Congenital midline nasal mass: cases series and review of the literature

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Encephalocele, glioma and dermoid cyst are the most common midline nasal masses. Given their potential for intracranial extension, prompt treatment is necessary to prevent complications. Herein, we present two cases of midline nasal masses. A comparison was made to delineate the differences between their clinical courses, treatments and outcomes. Case 1 was a baby girl with respiratory distress beginning at birth. Nasal glioma without definite intracranial extension was present. The mass was completely excised with the aid of a video-assisted endoscope without complications. At follow-up two years after surgery, no recurrence was noted. Case 2 was a two-year-old boy with a midline nasal dermoid cyst. Extirpation of the lesion through a vertical-dorsal approach was performed. He was discharged three days after surgery with a satisfactory aesthetic result.

Key words: nasal glioma, dermoid cyst, encephalocele.

Congenital midline nasal masses are rare, occurring once in every 20,000-40,000 births. They are often misinterpreted, and tend to be neglected. These tumors are mostly seen in newborns, infants and children, and are rarely found in adults. Midline nasal masses may be the result of dermoids, gliomas, encephaloceles, epidermoid cysts, and nasal polyps. Although most causes of midline nasal masses are relatively benign, the potential for congenital midline nasal masses to cause disfigurement and destruction of the cranial base should not be underestimated. Because of their possible connection to intracranial structures, a complete neuroradiologic evaluation is crucial before determining the proper management. The interpretation of computed tomographic (CT) and magnetic resonance (MR) images can be difficult, but imaging is useful in differentiating nasal gliomas from other congenital nasal masses. The presence of a fibrous stalk may be associated with an interconnection with the cranium, cranial defects and cerebrospinal fluid (CSF) leakage. Without prompt diagnosis and management, potential fatal meningitis may result. Treatment of nasal masses requires a well-planned, careful surgical resection, with endoscopic procedures playing an ancillary role[1].

Case Reports

Case 1

A three-day-old girl was seen shortly after her birth for the treatment of occasional central cyanosis with inspiratory stridor during feeding. At first, choanal atresia and laryngomalacia were suspected. Fiberoptic endoscopic examination was performed by an otolaryngologist and a whitish-pink, firm, slightly pulsatile mass was clearly seen over the nasopharynx, indicating a possible congenital nasal mass. Although the results of a Furstenberg test were negative, and the mass did not expand while crying, an actual or potential central nervous system connection

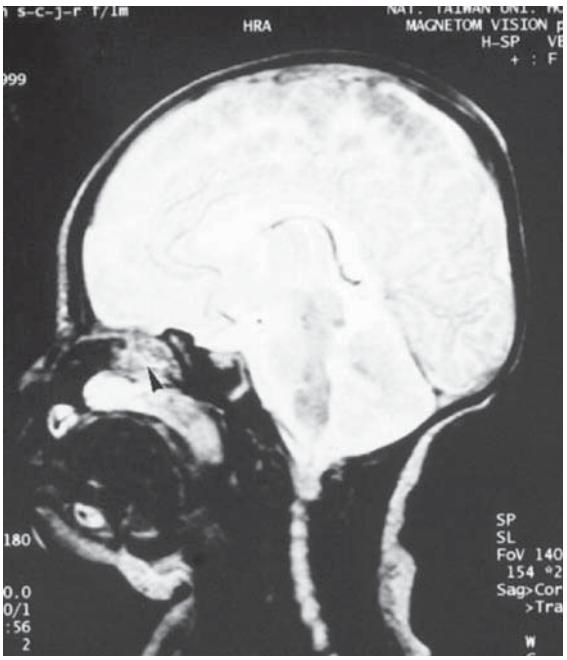

Fig. 1. Case 1. MRI without enhancement (T2WI), sagittal view. A mass lesion is noted in the nasopharyngeal region (arrowhead) without defect at the base of the anterior cranial fossa or sellar floor.

was considered. Subsequent microlaryngoscopic and bronchoscopic examination revealed the presence of an omega-shaped epiglottis and floppy arytenoid cartilage and suggested that airway obstruction such as laryngomalacia and tracheomalacia co-existed with the nasal mass. MRI of the brain revealed a midline mass lesion located in the nasopharyngeal region (Fig. 1). There was no direct evidence of intracranial extension of the lesion. CT study of the basal cranium found no apparent bony defect at the base of the anterior cranial fosse or the sellar floor. However, a 2 mm slit over the sphenoid was observed (Fig. 2), indicating a minor defect in the integrity of the skull base.

Surgical resection was performed under the guidance of a pediatric sinoscope. The tumor mass and its broad base attached to the upper septal portion of the choana and bulging anteriorly into the left nasal cavity was completely removed (Fig. 3). No interconnection between the intracranial space and the tumor itself was noted. Postoperatively, the patient showed remarkable improvement in respiratory function. There was no sign of nasal bleeding, CSF leakage or meningitis. The patient was discharged one week after the surgery. Histopathological examination was consistent with nasal glioma (Fig. 4). At the follow-up two years after surgery, no sign of tumor recurrence was noted.

Case 2

A two-year-old boy was seen in our clinic due to a right external nasal mass noted after a blunt nasal injury. The lesion fluctuated in size and pus-like discharge occasionally was expelled from a sinus-like opening. No symptoms such as nasal obstruction, dyspnea, periorbital swelling, or disturbed vision were observed by his parents or physicians. Physical examination revealed a tumor measuring 0.8 × 0.6 cm over the right nasal root (Fig. 5). The tumor was soft and displayed blended discoloration. CT of the head showed a soft tissue mass over the subcutaneous space of the right nasal root, extending into the dorsal aspect of the nasal bone.

No intranasal abnormality was found during sinoscopic examination. Extirpation of the lesion through a vertical-dorsal approach was performed, revealing a cystic mass containing hair appendages and greasy whitish materials

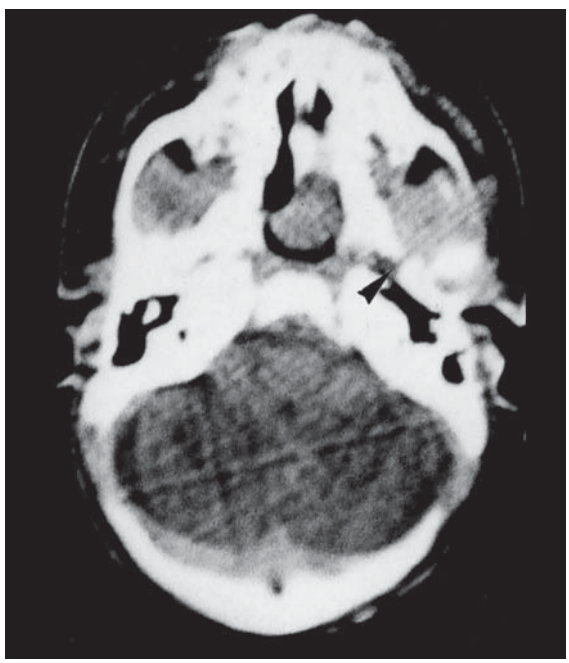

Fig. 2. Case 1. CT without enhancement, axial view. A mass is noted in the nasopharyngeal region without bony defect at the base of the anterior cranial fossa or the sellar floor, demonstrating a 2 mm slit over the sphenoid bone (arrowhead).

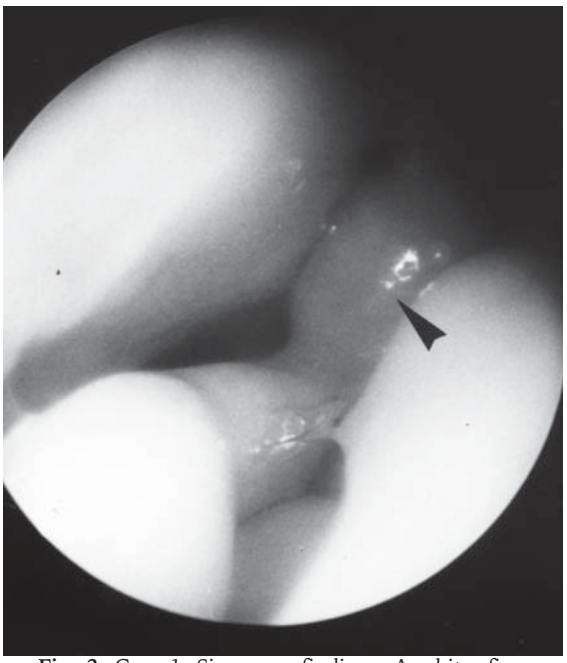

Fig. 3. Case 1. Sinoscopy findings. A white, firm mass (arrowhead) is seen raised from the upper septal portion of the choana and bulging anteriorly into the left nasal cavity and posteriorly to the nasopharynx.

(Fig. 6). The tumor was located in the subcutaneous space of the right nasal root. Extension of the tumor to the dorsum of the right nasal bone via a bony defect deep into the submucosal space of the contralateral nasal cavity was observed during the operation. He was discharged three days after surgery with a satisfactory aesthetic result. Histopathological examination was consistent with a dermoid cyst.

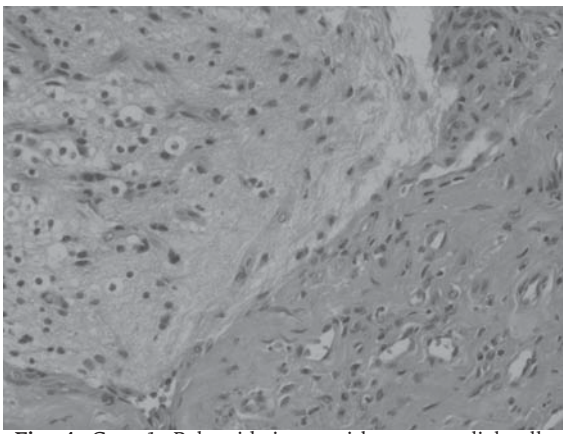

Fig. 4. Case 1. Polypoid tissue with mature glial cells is dominant. Immunohistochemical staining for glial fibrillary acidic protein (GFAP) and neurofilament (NF) is positive, and confirms the neurological origin of the lesion. Together, with the absence of a neuronal component, glial heterotopia is considered (x10).

Discussion

Laryngomalacia is the most frequent cause of congenital infantile stridor. However, its clinical course and associations with other conditions are not fully understood. It is first noted at birth and usually resolves spontaneously by the age of two years. In severe cases, it may lead to life-threatening situations such as airway obstruction, cor pulmonale and failure to thrive, and in these patients, surgery is mandatory.

In the first case presented, we treated a life-threatening emergency that manifested shortly after birth; the patient suffered from respiratory distress caused by an obstructing nasal tumor and laryngomalacia. Stridor in this infant was potentially serious and required urgent treatment. Resolution of respiratory symptoms immediately after removal of a mass does not

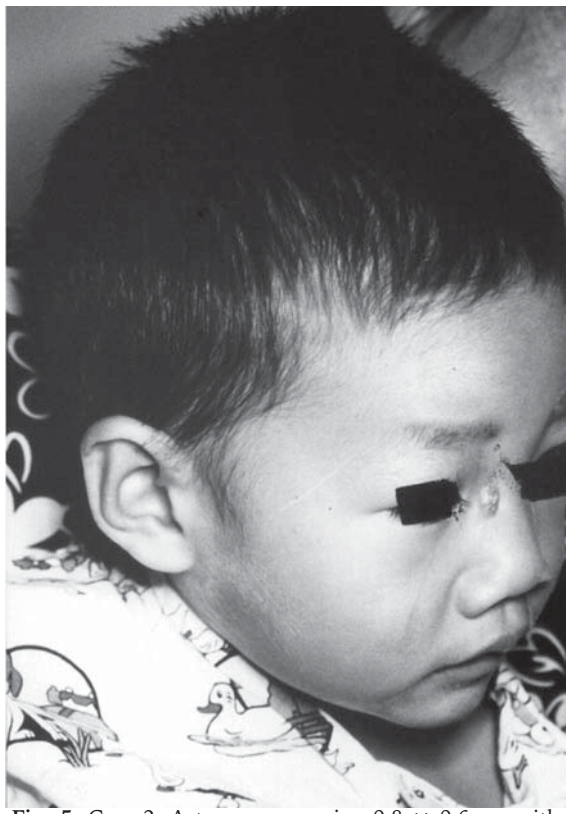

Fig. 5. Case 2. A tumor measuring 0.8 × 0.6 cm with blended discoloration is observed over the right nasal root.

always occur because tracheal deformity and laxity may sometimes take months or years to resolve. Fortunately, in this case, respiratory symptoms resolved as soon as the intranasal tumor had been excised surgically.

The differential diagnosis of congenital midline nasal masses that result from aberrant embryologic development includes nasal gliomas, dermoid cysts and encephaloceles[2]. Nasal gliomas are benign, congenital, midline tumors that have the potential for intracranial extension. They are thought to be encephaloceles that have lost their intracranial connection and meningeal continuity. In 15%-20% of cases, a fibrous stalk connects them to the intracranial space. These tumors are rarely associated with a bony cranial defect and CSF leakage. Sixty percent of gliomas are extranasal, 30% intranasal, and 10% are extranasal and intranasal. Clinically, these masses are firm, non-compressible, non-pulsatile, gray or purple lesions that obstruct the nasal cavity intranasally and cause deformity extranasally. Histologically, these tumors are made up of astrocytic neuroglial cells interlaced with fibrous and vascular connective tissue that is covered with skin or nasal respiratory mucosa. The treatment of choice is surgical excision. Inadequate primary excision results in a 4% to 10% recurrence rate. Hence, a thorough preoperative evaluation is indispensable to delineate the exact site and extension of the tumor and to plan an appropriate surgical approach. The preferred approach is excision via a frontal craniotomy for patients in whom the glioma has an intracranial connection, who have CSF rhinorrhea, or who suffer recurrent episodes of meningitis. Otherwise, a transfacial lateral rhinotomy is advised[3]. Intranasal endoscopic surgery is considered appropriate for the removal of intranasal gliomas that do not have intracranial extension. The procedure is less invasive and does not result in postoperative facial deformity[4]. Thus, unnecessary intracranial procedures are avoided without increasing the risk of ensuing complications[5]. In our first patient, the tumor appeared to be a benign lesion. Because recurrence is rare, a conservative cosmetic surgical technique was chosen for this glioma with no proven intracranial extension.

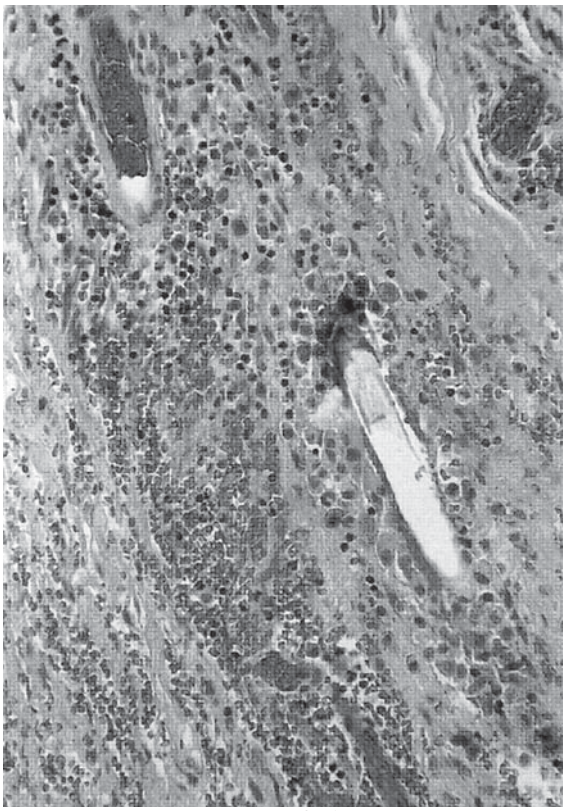

Fig. 6. Case 2. Operative findings. A cystic mass containing hair appendages and greasy whitish materials in the subcutaneous space of the right nasal root was noted. Microscopic examination was consistent with a dermoid cyst.

Dermoid cysts may present with a cutaneous midline nasal dimple or sinus, usually contain hair, and occasionally may expand the nasal septum or form a bifid septum[6]. These are rare developmental teratomatous lesions composed of ectodermally derived stratified squamous epithelium and mesodermally derived skin adnexal structures. Dermoid cysts affecting sites within the head and neck may be found in the frontotemporal/lateral brow area, central nasal area, oral cavity, and lateral neck, as well as other sites[7]. Surgical extirpation is the treatment of choice for nasal dermoids in the pediatric population. Approaches include vertical-dorsal, lateral, and transverse rhinotomy, and external rhinoplasty[8]. The last procedure, with a medial crura section, involves a wide surgical approach, with low recurrence rate and good aesthetic results[9].

The major considerations in the differential diagnosis of congenital midline nasal masses are encephalocele and nasal polyps. A nasal encephalocele is a protrusion of part of the

cranial contents through a defect in the skull, which may cause nasal obstruction. They are always transilluminative and have a dural connection, positive Furstenberg test, and are associated with meningitis[10]. Nasal polyps are very rare in children under five years old and, if present, are associated with cystic fibrosis. They usually do not cause osseous abnormalities.

Recent advances in CT and MRI have improved imaging of the airway in general, and in the pediatric population in particular. CT provides the best bony detail, but MRI may be needed to further identify intracranial extension. A nasal glioma typically appears as a mixed-intensity mass on CT scan that appears to be contiguous with intracranial structures. Nasal dermal sinuses can only be identified as they course through the skin and the subcutaneous soft tissue. Nasal encephaloceles are always identified as complex masses of mixed soft tissue and CSF intensity that are contiguous with intracranial structures[11].

Midline nasal masses in children may be difficult to manage where there is a connection with the central nervous system. Because of the rare incidence of nasal gliomas and dermoid cysts, many physicians are not familiar with their diagnosis and management. Respiratory abnormalities in newborns should always draw the physician's attention to their possibility. A thorough physical examination and complete neuroimaging studies are indicated, and prompt treatment should be carried out to avoid subsequent complications. Even though the understanding of these respiratory problems is in its infancy, and the problems may resolve spontaneously, pediatricians should be alert for any soft tissue lesion and consider the possibility of multiple lesions being present, even when one symptom predominates or occurs alone.

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