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Nasofrontal dermoid cyst

The incidence of midline frontonasal dermoid cysts is one in 20,000 to one in 40,000. These lesions may have intracranial extension. This is explained by the anatomy and embryology of nasofrontal development. Skin involvement may also be extensive.

Nasofrontal dermoid sinus cysts have a unique embryological origin. A midline basal frontal dermoid associated with a dimple on the nasal surface with or without protruding hair and sebaceous discharge is the pathognomonic presentation.

Although concomitant anomalies and familial clustering have been described, most cases are spontaneous occurrences.

Unlike other dermoid cysts, a NDSC can manifest as a cyst, sinus, or fistula, and may extend intracranially.

See Nasofrontal dermoid cysts with intracranial extension

Clinical features

They usually occur as a mass in the median nasofrontal area and may have a pit.

Diagnosis

If a dermoid cyst is presenting as a midline mass of the skull, preoperative imaging with computed tomography and/or magnetic resonance imaging is necessary to evaluate for possible intracranial extension, given the altered embryologic development behind the formation of these cysts.

Differential diagnosis

Includes epidermoid cyst, encephalocele, glioma, and sinus pericranii.

Treatment

The management of suspected dermoid cysts includes complete surgical excision, which may require a combined intracranial and extracranial approach.

Incomplete excision frequently leads to recurrence.

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Case series

2015

Retrospective case series of nasofrontal dermoid cysts in 4 patients treated at a single tertiary medical center from June 1, 2010, through July 31, 2012.

The mean age at surgery was 2.5 years. The anatomical location of the nasofrontal dermoid cysts differed:

- (1) supratip extending through the upper lateral cartilages to the cartilaginous septum
- (2) upper dorsum and subcutaneous tissue
- (3) tip and supratip extending deep to the nasal bones with involvement of the anterior cranial fossae and dura
- (4) nasal tip extending deep to the level of the rhinion and involving the upper lateral cartilages and below the left medial canthus. Preoperative imaging was performed on all patients. There was one case of intracranial extension. All patients underwent surgical excision with the vertical midline incision. Nasal reconstruction was performed with local soft-tissue flaps (1 patient), regenerative tissue matrix (2 patients), and bone dust pate (1 patient). The patient with intracranial involvement also underwent a frontal craniotomy. All lesions were histologically confirmed as dermoid cysts. Mean follow-up was 1.5 years. There were no complications or recurrences. All patients had cosmetically acceptable scars ¹⁾.

Fifty-five patients were treated. Magnetic resonance imaging or computed tomography was used to delineate the anatomy, and surgical excision was expedited if there was a history of infection, especially if imaging suggested intracranial extension. Twelve patients were treated endoscopically (one was converted to open). Eleven required transcranial approaches for intracranial extension (20 percent). Of these, one lesion breached the dura. The remaining 32 patients had dermoids excised with an open approach (direct, bicoronal, or rhinoplasty). There were no recurrences in the open group and there was one recurrence in the transcranial group. This was treated by reexcision ²⁾.

2002

2 patients:

The first patient, a 33-year-old woman, sought care for chemical meningitis. As a child, she was differentiated from her identical twin sister by a dimple on the tip of her nose. The second patient, a 34-year-old man, sought care for new-onset seizures. Since birth, he had a dimple on the tip of his nose. As a child, he had undergone resection of a nasal cyst. Imaging studies in both patients indicated a midline anterior cranial base mass within the falx and a defect in the crista galli.

Both patients underwent biorbitofrontal nasal craniotomy. A bifrontal craniotomy was performed first, then removal of the orbitonasal ridge. The dermoid and involved falx were resected. The sinus tract was followed through the crista galli and resected up to the osteocartilaginous junction in the nose.

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The remainder of the tract was resected via a small incision through the nares. The dura was closed primarily by mobilizing the dura along the sides of the crista galli. After surgery, both patients still possessed their sense of smell ³⁾.

1995

A series of 19 children with congenital nasal dermoid cyst or fistulas admitted in our department between 1978 and 1992 was reviewed in a retrospective study. They were 8 females and 11 males, ages ranged from 9 months to 7 years (mean: 2 years and 4 months). At diagnosis 8 children presented with infection (superficial in 6 cases, neurological in 2 cases), and in 11 cases the malformation was discovered by the family physician. In 14 cases the cyst was located at the mid nasal ridge. Four children had associated malformations: neurodevelopmental delay (2 cases), hypertelorism (1 case) and external ear agenesia with facial palsy. The cyst was subcutaneous in 5 cases and was operated on through a nasal approach. The 14 other children were operated through associated intra and extracranial approaches. Surgical exploration disclosed a dural extension of the fistula in 9 cases. When performed, preoperative radiological explorations including CT Scan and/or MRI had not always predicted the dural extension. The mean follow-up is 5.3 years. No recurrence was recorded and the cosmetic result was always excellent. Preoperative radiological investigations are necessary to precise the location of these malformations and to detect associated disorders, but they appear unable to prove the extension of the fistula to the dura. Intracranial and extracranial approaches should be always planned to perform a total excision of the lesion ⁴).

1994

In 5 patients, ages 4 to 48 months (mean 25 months), computed tomographic scans indicated indirect signs of intracranial extension, which were confirmed intraoperatively and histologically in all 5 patients. After neurosurgical consultation, a combined single-stage intracranial-extracranial approach was used to excise the lesion. No perioperative complications occurred. Clinical assessment (follow-up 20 to 40 months, mean 31 months), confirmed by postoperative CT scan 1 year after surgery, indicated no evidence of recurrence, residual skeletal contour defects, or deformity; forehead and nasal growth were qualitatively normal, and scar appearance was satisfactory. The experience indicates that intracranial extension of nasal dermoid sinus cysts seen at a tertiary care referral center are not rare, that computed tomography scan permits accurate diagnosis, and that the single-stage intracranial-extracranial approach to resection is effective and results in minimal morbidity ⁵⁾.

Case reports

2022

An 8-year-old previously healthy female was initially found to have a small pit on her nasal dorsum. The lesion developed a local infection and she was initially treated with antibiotics two years prior to the current presentation. The lesion was diagnosed as a dermal sinus tract, and surgical removal was conducted at an outside hospital. In retrospect the pre-operative work-up imaging showed an occult intraosseous nasal bone extension, however, this was not appreciated at the initial surgery. She experienced repeat infections and underwent a second surgery with exploration under the nasal bones, however, the patient experienced recurrent postoperative local infections. The family

presented to our institution for a second opinion. Our images interestingly illustrate the nasal dermoid cyst extending into the nasal bone at the nasofrontal junction without detectable extraosseous subnasal bone extension on the imaging. The patient proceeded with a third surgery to completely remove the lesion via an extended vertical nasal incision. The nasal bones were removed in their entirety, the occult dermoid cyst with a small tract was located in the nasal bones and the undersurface of the bones was debrided entirely. No intracranial extension was observed after a careful investigation of the skull base. Particulate corticocancellous bone was used with fibrin sealant to reconstruct the defect. The nasal bones were then replaced. The pathology results were consistent with a dermoid cyst. The postoperative course was uncomplicated and she has not had a recurrence after the third surgery.

This case, despite an exploration under the nasal bones, initially received incomplete resection and experienced multiple infections because of failure to appreciate the portion hidden in the nasal bones. Our case was successfully treated with ostectomy of nasal bones without recurrence and complications. This procedure allows clear visualization of the entire cyst leading to the complete removal of the lesion. This is an instructive case to show that portions of the cyst may remain hidden and lead to recurrent infection, and complete resection with sufficient exposure of the entire lesion is needed to successfully treat this condition ⁶⁾.

2012

A 15-year-old boy presented with a fistula, present from birth, in the nasal soft triangle. This report describes a case of NDSC occurring in the nasal septum and a fistula opening in the soft triangle, which is very rare, without recurrence after 4 years ⁷⁾.

2011

Nasofrontal dermoid fistula in a child: report of a case 8).

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