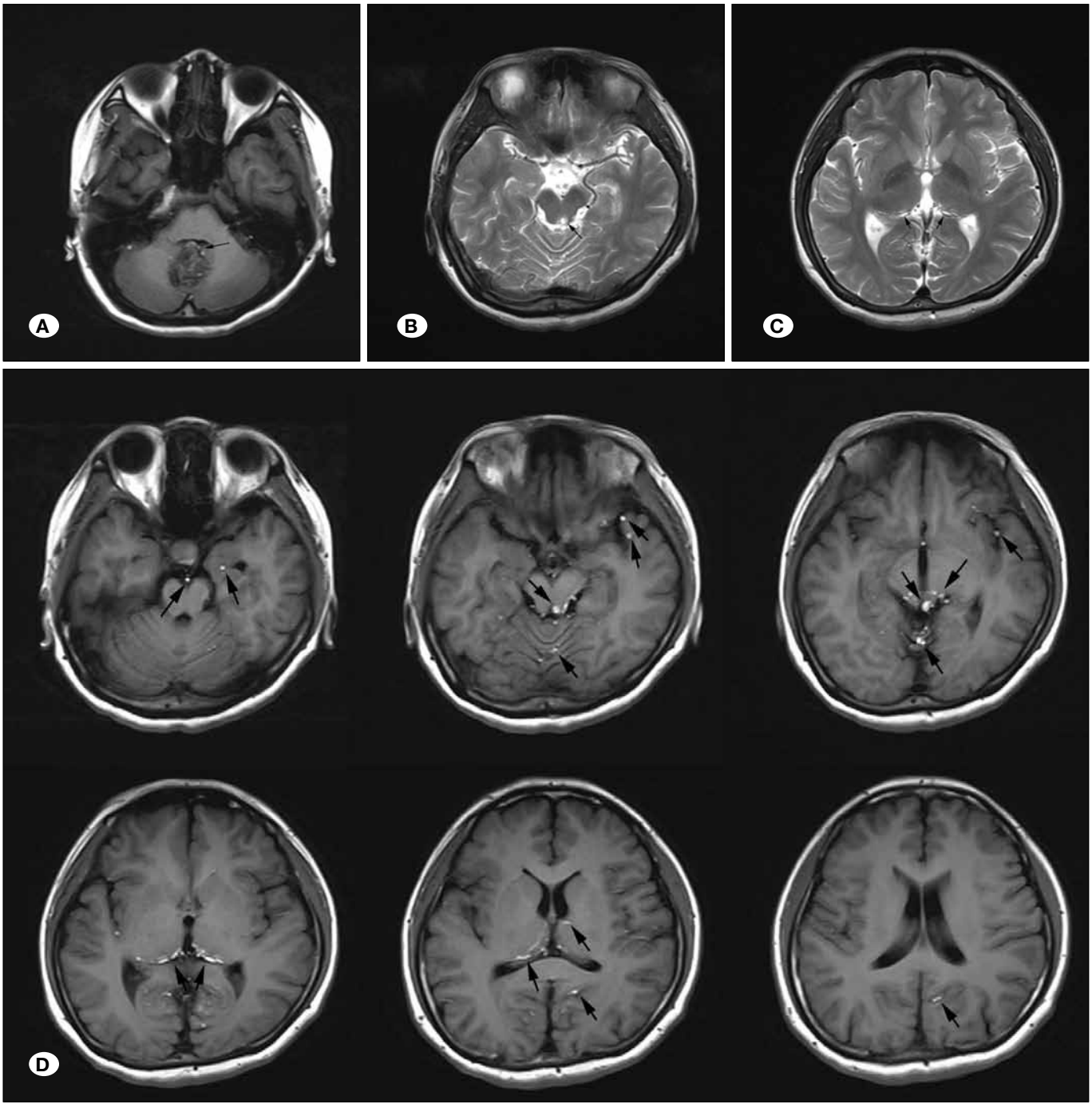

Figure 5: A series of MR images showing the scattered fat droplets. **A)** There is an inconspicuous cleft (*arrow*). **B**, **C)** They show the hyperintense fat droplets (*arrows*) on T2WI. **D)** It describes the hyperintense fat droplets (*arrows*) on T1WI.

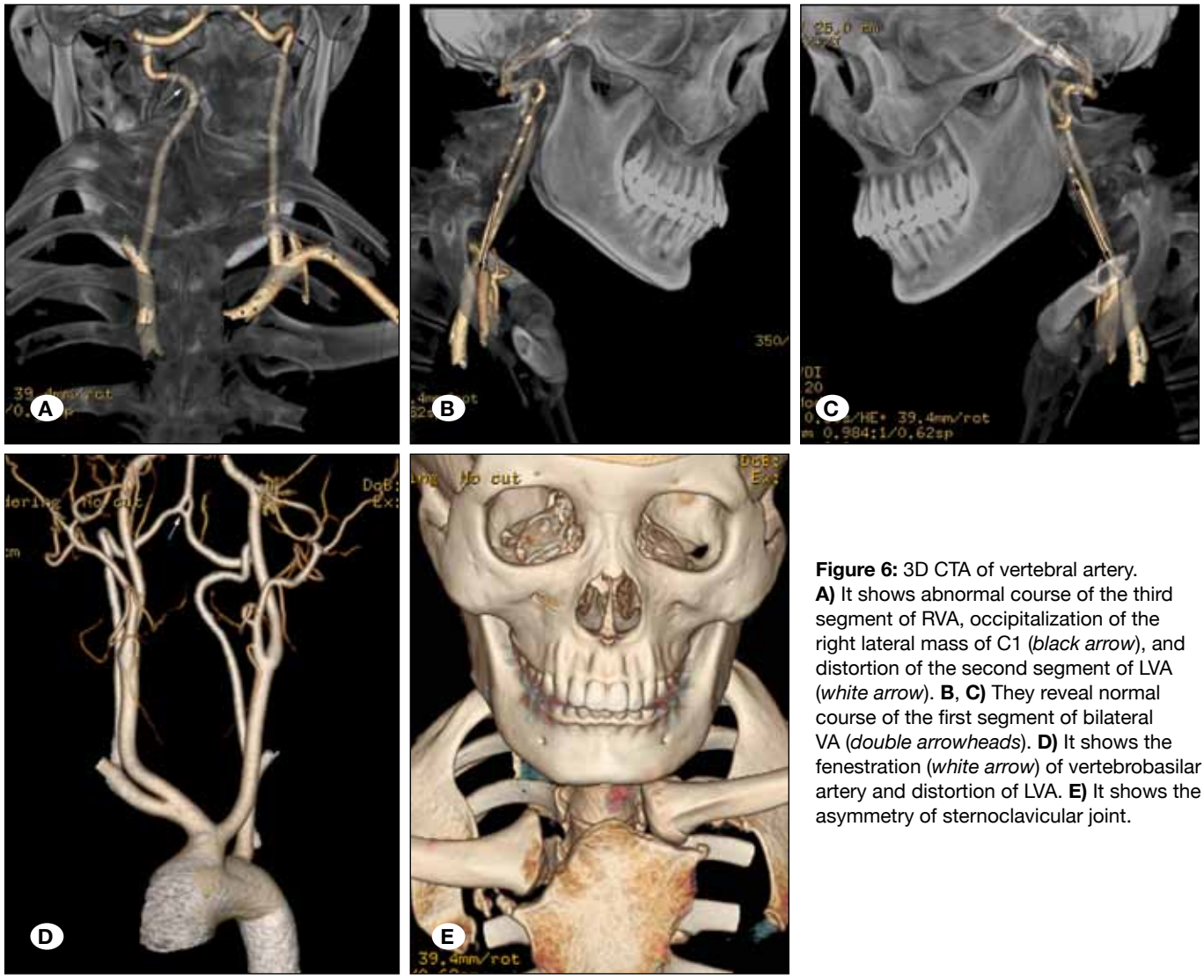

Figure 6: 3D CTA of vertebral artery. **A)** It shows abnormal course of the third segment of RVA, occipitalization of the right lateral mass of C1 (*black arrow*), and distortion of the second segment of LVA (*white arrow*). **B**, **C)** They reveal normal course of the first segment of bilateral VA (*double arrowheads*). **D)** It shows the fenestration (*white arrow*) of vertebrobasilar artery and distortion of LVA. **E)** It shows the asymmetry of sternoclavicular joint.

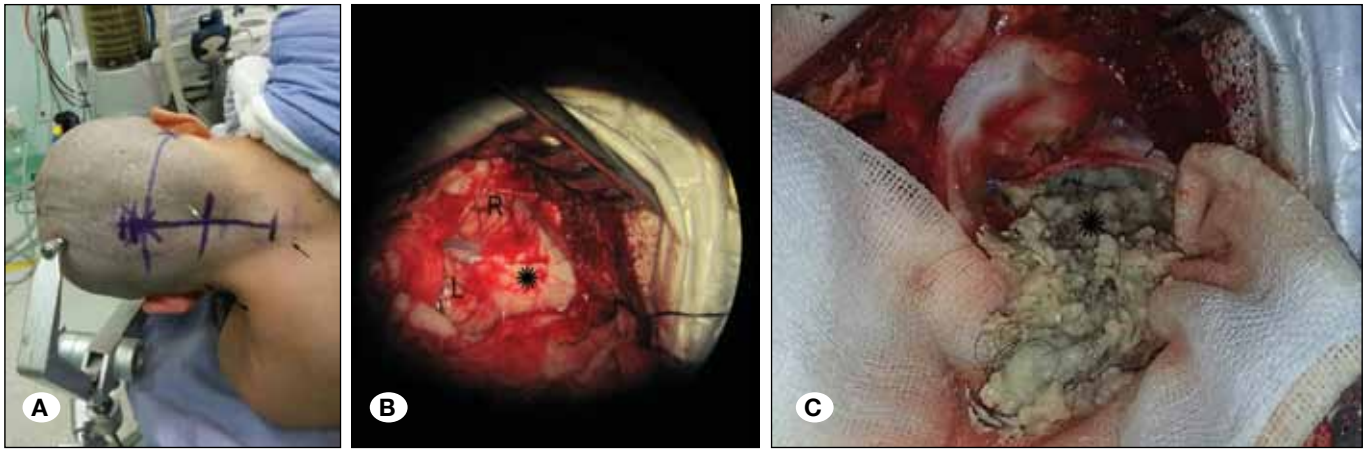

Figure 7: Intraoperative images of the patient. **A)** It shows the low occipital headline (*black arrow*), short neck (*double arrowheads*) and one apparent pit (*white arrow*). **B)** It reveals bulgy cerebellar tissue (*R, L*) and tumor (*asterisk*). **C)** the contents in the cyst (*asterisk*). (*R, L* shorting for right, left cerebellar hemisphere).

DISCUSSION

Analysis of the 25 cases in Table II present more details concerning the association of KFS with DCs. These cases included 14 women and 11 men, with an average age of 19.7 years (ranging from 1 to 61 years). Pediatric cases represented 56% of patients, indicating that approximately half of such patients are not diagnosed until adulthood. The follow-up period ranged from 3 months to 6 years among the 17 cases with sufficient information. Tumor recurrence was not reported in these patients during the follow-up period.

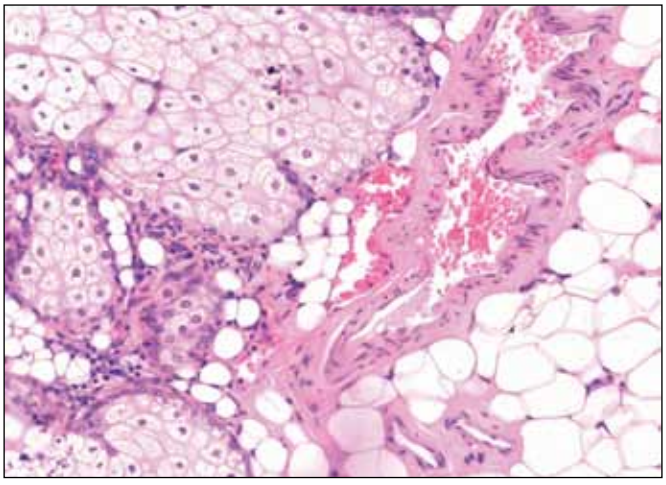

Figure 8: Photomicrograph demonstrating a classic DC with a lining of smooth muscle cells, sebaceous gland and keratinous material with calcification (H& E, × 200).

Klippel-Feil Syndrome

KFS is associated with the classic triad of a short neck, a low posterior hairline and a limited range of motion of the neck; it is caused by congenital fusion of two or more cervical vertebrae (11, 26). Analysis of the 25 cases shows that the classic KFS triad is present in 37.5% of patients. The lack of population screening studies has made it impossible to determine the exact incidence and prevalence of KFS (39). KFS can be classified into three subtypes (11)—type I, II or III:

Type I is defined as having multiple cervical or upper thoracic vertebral fusions;

Type II is defined as having isolated fusions at 1 or 2 cervical interspaces;

Type III is defined as having fusions in the cervical spine combined with lower thoracic or lumbar fusion.

Therefore, the current case was classified as Type I of KFS. As shown in Tables I and II, vertebral fusions were described in 21

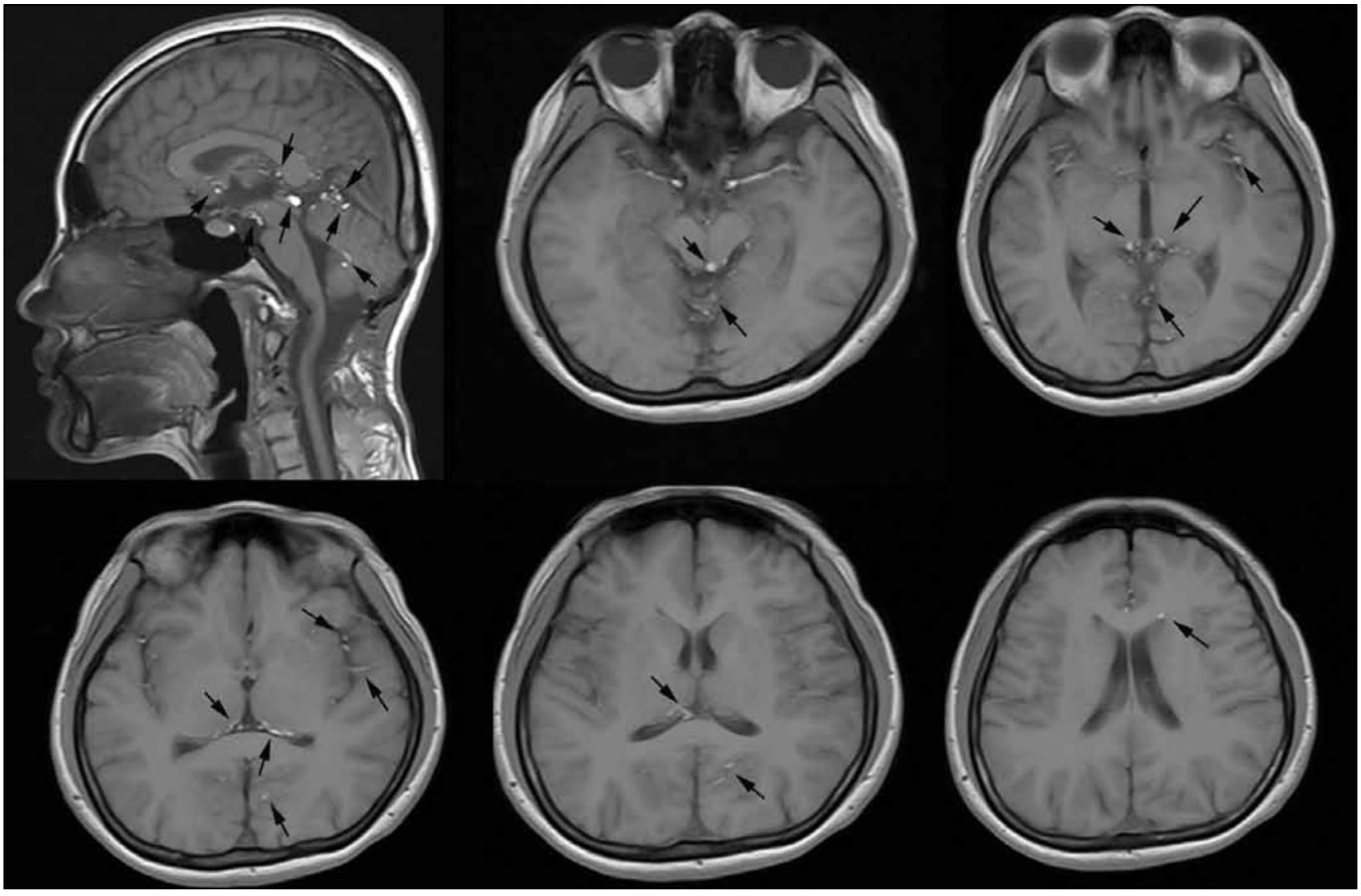

Figure 9: T1 MRI follow-up scans show the fat droplets (arrows) in the brain.

cases, with 62% classified as type I, 33.3% classified as type II, and 4.7% classified as type III. Furthermore, it is unclear whether the diagnosis of KFS is a discrete entity or if it is one condition in a spectrum of congenital spinal deformities (12). As described in Table I, congenital deformities associated with KFS and DCs include Sprengel's deformity, asymmetry of the sternoclavicular joint, fusion of the ribs or rudimentary cervical ribs, pigeon chest, palatal fissure, and basilar artery fenestration et al.

In recent years, numerous studies of KFS have been reported, particularly regarding the genetic etiology, such as a paracentric inversion on 8q (6), mutations of MEOX1 gene (2, 24), Notch signaling pathway (33, 36), Pax 1 and Pax 9 (5), or HOX gene (21). However, most of these conditions have only been confirmed in animal studies, and further studies are warranted to investigate the exact genetic origins of KFS.

Dermoid Cysts

The common view is that there is a focal failure of cleavage between the neuroectoderm and cutaneous ectoderm between the third and fifth weeks of embryogenesis (15, 41). This results in the incorporation of cutaneous ectoderm, which forms the skin epithelium and all skin appendages, into the neural tube. Additionally, dermoid or epidermoid cysts of the CNS may be produced (4, 10, 34, 37). The clinical manifestations of DCs are non-specific and insidious, unless rupture occurs (14). Symptoms in patients with unruptured DCs result from compression of adjacent neurovascular structures. As shown in tables 1 and 2, DC patients were often admitted with signs (41.7%) of elevated intracranial pressure (rICP), in addition to cerebellar signs (33.3%). Rarely, ruptured intracranial DCs could result in dissemination of fatty material into cisterns, producing variable neurological symptoms, such as chemical meningitis (43), seizures (18), visual deficits (38), hydrocephalus (14), transient cerebral ischemia (45), or no symptoms, just as in our case. Additionally, rupture is usually spontaneous and is due to an increase in the size of the tumor's contents, together with head movements and/or brain pulsations; however, rupture has been reported following head trauma (16, 30). According to Table II, DCs located in the midline of the posterior fossa were found in 21 cases (84%), DCs extending to the craniocervical junction (including the present case) were found in 3 cases (12%), and a DC in the cerebellar hemisphere was found in one case (4%), showing that the midline of the posterior fossa is the most common site of DCs. Complete resection of DCs was achieved in 86% of cases. Additionally, 87.5% (21/24) of patients did not require OC fusion and did not demonstrate instability; instead, there was one patient with mild restriction of neck movement after OC fusion.

Craniovertebral Junction Anomalies

The bony CVJ can be divided into two components: a central pillar, which consists of the clivus and a central pivot (including the dens and C2 vertebral body), and a bistratal ring around the central pivot. They are the foramen magnum ring superiorly and the atlantal ring inferiorly (29). Therefore, developmental anomalies at the CVJ can also be classified into those related to the two components. As shown in Tables I and II, atlas assimilation is the most common CVJ anomaly, followed by platybasia, basilar invagination, atlantoaxial dislocation (AAD) and spina bifida occulta. A defect in the posterior arch of C1 also occurred in the present patient.

Mirror Movement

Mirror movement, also known as synkinesia or Kallmann's syndrome (22), occurs when voluntary movements of one side of the body are copied involuntarily by the other side of the body. This synkinesia manifests as a rare disorder that is usually associated with KFS (31, 35). It may represent neurological impairment or agenesis of the pyramidal decussation (44), cerebral cortex, corpus callosum, medulla oblongata or spinal cord (31).

To date, however, according to Table II, only three cases of KFS and posterior fossa DC have been reported with mirror movement. In our case, we believe that neurological impairment in the pyramidal decussation and the medulla oblongata, due to long-term compression by a congenital tumor, may be the cause of mirror movement.

■ CONCLUSION

This is the first reported case of associated CVJ anomalies, KFS, ruptured DC and mirror movement in one patient. A literature review discusses these conditions from multiple aspects. Several hypotheses (4, 15) have been proposed to explain the association among posterior fossa DC, KFS, and CVJ anomalies. Additionally, the combination of CVJ anomalies, KFS and DC may represent a new syndrome that has previously gone unnoticed.

Table I: Literature Review of 25 Cases of Klippel-Feil Syndrome with Dermoid Cyst of the Posterior Fossa (Including the Present Case)

Authors & year	Age (yrs) Sex	Clinical manifestation	Dermal sinus/ Infection	Radiological modality	Location	Rupture	Fusion	CVJ Malformations	Scoliosis	Other Malformations	Treat-ment	Complica-tions	Follow-up
Love et al., 1936 (20)	30, F	rICP rigidity of neck	No/No	Plain x-ray ventriculography	Midline of posterior fossa	No	C2-C3	--	No	No	Total res	No	No rec of 7 mons
Logue et al., 1952 (19)	61, F	rICP 2mm dimple above occipital protuberance	Yes/No	Plain x-ray	Midline of posterior fossa	No	Bodies and laminae of C2-C5	--	--	--	Total res	--	--
Tytus et al., 1956 (41)	5, F	rICP	Yes/No	Plain x-ray	Midline of posterior fossa	No	Multiple fusions through the whole spine	--	--	--	Total res	No	No rec of 4 yrs
Roberts et al., 1957 (35)	2yrs 10mons, M	rICP meningitis a swelling below the occipital protuberance	Yes/Yes	Plain x-ray	Midline of posterior fossa	No	Multiple fusions through whole cervical vertebrae	--	--	--	Total res	--	--
Whittle et al., 1983 (44)	2, F	head tilt to right mirror movement gait ataxia triad of KFS	No/No	Plain x-ray CT scan Metrizamide myelography	Midline of posterior fossa	No	Bodies of C2-C5	Spina bifida occulta at level of C6-C7	--	--	Total res	No	No rec of 1 yrs
Diekmann-Guiroy et al., 1989 (8)	42, F	aseptic meningitis triad of KFS	No/Yes	Plain x-ray MRI	CVJ	--	C2-C5	Spina bifida	--	--	Total res	No	--
Dickey et al., 1991 (7)	14, F	rICP vertical nystagmus dysdiadochokinesia mirror movement	Yes/No	Plain x-ray CT scan	Midline of posterior fossa	No	Bodies and laminae of C4-C6 bodies of C1-C2 laminae of C2-C3	Occipitalization of posterior arch of C1	Scoliosis of chest, concave to the right	Sprengel's deformity anterior fusion of first and second ribs hemi-vertebra at T3	Partial Res	No	--
Kennedy et al., 1998 (15)	40, F	headache gait ataxia absence of the right corneal reflex and gap reflex	--/--	Plain x-ray MRI	Midline of posterior fossa	No	--	--	--	--	--	--	--
Muzumdar et al., 2001 (25)	12, F	torticollis short and webbed neck high arched palate	No/No	Plain x-ray MRI	Midline of posterior fossa	No	bodies of C2-C5 laminae of C2-C7	AAD Occipitalization of the atlas	--	bifid uvula pigeon-Chest	Total res OC fusion		No rec of 1 yrs
	8, M	headache short neck torticollis truncal ataxia	Yes/No	MRI	Midline of posterior fossa	No	--	--	Scoliosis of cervical spine	--	Partial res	No	No rec of 9 mons

Table I: Cont.

Authors & year	Age (yrs) Sex	Clinical manifestation	Dermal sinus/ Infection	Radiological modality	Location	Rupture	Fusion	CVJ Malformations	Scoliosis	Other Malformations	Treat-ment	Complica-tions	Follow-up
Aksoy et al., 2001 (1)	16, M	suboccipital lump triad of KFS	Yes/No	Plain x-ray CT scan MRI	Midline of posterior fossa	No	C3-C7	Occipitalization of posterior arch of C1	--	Rudimentary cervical ribs on the left	--	--	--
Hinojosa et al., 2001 (13)	23, F	rICP 2mm dimple above occipital protuberance triad of KFS	Yes/No	Plain x-ray MRI	Midline of posterior fossa	No	Bodies and laminae of C4-C5	--	--	--	Total res	--	No rec of 1 yrs
Sharma et al., 2001 (37)	4, M	rICP head tilt to left pea-siaed occipital lump restricted neck mobility and stiffness truncal ataxia	--/--	Plain x-ray CT scan MRI	Midline of posterior fossa	No	C2-C3	Occipitalization of posterior arch of C1	Scoliosis of cervical spine, concave to the left	hemi-vertebra at the right side of C5-7	Partial Res	No	--
González-Darder et al., 2002 (10)	36, M	rICP juvenile diabetes ataxia triad of KFS	Yes/No	Plain x-ray CT scan MRI	Midline of posterior fossa	No	C3-C6	absence of posterior arch of C1	--	Cervical and dorsal ribs	Total res	No	No rec of 1.5 yrs
Oertel et al., 2002 (27)	54, F	--	No/--	MRI	Midline of posterior fossa and cer hemisphere	No	C3-C6	No	--	--	Micro-surgical res	No	No rec of 3 mons
Caldarelli et al., 2004 (3)	5, M	Recurrent hyperpyrexia	Yes/Yes	MRA	Midline of posterior fossa	No	--	--	--	--	Total res	CSF leak-age	No rec of 41 mons
Chandra et al., 2005 (4)	25, M	torticollis bilateral papilledema	No/No	Plain x-ray MRI	Midline of posterior fossa	No	C2-C3	AAD Occipitalization of the atlas platybasia	--	--	Total res OC fusion	No	No rec of 8 mons and mild restric-tion of neck move-ments
	23, M	headache neck pain restricted neck movements	No/No	Plain x-ray MRI	Midline of posterior fossa	No	C2-C3	basilar invagination Occipitalization of C1 arch AAD	--	--	res OC fusion	Aseptic meningitis	No rec of 15 mons

Table I: Cont.

Authors & year	Age (yrs) Sex	Clinical manifestation	Dermal sinus/ Infection	Radiological modality	Location	Rupture	Fusion	CVJ Malformations	Scoliosis	Other Malformations	Treat-ment	Complica-tions	Follow-up
Pai et al., 2007 (28)	3, M	rICP intermittent irritability	No/No	Plain x-ray CT scan MRI	Midline of posterior fossa	No	C2-C4	--	--	Cerebellar tonsillar herniation	res	--	No rec of 2 yrs
	1, M	fever vomiting occipital lump	Yes/--	Plain x-ray CT scan MRI	Right cer hemisphere	No	Fusion of several cervical vertebral	--	--	Several hemi-vertebrae of thoracic spine	res	Pseudo-meningo-cele	--
	2, M	hesdache epistaxis occipital lump	Yes/--	CT scan MRI	Midline of posterior fossa	No	--	--	--	--	Total res	No	No rec
Turgut , 2009 (40)	4, F	headache head tilt to left pea-sized occipital lump triad of KFS	Yes/No	Plain x-ray CT scan MRI/MRS Ultrasound Echocardio-graphy	Midline of posterior fossa	No	C4-C7	No	Scoliosis of cervical spine, concave to the left	No	Total res	CSF leak-age	No rec of 10 mons
Ramzan et al., 2011 (34)	5, F	Torticollis to left occipital lump triad of KFS cer signs on the right	Yes/No	Plain x-ray CT scan MRI	Midline of posterior fossa	No	C3-C5	No	--	No	Total res	No	No rec of 3 mons
Mclaughlin et al., 2013 (23)	47, F	rICP occipital lump triad of KFS cer signs paraparesis of lower extremity with marked patellar hyperreflexia sensory deficit	Yes/No	Plain x-ray CT scan MRI	Craniocer-vico Thoracic	No	Bodies and laminae of C2-T6	No	40° thoracic kyphosis	.. palatal fissure ¨ vertebral artery anterior to the lesion	Total res	No	No rec of 6 yrs
Current case	28, F	head tilt to left mirror movement triad of KFS cer signs	No/No	Plain x-ray CT scan CTA MRI Ultrasound Echocardio-graphy	Craniocervi-cal junction	Yes	C1-C7 T1-T3	Platybasia Basilar invagination bifid anterior C1 arch defects of posterior C1 arch Spina bifida occulta at level of C1-C7	Scoliosis of cervical spine, concave to the left, Scoliosis of first to third spine, concave to the right	Basilar artery fenestra-tion asymmetry of sternoclavicular joint intraspinal lipoma	Total res	No	No rec of 6 mons

Abbreviations: ***AAD*** *atlantoaxial dislocation;* ***cer*** *cerebellum;* ***C*** *cervical vertebra;* ***CSF*** *cerebrospinal fluid;* ***CT*** *computed tomography;* ***CTA*** *computed tomography angiography;* ***CVJ*** *craniovertebral junction;* ***KFS*** *Klippel-Feil syndrome;* ***mons*** *months;* ***MRI*** *magnetic resonance imaging;* ***MRS*** *magnetic resonance spectroscopy;* ***No.*** *number;* ***OC*** *occipitocervical;* ***rICP*** *raised intracranial pressure;* ***rec*** *recurrence;* ***res*** *resection;* ***T*** *thoracic vertebra;* ***yrs*** *years; -- unmentioned.*

Table II: Further Analysis of the 25 Reported Cases (Including the Present Case)

Details	No. (%)	No. Cases where data is specified
No. of Patients and Avg Age [range]yrs	25 and 19.7 [1-61]	
Female [Avg age] yrs	14 [26.3]	25
Male [Avg age] yrs	11 [11.4]	
Patients≤18yrs [Avg age] yrs	14 [6] (56%)	
Clinical manifestation	14 (58.3%)	
Dermal sinus	10 (41.7%)	
Occipital dimple or lump	10 (41.7%)	
rICP	9 (37.5%)	24
Triad of KFS	8 (33.3%)	
Cerebellar signs	7 (29.2%)	
Short neck or restriction of neck or head titling to one side meningitis	1 (4.2%)	
DC location		
Midline of posterior fossa	21 (84%)	25
CVJ	3 (12%)	
Cerebellar hemisphere	1 (4%)	
Types of KFS		
I	(62%)	
II	(33.3%)	21
III	(4.7%)	
unmentioned	4	
CVJ malformations		
Atlas assimilation	6 (60%)	
Basiocciput dysplasia (platybasia, basilar invagination)	3 (30%)	
Dysplasia of the lateral sclerotome or/and hypochordal bow (hypoplasia or Defects of anterior or/and posterior C1 arch)	2 (20%)	10
AAD	3 (30%)	
Spina bifida occulta	3 (30%)	
Ruptured DC	1	25
Mirror movement	3	24
Surgical resection		
Resection and OC fusion	3 (12.5%)	24
Detailed described resection	21	24
Total resection	18 (86%)	21
Partial resection	3 (14%)	21
Complication of post-operation		
CSF leakage	(8.3%)	24
Aseptic meningitis	1 (4.1%)	24
Pseudo-meningocele	1 (4.1%)	24
Follow-up		
Avg time [range] months	[3-72]	17
Tumor recurrence	0	
Mild restriction of neck movement of OC fusion	1	

Abbreviations: AAD *atlantoaxial dislocation;* ***Avg*** *average;* ***CSF*** *cerebrospinal fluid;* ***CVJ*** *craniovertebral junction;* ***DC*** *dermoid cyst;* ***No.*** *number;* ***OC*** *Occipitocervical;* ***rICP*** *raised intracranial pressure.*

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