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Posterior fossa dermoid: yet another cause of difficult airway?

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Abstract
Klippel-Feil syndrome and craniovertebral junction anomalies are known to pose challenges while the airway is being secured. These anomalies may occur in association with dermoid and epidermoid cysts in the posterior fossa. We present a case of posterior fossa dermoid with extracranial extension that caused severely restricted neck movement. As these anomalies seem to form part of a single, unnamed syndrome, the possibility of upper cervical spine abnormality leading to a difficult airway should be anticipated in patients with posterior fossa dermoid.

Key words Posterior fossa dermoid · Klippel-Feil syndrome · Difficult airway

Introduction

The most common site for an intracranial dermoid is the midline infratentorial segment of the posterior cranial fossa [1]. An association of dermoid and epidermoid cysts in the posterior fossa with Klippel-Feil syndrome [2–4] and craniovertebral junction anomalies [5,6] has been reported. Whether or not these anomalies are associated with dermoid cysts, they are known to pose challenges while the airway is being secured. However, a literature review failed to reveal mass lesion in the posterior fossa as a cause of difficult airway. We present a case of posterior fossa dermoid with extracranial extension that caused severely restricted neck movement.

Case report

A 30-year-old male nonobese patient, 168 cm tall, weighing 70 kg, was admitted to our hospital with the chief complaint of a swelling over the nape of the neck, which had been present since birth, and which was gradually increasing in size. He had had two episodes of generalized tonic clonic seizures in the past 5 years, for which he was taking phenytoin. He also had complaints of blurred vision, right upper-limb weakness, and ataxic gait. His past medical history was unremarkable. There were no other associated neurological abnormalities. Examination of the neck revealed a 5 × 4-cm swelling over the posterior aspect of the neck (Fig. 1). It was nontender, irreducible, and firm, with no leak, and negative transillumination. A dimple of 3 mm was present on the overlying skin, with a tuft of hair. The patient had a Mallampati grade II airway with an interincisor gap of 4.5 cm. The thyromental distance was found to be more than 6.5 cm. He had a short neck with low posterior hair line and severely restricted neck movements, indicating the presence of Klippel-Feil anomaly. All the routine preoperative investigations (ECG, peripheral oxygen saturation [S_{pO_2}] noninvasive blood pressure [BP]) were within normal limits. Preoperative magnetic resonance imaging (MRI, Fig. 2) showed a large posterior fossa space-occupying lesion (10 × 9 cm) with scalloping of the occipital bone. The lesion was well-encapsulated, and there was a clear-cut margin between the lesion and the floor of the fourth ventricle.

A provisional diagnosis of posterior fossa dermoid with extracranial extension was made, and excision of the same was planned. Awake fiberoptic intubation (FOI) was explained, but the patient was very apprehensive, and refused to consent to the procedure. Hence, FOI under general anesthesia was planned. The patient was premedicated with intramuscular glycopyrrolate 0.2 mg 1 h before the induction of anesthesia. In the operating theater routine monitors were attached. Anesthesia was induced with intravenous fentanyl 2 $\mu g \cdot kg^{-1}$ and propofol 2 $mg \cdot kg^{-1}$. Direct laryngoscopy revealed a Cormack-Lehane grade-III view of the

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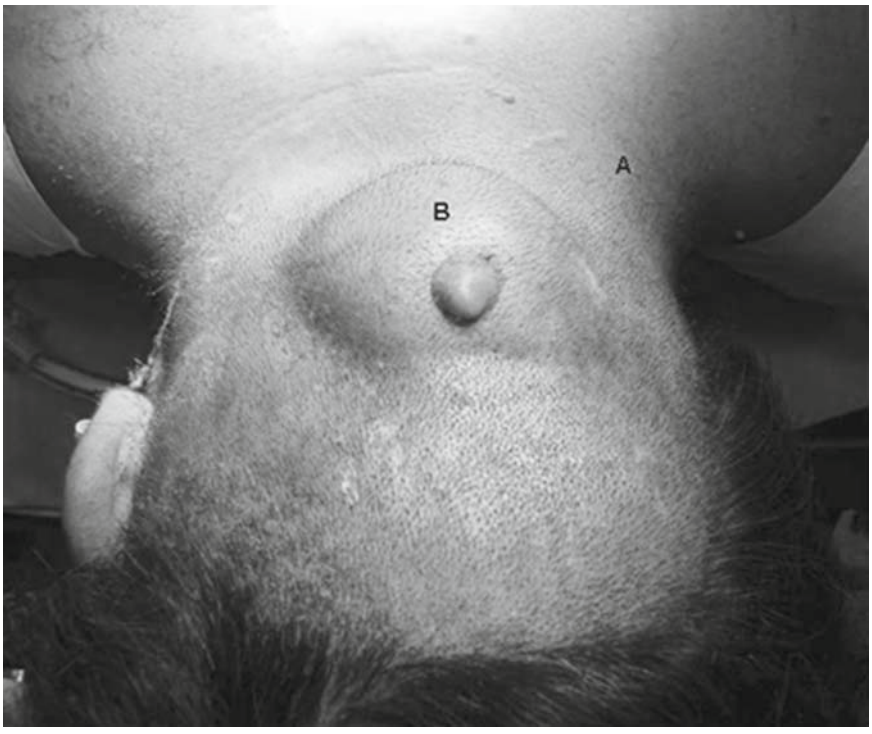

Fig. 1. Patient positioned prone with head fixed on a Mayfield clamp, showing short neck (*A*) and a mass protruding from the posterior fossa (*B*)

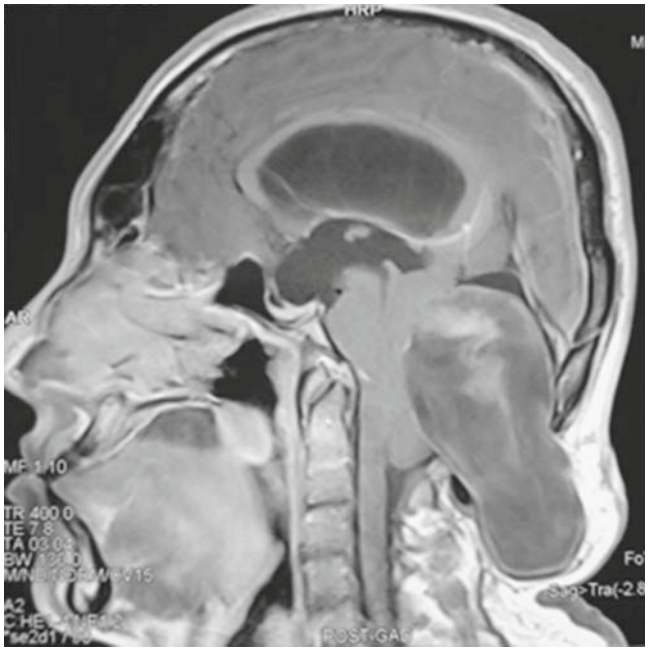

Fig. 2. Sagittal T1-weighted magnetic resonance image, showing well-defined, hyperintense posterior fossa mass lesion, compressing the cerebellum and fourth ventricle and protruding extracranially through scalloped occiput

larynx. As the patient's neck extension was severely restricted, further attempts at laryngoscopy were avoided. A classic laryngeal mask airway (LMA), size 4, was placed for ventilation, as the ventilation with a face mask was difficult. Rocuronium 1 mg·kg^{-1} was administered and anesthesia was deepened with a further bolus of propofol 50 mg. The LMA was removed and orotracheal intubation was performed by using a fiberoptic bronchoscope, at the first attempt. Anesthesia was maintained with oxygen/nitrous oxide, isoflurane, and intermittent doses of fentanyl and rocuronium. Other monitoring parameters included end-tidal CO_2, arterial BP, temperature, and urine output. The patient was placed in the prone position. A sub-occipital craniectomy was performed to excise the lesion. The uneventful surgery lasted for 4 h. At the end of the procedure, the patient was turned to the supine position and the anesthetic agents were discontinued. Residual neuromuscular block was reversed and the trachea was extubated after the patient became fully awake. After a quick neurological assessment, the results of which were normal, the patient was shifted to the post-anesthesia care unit. The further hospital stay of the patient was unremarkable, and he was discharged on the eighth post-operative day with advice for follow-up.

Discussion

Intracranial dermoid tumors constitute 0.1% to 0.7% of all intracranial neoplasms [2]. These tumors are developmental abnormalities; they result from the trapping of the surface ectoderm during closure of the neural tube around day 28 of gestation. Klippel-Feil syndrome,

which is characterized by the classic triad of short neck, low posterior hairline, and restricted range of neck movement, is a developmental anomaly of the craniovertebral region. Other features include central nervous system, cardiac, and renal abnormalities; fused cervical vertebrae; scoliosis; torticollis; and deafness. The cause of this anomaly is thought to be failure of division of the mesodermal somites in the cervical region, which causes abnormal fusion of the cervical vertebrae and the restricted neck movements. This failure also occurs towards the end of the fourth week of gestation, i.e., at 26–28 days. Thus, the maldevelopment which leads to the formation of posterior fossa dermoids and Klippel-Feil anomaly occurs in the identical location and the identical time period of embryological development. This fact may possibly be responsible for the occurrence, albeit uncommon, of both conditions together. This idea is borne out by a review of the literature, which reveals reports of posterior fossa dermoid associated with Klippel-Feil syndrome [2–4] and other craniovertebral anomalies [5,6]. In our patient, the classic triad of Klippel-Feil anomaly was present, though the common feature of fused cervical vertebrae was not evident. However, the presence of restricted neck movement with a difficult airway suggested an abnormality of the cervical spine, which was not evident from the MRI scan. The reported occurrence of craniovertebral junction anomaly and upper cervical anomaly with posterior fossa dermoid [5,6], and the fact that both conditions are related embryologically, led us to believe that such an association was present in our patient. Nevertheless, in this patient, the large size of the lesion in the posterior fossa seemed to be the culprit that was responsible for the restricted neck extension.

To conclude, because posterior fossa dermoid, Klippel-Feil syndrome, and craniovertebral junction anomalies seem to form part of a single unnamed syndrome, the presence of any one of them should lead to suspicion about the existence of the other. Hence, we suggest that, in patients with posterior fossa dermoid, the possibility of upper cervical spine involvement leading to a difficult airway may be anticipated.

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