A 2-year-old previously healthy girl was brought to her pediatrician with the chief complaint of persistent noisy breathing. Two months earlier, the child had an upper respiratory tract infection (URI) with rhinorrhea, cough, noisy breathing, and wheezing. All symptoms had resolved except the abnormal breathing. Physical examination findings were unremarkable. A lateral neck x-ray film demonstrated subglottic narrowing, thought to be consistent with croup. Laryngoscopic examination by an otolaryngologist did not reveal any pathology.

The child continued to have stridor for 2 months after the URI—especially at night—and for an additional month following administration of a short course of outpatient oral prednisone. The corticosteroid relieved the stridor only temporarily, and the patient returned for evaluation. Stridor occurred day and night but was more pronounced at night—particularly when the child was supine. Her parents noted that the stridor was of variable intensity interspersed with periods of quiet. A review of systems was negative. There was no history of hoarse voice, foreign-body aspiration, problems with swallowing, pain, emesis, or gait changes. The child's parents considered her voice to be normal. Vital signs were also normal: the child appeared comfortable and without distress, despite loud inspiratory stridor at rest. Her oropharynx was clear, with mildly hypertrophic tonsils. There were no masses or lymphadenopathy on neck examination. Other than the inspiratory stridor, chest auscultation revealed clear, equal breath sounds bilaterally. There were no other remarkable findings. Extraocular movements were intact, and the tongue was in midline. There was no papilledema, ptosis, or facial asymmetry.

Because laryngoscopy had revealed no abnormalities, the child underwent flexible bronchoscopy to evaluate the lower airway. This revealed collapsing, inflamed, edematous arytenoids and paradoxical movement of the vocal cords on inspiration. There was no overt collapse of the epiglottis, no mass or hemangioma, and no tracheal stenosis or compression. The bronchi appeared normal. After bronchoscopy, significant stridor developed and the child required supplemental oxygen. Her symptoms improved a day after corticosteroid and epinephrine administration. She was discharged with the diagnosis of laryngomalacia with paradoxical movement of the vocal cords and was treated with lansoprazole for suspected gastroesophageal reflux. Despite proton pump inhibitor use, the patient continued to have stridor. Six weeks later, dyspnea on exertion developed. However, there was no stridor or hoarseness during vocalization. The patient underwent microlaryngoscopy. Paradoxical movement of the cords with inspiration was again present but was noted to disappear during deep anesthesia induction and with application of topical lidocaine to the vocal cords. No other pathology was found. Increased stridor again developed postoperatively. The patient required oxygen supplementation and responded well to corticosteroid and epinephrine therapy.

Because of the reversible, paradoxical vocal cord movements found during microlaryngoscopy, MRI scans were obtained to evaluate for possible Arnold-Chiari malformation. Scans revealed posterior displacement of the medulla by a 0.8 X 1.6 X 2.7-cm homogeneously enhancing extra-axial mass surrounding the anterior and lateral medulla (Figure). The mass encased both vertebral arteries, the posterior inferior cerebellar artery, and cranial nerves IX through XII bilaterally.
Figure - A T1-weighted MRI scan with gadolinium contrast demonstrates posterior displacement of the medulla by a homogeneously enhancing mass that extends anterior and superior to the medulla.

Progressive respiratory failure soon ensued, and the patient required intubation and mechanical ventilation. She underwent a left lateral craniotomy, C1 laminectomy, and intradural exploration, with partial resection of the tumor and decompression of the cervicomedullary and vertebral artery junction. The tumor was adherent to the brain stem without clear cleavage planes and was therefore deemed unresectable. On review by neuropathology, the tumor was classified as an anaplastic astrocytoma with features of a pleomorphic xanthoastrocytoma, World Health Organization grade III. After tracheostomy and gastrostomy tube placement, radiation and chemotherapy were initiated. Now, 1 year later, the tumor has minimally increased in size and the patient is still receiving chemotherapy. She is alert, cooperative, has full visual fields, and can successfully talk "over her tracheostomy." She has normal tone and strength in her upper extremities but has decreased tone and strength in her lower extremities. She is nonambulatory.

**STRIDOR**

Stridor is a high-pitched sound heard during inspiration caused by turbulent airflow through a narrowed segment of the upper respiratory tract. Acute stridor is a common symptom that affects many children and often reflects an infectious cause or foreign-body aspiration. Chronic stridor is a
rare finding that often indicates more permanent structural abnormalities.

Laryngomalacia is the most common cause of chronic stridor in the newborn and young infant and accounts for approximately 27% to 39% of cases. Nasopharyngoscopy often reveals inward collapse of the aryepiglottic folds and cartilage with supraglottic structures collapsing during crying or while the infant is supine. Approximately 80% of cases resolve by 18 months of age. Gastroesophageal reflux may also exacerbate laryngomalacia or may directly cause stridor from acute airway inflammation.

Tracheomalacia, another common source of chronic stridor, is caused by significant malformation of the cartilage rings and narrowing of the compliant trachea. Laryngomalacia and tracheomalacia often represent delayed maturation of the supporting structures of the larynx and trachea. Paradoxical vocal cord movement—also known as vocal cord dysfunction—is abnormal adduction of the vocal cords during the respiratory cycle, usually during inspiration, that produces airflow obstruction at the level of the larynx. Signs and symptoms include cough, inspiratory/expiratory wheeze, stridor, throat tightness, and reflux. Patients with paradoxical vocal cord movement are often treated for asthma or spasmodic croup with corticosteroids or bronchodilator medications before the diagnosis of paradoxical vocal cord movement is made by direct visualization of the cords.

Causes of paradoxical vocal cord movement include gastroesophageal reflux, asthma, cortical injury of upper and lower motor neurons, brain stem compression (by Arnold-Chiari malformation, cerebral aqueductal stenosis, or tumors), and movement disorders, as well as stress, anxiety, and other psychological factors. During normal inspiration, the vagus nerve (cranial nerve X) stimulates the posterior cricoarytenoid muscles to abduct the vocal cords. On expiration, tonic contraction by the lateral cricoarytenoid adds the vocal cords and the majority of the glottic aperture narrows as the posterior cricoarytenoid muscles relax. Changes in vagal innervation may alter laryngeal tone and lower the threshold for stimuli to produce vocal cord spasm or to precipitate abnormal adduction of the vocal cords. Of note, left vagus nerve stimulation-used to reduce seizure frequency in patients with refractory seizures-has been shown to cause left vocal cord adduction. Both hydrocephalus itself and ventricular-peritoneal shunting to relieve increased intracranial pressure can cause stridor in children; this may be related to compression of or traction on the vagus nerve.

VOCAL CORD PARALYSIS SECONDARY TO A CNS TUMOR

Vocal cord paralysis or paresis as the initial presentation of a CNS tumor is rare. Ours is the only reported case in the English literature of a brain stem tumor presenting as isolated chronic intermittent stridor. This was determined by a computer search of the literature from 1950 to 2007, using the key words brain tumor, stridor, ependymoma, medulloblastoma, astrocytoma, and pleomorphic xanthoastrocytoma.

In 1987, Stern reported a case of a 60-year-old man with acquired hydrocephalus, hiccups, and paroxysmal laryngeal stridor who was found to have a calcified ependymoma on the floor of the fourth ventricle. In 1990, Ross and Ward reported 2 cases of children with intracranial tumors who initially presented with hoarseness and stridor. In a retrospective chart review of 200 children with brain tumors diagnosed between 1975 and 1993, Keene and coworkers reported 19 patients (9.5%) with brain stem tumors. Signs and symptoms at presentation included gait difficulties (83%), "cross eyes" (50%), headaches (25%), vomiting (25%), and swallowing difficulties (8%). No cranial nerve X involvement was noted on initial examination. In a retrospective chart review of 200 children with brain tumors diagnosed between 1998 and 2001, Wilne and associates reported that the initial signs and symptoms at the time of presentation were headache (41%), vomiting (12%), ataxia (11%), visual problems (10%), educational or behavioral problems (10%), and seizures (9%). Of 175 children with abnormal neurological findings at the time of presentation, 86 (49%) had cranial nerve abnormalities. According to personal communication with the authors, 13% of these children had single cranial nerve findings; no vagal nerve abnormalities were noted. Habermann and coworkers reported late detection of an astrocytoma of the cerebellum and brain stem in a 6-year-old boy who initially presented with hypernasal speech that did not respond to speech therapy. One-sided vocal fold palsy with ipsilateral soft-palatal palsy led to a search for a CNS
A Brain Tumor Presenting as Chronic Intermittent Stridor
Published on Psychiatric Times (http://www.psychiatrictimes.com)

Faddoul and Meguid\textsuperscript{16} reported a case of a brain stem tumor in a 4-month-old boy who initially presented with recurrent aspiration pneumonia. The tumor was discovered after the development of additional neurological abnormalities, including left-sided weakness, left head tilt, left eye ptosis, right tongue deviation, and abnormal gag reflex.

Astrocytomas are the most common primary brain tumors, accounting for 52\% of primary malignant brain tumors.\textsuperscript{17} Infratentorial location predominates in the pediatric population, whereas in adults, most tumors arise above the tentorium.\textsuperscript{18} Long-term survival for patients with high-grade lesions, such as anaplastic astrocytoma, is poor despite advances in surgery, radiation, and chemotherapy. Pleomorphic xanthoastrocytoma (PXA) is typically a low-grade tumor,\textsuperscript{19} but high-grade anaplastic lesions such as our patient's are not uncommon. PXA generally occurs in children and young adults and appears superficially along the cortex or leptomeninges with a predilection for the temporal lobe.\textsuperscript{20} However, infratentorial lesions have been reported.\textsuperscript{21,22} Because these tumors tend to grow slowly, affected patients often come to medical attention months after symptoms first appear. The extent of surgical resection is the most important prognostic factor. Overall survival is 81\% at 5 years and 70\% at 10 years.\textsuperscript{23} Unlike the more common pilocytic astrocytoma, PXAs may undergo malignant transformation in the pediatric population, especially at the time of recurrence. The rate of malignant transformation is 10\% to 20\%. Time to transformation varies from months to years.\textsuperscript{24,25}

**BOTTOM LINE**

We present a child with isolated chronic, intermittent stridor whose underlying diagnosis was a brain stem astrocytoma. The typical neurological signs and symptoms seen with CNS tumors (headache, irritability, vomiting, ataxia, papilledema, focal neurological defects) were absent. The clue to final diagnosis was anesthesia-reversible paradoxical movement of the vocal cords that suggested ongoing bilateral vagal nerve stimulation as the cause of stridor.

**References:**

A Brain Tumor Presenting as Chronic Intermittent Stridor

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