Familial frontonasal dermoid cysts

Report of four cases

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A family group, comprised of father and all three children, had frontonasal dermoid cysts. Clinical features and x-ray findings are discussed, and the similarity to a slow-growing intracranial space-occupying lesion is emphasized. Plain x-ray films may show a widened nasal septum, and contrast radiology will help outline an avascular mass arising from the cribriform plate. Resection utilizing a synchronous nasal and frontal operation proved effective.

Key Words · dermoid cysts · frontonasal · familial

Intracranial dermoid cysts, although rare, are well documented.\(^6,12,15,16\) They are usually located in the posterior fossa and the spinal canal, but have also been reported in the subfrontal\(^11\) and median nasal regions. It is difficult to ascertain the true incidence of these tumors in either the frontal or nasal sites as some are not reported and some may be incorrectly diagnosed, particularly the nasal variety.

Familial nasal dermoid cysts have also been described. In all, seven families are documented, with 19 patients affected.\(^1,5,8-10,14\) No familial series of frontal dermoid cysts has been published, and in none of the nasal series is the combined frontal and nasal situation described. We wish to record a family in which the father and all three children were found to suffer from both frontal and nasal dermoid cysts. The paternal grandparents were first cousins, but otherwise no other features seemed significant in the family history. No other members of the family have had similar lesions.

Case Reports

Case 1 (Father)

This 33-year-old man was seen in 1961 with a 10-year history of epilepsy. A "nasal cyst" had been removed some years previously elsewhere; no precise information was available on the nature of the lesion. The epileptiform attacks were nocturnal with hallucinations and postictal dysphasia.

Examination. There was bilateral anosmia and early papilledema. No other abnormalities were detected, and general examination was normal. Skull x-ray films revealed a translucency in the right frontal region together with erosion of the anterior clinoids. An electroencephalogram demonstrated a right posterotemporal focus. Bilateral carotid arteriography showed a large suprasellar subfrontal mass with considerable posterior displacement of the anterior part of the third ventricle.

Operation. Right frontal craniotomy was performed on October 31, 1961. A large
dermoid cyst was encountered extending mainly to the right of the midline and posteriorly as far as the optic chiasm. A small perforation was noted in the cribriform plate. The cyst was excised, and histological examination confirmed that it was a simple dermoid.

Postoperative Course. The immediate postoperative period was uneventful, and the patient was neurologically normal apart from anosmia on discharge. Subsequently, grand mal epileptic attacks have occurred and are now controlled with anticonvulsant therapy.

Case 2 (Son)

This 13-year-old boy, the eldest sibling, was seen by the department of otolaryngology staff in 1967 because of complaints of recurrent episodes of nasal obstruction; a submucous resection of a septal swelling had previously been carried out in 1963 when deviation of the septum to the right with a high left-sided cystic swelling was noted.

Examination. X-ray films of the skull and facial bones showed a widened nasal septum, maldevelopment of the right frontal sinuses, and incomplete fusion of the frontonasal process (Fig. 1 left). A diagnosis of a midline dermoid cyst of the nose was made.

Operation. On February 20, 1968, through a transnasal approach, submucous resection of the septum was carried out, and a large high nasal cyst extending well into the anterior cranial fossa excised.

Postoperative Course. Cerebrospinal rhinorrhea and meningitis developed. The patient was treated conservatively and recovered. Five months later remnants of the cyst were removed at a second transnasal operation, and were confirmed histologically to be those of a dermoid cyst. Two years later the patient is well except for anosmia. No further episodes of rhinorrhea or meningitis have occurred, although he remains under observation.

Case 3 (Daughter)

This 12-year-old girl, the second sibling, was first seen in 1970. Earlier when she was a few months old, she had suffered a minor head injury followed by a white discharge "from the bridge of the nose" which continued intermittently throughout childhood. Her mother had noticed recent personality changes, and it was largely for this reason that the patient was referred to the hospital.

Examination. The patient was a simple girl with poor recall of recent events. There was hypertelorism with a widened nasal bridge and small dermal sinuses just below the glabella. Apart from anosmia, no neurological abnormality was detected. Widening of the nasal septum was easily visualized. Plain x-ray films and tomograms of the frontonasal region confirmed the presence of a widened nasal septum and revealed a small defect in the midline frontally, as well as a large defect in the cribriform plate (Fig. 1 right). An electroencephalogram demonstrated a bifrontal slow wave abnormality, maximal on the left. A diagnosis of frontonasal dermoid cyst was made, and a synchronous nasal and intracranial operation was advised.

Operation. Bifrontal craniotomy was performed on January 24, 1970. A midline interfrontal dermoid cyst extending through the cribriform plate was found and removed in toto; a transnasal approach was then used to excise the septal extension of the lesion.

Postoperative Course. Progress was uneventful. The personality changes and memory deficits previously noted have been resolved, and anosmia remains as the only deficit.

Case 4 (Daughter)

This 9-year-old girl, the third sibling, is presently asymptomatic, but examination reveals anosmia, widening of the nasal septum, and radiological changes similar in most respects to those of her two siblings and father. Prophylactic excision of the lesion has been advised. The parents have declined until more overt symptoms present.

Discussion

The average age of patients with intracranial dermoids is about 22 years. Spinal dermoids tend to present earlier. Nasal dermoids have most frequently been described in the first and second decades of life, but as they enlarge slowly and do not cause space-occupying symptoms of significance, a fair number have presented at an older age. There is a significant sex ratio for nasal dermoids, with a male preponderance of 2 to 1, but no such preponderance seems to occur in the intracranial lesions.
Paranasal dermoids occur in one of four sites, namely, the glabella, bridge of the nose, tip of the nose, and deep septum. The pattern of presentation depends on the site. Nasal tip cysts result in a swollen drooping nose, those at the nasal bridge in marked broadening of the nose, and deep septal cysts in nasal obstruction. Finally, as these cysts communicate with the surface of the skin by small sinuses, inflammation and frank abscess formation may occur.

The clinical features of intracranial dermoid cysts are similarly related to their site of origin and to the occurrence of dermal sinuses in some. They are slow-growing tumors, and in many instances a preoperative pathological diagnosis is not possible. Intracranial infection is a particular hazard. Radiological changes in those cysts without a dermal sinus are nonspecific. Plain skull films may show evidence of raised intracranial pressure. Enlargement of the optic foramina, intracranial calcification, and erosion of the temporal bone have been described. If a dermal sinus is present, a small skull defect may be demonstrated but this is not invariable. Contrast radiology may also show evidence of a mass, but the signs are not pathognomonic. The lesions demonstrated are avascular. A broadened nasal septum, and destruction, separation, or distortion of the nasal bones are the main radiological features of nasal dermoids. In Case 3, a defect in the cribriform region was demonstrated; the small prefrontal translucency in this case was misleading, for the anticipated sinus was not present.

The only available therapy for these lesions is surgical excision. The deep nasal dermoids are relatively inaccessible and difficult to excise. Case 2 illustrates the serious problems that can arise if the possibility of an intracranial extension is not realized preoperatively. A combined simultaneous intracranial and nasal procedure is the best approach to these extensive lesions; this was used in Case 3. A similar procedure has been recommended for Case 4 after more precise delineation of the intracranial extension is possible.
Summary
We have reported the occurrence of frontonasal dermoid cysts in a father and his three children. The diagnostic features of this syndrome have been discussed and a combined bifrontal craniotomy and transnasal excision recommended as the treatment.

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References

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