Case Report

Intramedullary inflammatory mass dorsal to the Klippel–Feil deformity: error in development or response to an abnormal motion segment?

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Study design: Case study on a 45-year-old female with progressive weakness and paresthesias in her lower extremities and a magnetic resonance image of an intramedullary mass at the level of C5–6 and a Klippel–Feil (KF) deformity ventral to the lesion.

Objective: Present an interesting case of an intramedullary mass coexisting with a ventral KF deformity with review of the medical literature on intramedullary masses and cervical spine biomechanics.

Setting: New York city, New York, USA.

Methods: Case study with discussion of neurosurgical and neuropathological findings and review of the literature.

Results: The patient underwent open neurosurgical excisional biopsy of the intramedullary mass which revealed a non-neoplastic inflammatory mass that slowly resolved with medical management.

Conclusions: While no definitive etiology was found in this case we offer two interesting mechanisms: (1) maldevelopment of the cervical spine or (2) this inflammatory mass is in response to an abnormal motion at the level of the Klippel–Feil.

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Introduction

Klippel–Feil syndrome (KFS) has been associated with multiple other deformities or clinical abnormalities in the body.^{1–8} The KFS deformity is most often associated with other neurological deformities such as posterior fossa cysts or diastematomyelia.^{2,5} There are a few cases of KFS being associated with spinal cord tumors.^{3,4} To date most cases are thought to occur through an error in neurulation.^{2,3} We present the first case of an intramedullary nonspecific inflammatory mass occurring dorsal to the KF deformity in a developmentally normal adult female. The possible etiologies of this inflammatory mass and a review of previous spinal cord tumors in KFS will be discussed herein.

Case report

A 45-year-old female presented to the neurosurgery clinic with complaints of paresthesias and progressive weakness in her lower extremities. The patient had no other complaints and no significant medical history. The patient denied any history of trauma. On physical

examination, the patient had 4/5 strength bilaterally in lower extremities, increased reflexes 3+ in her lower extremities as compared to upper extremities and diminished light touch in lower extremities bilaterally, the remainder of the examination was normal. The patient brought with her a magnetic resonance image of her spine from an outside facility. The images revealed an enhancing expansile intramedullary mass at the level of C5–C6 with an intramedullary cyst extending above and below the mass with an incidental KF deformity at the level of C6–T1 (Figure 1). The patient was informed that a cervical laminectomy with excisional biopsy would be the best option for elucidation and possible treatment of this lesion. Owing to the KF deformity preoperative flexion–extension plain films were performed, which demonstrated no instability. Upon informed consent the patient underwent a C4–C7 laminectomy with excisional biopsy of the intramedullary mass. Multiple separate frozen sections were sent during the operation which revealed an inflammatory mass. After substantial specimens were obtained, a decision for subtotal resection was made to prevent postoperative deficits from a radical resection and extensive myelotomy. The patient suffered no deficits

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Figure 1 (a) Sagittal T1-weighted MRI with contrast demonstrating a intramedullary mass at the level of C5–6 with cord expansion and corresponding ventral KF deformity (b) Axial T1-weighted MRI revealing an expansile intramedullary mass at the level of C5–6

postoperatively and was discharged on postoperative day 2 from the hospital. Final pathology revealed a nonspecific inflammatory mass with reactive tissue. Postoperative MRI performed 1 month after surgery revealed total resolution of the mass. The patient has been followed for approximately 1 year postoperatively with no symptoms and complete resolution of the mass.

Discussion

KFS has been reported to be associated with numerous other developmental defects in multiple organ systems.^{1–8} Most cases of KFS are associated with anomalies that involve the central nervous system. There are very few cases of spinal cord tumors associated with KF deformity and to date, no intramedullary masses have ever been reported.^{3,4} Vaquero *et al* reported on a patient with KFS and an associated epidural fibroblastoma in the area of KF deformity.⁶ There is an interesting case of multiple meningiomas occurring in the cervical spine of a patient with KFS and atlantooccipital assimilation, however, a form fruste of another disorder was also entertained.⁴ Fibrous diastematomyelia has been reported in several cases of KFS and including a recent case of intradural epidermoid tumor associated with the diastematomyelia.^{1,4,8} Does our case just represent another example of maldevelopment or could this inflammatory mass be a response to a chronic biomechanical problem?

Abnormal motion with KFS deformity is well recognized.^{9,10} There are numerous reports of spinal cord injury occurring secondary to the predisposition for injury in KFS.^{9,10} Most surgeons would agree with a recent report recommending surgical intervention in patients with KFS only upon developing deficits.¹¹ KF deformities often have hypermobile segments above or below the level of ‘block’ vertebrae leading to increased chances for injury.¹⁰

A recent magnetic resonance imaging study demonstrated that cervical fusion produces abnormal motion at the level above and/or below the level of fusion.¹² Abnormal motion in any joint may incite an inflammatory reaction leading to the development of arthropathy or pannus formation.^{13,14} Spinal cord injury studies also demonstrate an inflammatory reaction leading to macrophage invasion and development of scar formation.¹⁵ Benzel coined the term ‘glacial instability’ which is spinal instability that is not overt and that does not pose a significant risk for the rapid development or progression of deformities but progresses gradually as in the motion of a glacier.¹⁶ Could this case represent glacial instability? Of interest is the development of this intramedullary lesion right at the tip of the KF deformity, that is, fulcrum. Could our patient at 45 years of age be suffering from merely an intramedullary inflammatory response secondary to abnormal motion?

This is the first report of a non-neoplastic inflammatory mass occurring adjacent to a KF deformity. How common are intramedullary inflammatory lesions? In a review of 212 patients operated on for spinal cord masses, nine were found to be non-neoplastic intramedullary lesions, of which only one was an inflammatory mass.¹⁷ Interestingly, they concluded that lack of spinal cord expansion associated with an intramedullary mass is the hallmark of a non-neoplastic lesion.¹⁷ A recent review of 38 patients who underwent spinal cord biopsy for intramedullary lesions found that

only 87% of neoplastic cases demonstrated cord expansion.¹⁸ In addition, they also found that 94% of patients with inflammatory lesions demonstrated spinal cord expansion.¹⁸ Our case further supports non-neoplastic intramedullary lesions ability to induce cord expansion.

We have presented the first case of a non-neoplastic intramedullary inflammatory mass dorsal to a KF deformity. While no definitive etiology was found in this case, we presented two interesting mechanisms: (1) maldevelopment or (2) response to an abnormal motion segment. It would be of great interest to retrospectively examine the previous reports of non-neoplastic inflammatory masses to determine the incidence of possible biomechanical etiologies or associated KF deformities.

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