Nasal Dermoid Cyst and Nasal Glioma with Intracranial Extension

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BACKGROUND

The most common congenital midline nasal masses are dermoids, gliomas, and encephaloceles.1 These are rare, and in the United States they occur in approximately 1:30,000 live births.2 Gliomas and encephaloceles are of neurogenic origin, and dermoid cysts originate from ectoderm and mesoderm.

These congenital midline nasal masses are the result of a failure of normal neural crest cell embryology. The ectodermal anterior neuropore (primitive frontonasal region) forms medial to the optic recess of the sphenoid sinus in the third week of gestation, and the frontal, nasal, and ethmoid structures form in the area immediately proximal to the anterior neuropore.3 Through the eighth week of gestation, the suture line is not yet defined. The fonticulus frontalis (gap between frontal and nasal bones) fuses with the foramen cecum (region between ethmoid and frontal bones connecting with the prenasal space) in the area of the future cribiform plate, separating intracranial contents from the nose and extracranial structures. Failure of closure here can lead to encephaloceles. Gliomas can form when faulty closure of the anterior neuropore leads to the development of rests of CNS tissue extracranially. Dermoids may develop after faulty closure of the fonticulus frontalis, which allows dermal elements to invaginate between the nasal bones and cartilage. Intracranial connections can be seen with all of these. None should be biopsied until radiographic imaging is obtained to determine intracranial extension, as this could be associated with CSF leak. Intracranial connection is also associated with an increased risk of meningitis.4

Magnetic resonance imaging (MRI) is the test of choice for determining intracranial soft tissue connections associated with congenital midline nasal masses.5 In the first 6-8 months of life, the frontal, nasal, and ethmoid bones are not ossified and have similar CT imaging attenuation as brain and nasal cartilage, which may falsely appear as if there is a bony dehiscence in the frontonasal region.6 Dermoids may exhibit an intracranial connection 26-30% of the time.5,6 Gliomas are associated with an intracranial connection 15% of the time and tends to be associated with intranasal gliomas.7 Encephaloceles are always associated with a significant skull base defect.8 It is of utmost importance to determine if there is an intracranial connection, as neurosurgical consultation then becomes mandatory.

Surgical resection is the treatment of choice, to minimize risks of infection, growth, and destruction of adjacent tissues.9 If there is intracranial extension, surgical resection should proceed without delay to minimize complications; if there is no intracranial extension, surgery may be delayed for 2-5 years.3

RESULTS

Case 1

A 3 year old boy presented with a nasal mass since birth, just to the right of midline, extending from the nasal frontal suture down to the nasal tip (Figure 1). Magnetic resonance imaging (MRI) revealed a 3x1.1cm nasal mass with an intracranial component measuring 1.2cm extending through the foramen cecum (Figure 2). This was completely resected surgically through a frontal craniotomy approach combined with a midline dorsal nasal incision. Pathology revealed a nasal dermoid cyst. At 4 month follow-up, the patient was doing well with no evidence of residual disease or recurrence.

Case 2

A newborn boy presented with a 2cm mass overlying the nasal dorsum and left nasal bone (Figure 3). Although MRI at birth showed some suggestion of intracranial extension, repeat MRI at 3 months of age more clearly demonstrated extension of the mass deep to the nasal bone with a soft tissue stalk extending to the dura in the midline (Figure 4). This was completely resected surgically through a frontal craniotomy approach combined with an extranasal incision and intranasal endoscopic approach (Figure 5). Pathology revealed a nasal glioma. At 7 month follow-up, the patient was doing well with no evidence of residual disease or recurrence.

Figure 1: Nasal dermoid


Figure 2: MRI nasal dermoid

MRI showing nasal (solid arrow) and intracranial components of nasal dermoid (dashed arrow).

Figure 3: Nasal glioma

A) At birth. B) At 3 months of age.

Figure 4: MRI nasal glioma

MRI showing nasal component of glioma (solid arrow) and soft tissue stalk (dashed arrow) extending to dura.

Figure 5: Intraoperative photos nasal glioma


CONCLUSIONS

Congenital midline nasal masses are rare entities, with intracranial extension rarer still. Radiographic evaluation is invaluable in determining intracranial extension. If identified, a combined surgical approach between otorhinolaryngology and neurosurgery is essential for complete resection.

REFERENCES