Atypical presentation of a posterior fossa tumour: A case report

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Abstract

BACKGROUND
We describe a case of a patient with a meningioma in the posterior fossa presenting atypically with an isolated unilateral vocal cord palsy causing severe respiratory distress. This is of interest as the patient had no other symptomatology, especially given the size of the mass which would typically cause a pressure effect leading to neurological and auditory symptoms.

CASE SUMMARY
This case report describes a 48-year-old male who is married with two children and employed as a car guard. He has a background medical history of asthma for the past ten years controlled with an as needed beta 2 agonist metered dose inhaler. He initially presented to our facility with severe respiratory distress. He reported a one-week history of shortness of breath and wheezing that was not relieved by his bronchodilator. He had no constitutional symptoms or impairment of hearing. On clinical examination, the patient’s chest was “silent”. Our initial assessment was status asthmaticus with type 2 respiratory failure based on the history of asthma, a “silent chest” and the arterial blood gas results.
Management was commenced with continuous nebulisations, intravenous magnesium sulphate infusion and intravenous hydrocortisone. Pharmacological management was ineffective, leading to the patient being subsequently intubated, ventilated and transferred to the intensive care unit.

During his stay in the intensive care unit, the patient had a persistent respiratory acidosis with subsequent Klebsiella pneumonia and Ciprofacter koseri cultured on the endotracheal aspirate. Blood culture was positive for Acinetobacter baumantii for which he was treated with intravenous amoxycillin-clavulanate and gentamycin. The patient coped well while ventilated and was assessed as being fit for extubation. Upon extubation he immediately became distressed and developed stridor; he was promptly re-intubated and ventilated. Examination of his vocal cords under anaesthesia showed a left vocal cord palsy with associated oedema over the arytenoids, granulation tissue over the left vocal cord and trauma to the anterior trachea. The sub-glottis was normal. He underwent a tracheostomy and was weaned off the ventilator and stepped down to the general medical ward, where he was managed by a multidisciplinary team including a speech therapist, otorhinolaryngology, specialist physicians and a nursing team who provided tracheostomy care. He was referred to the speech therapist as part of rehabilitation and for tracheostomy care to assist with speech, communication and swallowing issues.

As part of the diagnostic workup of the unilateral left vocal cord palsy, we did a contrasted computed tomography scan of his head, neck and chest. The scan identified a large densely calcified extra-axial right mass in the posterior fossa measuring 4x4x4 cm that extended into the internal auditory meatus, jugular fossa and hypoglossal canal with inferior extension into the upper spinal canal via the foramen magnum, causing right supratentorial hydrocephalus. A subsequent MRI revealed a large mass in the right cerebellar pontine angle with associated expansion of the internal acoustic meatus and extension into the upper cervical canal, jugular fossa and right paravertebral region.
(figure 1). The patient underwent neurosurgery for de-bulking and an excision biopsy was done. The tumour tissue demonstrated extensive dystrophic calcification forming concentric lamellate psammoma bodies, and cellular areas showed nuclei pseudo-inclusions. These features are consistent with a psammomatous meningioma (World Health Organisation Grade 1). Post neurosurgery, he was referred for radiation therapy.

CONCLUSION

A posterior fossa meningioma of such a large size and with extensive infiltration rarely presents with an isolated unilateral vocal cord palsy. The patient’s chief presenting feature was severe respiratory distress, which combined with his background medical history of asthma, was misleading. Clinicians should thus consider meningioma as a differential diagnosis for a unilateral vocal cord palsy even without audiology involvement.

INTRODUCTION

We describe a rare case of unilateral vocal cord palsy secondary to a posterior fossa tumour presenting atypically, with severe respiratory distress. The patient had a background history of asthma, and initial assessment was that of status asthmaticus based on history of asthma, severe respiratory distress and a “silent chest.” He was intubated and ventilated due to respiratory failure. In retrospect, we assessed him as having upper airway obstruction secondary to unilateral vocal cord paralysis, based on stridor immediately after extubation and as evidenced by direct laryngoscopy. To identify the underlying cause of the unilateral vocal cord palsy, we undertook imaging of the head, neck and chest, which revealed a large cerebellar-pontine tumour with abutment of adjacent structures. Debulking neurosurgery with excision biopsy demonstrated a psammomatous meningioma. The patient is currently awaiting radiotherapy.
CASE PRESENTATION

Chief complaints
acute onset shortness of breath

History of present illness
none, sudden onset

History of past illness
known asthmatic

Personal and family history
none

Physical examination
no air entry bilaterally
severe respiratory distress

Laboratory examinations
Type 2 respiratory failure on blood gas

Imaging examinations
N/A

FINAL DIAGNOSIS
psammomatous meningioma

TREATMENT
Debulking neurosurgery with excision biopsy followed by radiotherapy
OUTCOME AND FOLLOW-UP
Patient transferred to Quaternary level facility

DISCUSSION
Isolated left sided vocal cord palsy with respiratory failure is a rare presentation of a tumor in the posterior fossa. In this patient, the tumor resulted in a mass effect with resultant effacement and displacement of the pons and medulla to the left; this may explain the left sided vocal cord paralysis. An excision biopsy demonstrated a psammomatous meningioma.

Unilateral vocal cord paralysis can be asymptomatic or can present with hoarse voice, dysphonia, dysphagia, aspiration and coughing. (1,2) Patients may recover spontaneously in some instances, or the contralateral cord may compensate for its dysfunctional counterpart. (1) The most commonly affected side for an isolated vocal cord paralysis is the left side, as was the case in our patient. (1) This is due to the longer course of the left recurrent laryngeal nerve compared to the right side making it more vulnerable to damage, especially in the mediastinum. Most patients present in the 5th or 6th decade of life, on average at age 53 years. (2) Diagnosis of a vocal cord paralysis is dependent on confirmation during indirect laryngoscopy or laryngeal endoscopy, (2) as confirmed in our patient.

The underlying cause of vocal cord palsy varies based on geographic location. Malignancy contributes to 34% of cases of vocal cord palsies. (2) Primary malignancy only accounts for 7.5% while secondary pressure effects and nerve damage accounts for 85% of these cases presenting with cord paralysis. (3) If not laryngeal in origin, abnormalities of the thyroid gland, oesophagus, mediastinum and lung are the most common causes for this presentation. An otolarynological assessment includes otoscopy to exclude a cholesteatoma. Flexible nasolaryngoscopy or rigid laryngoscopy can also
be done to exclude infiltration of the primary lesion. (4). Radiological investigation, namely, a computed tomography scan from the base of the skull to the upper mediastinum, is needed to identify the possible cause of unilateral vocal cord palsy. This is the approach that we followed.

Senior et al (4) reported a case involving a 78-year-old female who presented with progressive dysphonia and dysphagia. She also had further neurological fallout which included progressive left-sided hearing loss with normal otoscopic examination. Flexible naso-endoscopy showed a unilateral vocal cord palsy. Magnetic resonance imaging (MRI) revealed a primary cerebellopontine angle meningioma arising from the jugular foramen. Another case report described a 34-year-old man who presented with dysphagia, loss of taste and dysarthria. Laryngoscopy showed a unilateral left vocal cord palsy. Electrodiagnostic study confirmed paralysis of the lower cranial nerves (IX to XII). Further imaging with brain MRI revealed a left cerebellopontine angle meningioma. This patient was subsequently diagnosed with Collet-Sicard syndrome (unilateral lower cranial nerve paralysis) secondary to the cerebellopontine angle mass. (5)

Our patient differed from the aforementioned cases in that he had vocal cord palsy with respiratory failure, eventually requiring a tracheostomy. The late presentation may have accounted for the severe respiratory distress that, to the best of our knowledge, has not been reported with other cases of meningioma. One may speculate that his asthma may have also contributed to his severe respiratory distress.

CONCLUSION

A meningioma in the posterior fossa of such size and infiltration as described in this case, rarely presents asymptptomatically or with a unilateral vocal cord palsy. Larger meningiomas are mostly symptomatic due to tissue compression and subsequent
oedema. Despite this patient having both of these findings, the symptoms were limited. Cases described previously in the literature presented with audiological and vocal cord involvement, but the presentation of vocal cord involvement in isolation is a rare finding. Early recognition in such cases would lead to better prognosis; especially with the possibility of resection. Imaging, histopathology and then prompt referral to oncology is required for management planning. Clinicians should thus consider meningioma as a differential diagnosis for a unilateral vocal cord palsy, even without audiology involvement. In this case, the diagnosis in a man with isolated unilateral vocal cord palsy and respiratory failure was confounded by the history of asthma.
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