International Journal of Otorhinolaryngology and Head and Neck Surgery
Faoury M et al. Int J Otorhinolaryngol Head Neck Surg. 2019 May;5(3):785-788
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

Case Report

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20191016

Craniofacial dermoid cyst: a case report

Morad Faoury[1]*, Stefan Mitrasinovic[1], William Hellier[1], Nijaguna Mathad[2], Madanagopalan Ethunandan[3]

[1]Department of Otolaryngology Head and Neck Surgery, [2]Department of Neurosurgery, [3]Department of Oral and Maxillofacial Surgery, University Hospital Southampton, UK

Received: 20 February 2019
Accepted: 08 March 2019

***Correspondence:**
Dr. Morad Faoury,
E-mail: moradfaoury@gmail.com

ABSTRACT`

We describe a case of a craniofacial dermoid cyst in a 16-month boy treated at our tertiary referral centre. The patient presented with a soft tissue swelling in the mid forehead extending down to the glabella. Computed tomography and magnetic resonance imaging scans demonstrated a peripherally enhancing cystic lesion with a defect in the underlying frontal bone. The clinical and imaging features were suggestive of a dermoid cyst with intracranial extension. The cyst became infected pre-operatively and this episode was managed by aspiration and antibiotics. Definitive management was by excision of the extra and intra-cranial components of the lesion via a bifrontal craniotomy. The presentation, investigations and management of this lesion is discussed.

Keywords: Dermoid cyst, Nasoglabellar dermoid cyst, Extracerebral dermoid cyst

INTRODUCTION

Dermoid cysts are benign congenital 'tumours' that occur in the extradural and intradural cranial spaces. Craniofacial dermoid cysts represent about 7% of all dermoids, with common locations including the periorbital, nasal, scalp and postauricular regions.[1] They typically present during infancy as non-tender, subcutaneous masses along embryonal skin fusion lines. The definitive diagnosis is made by histopathology, however computed tomography (CT) and magnetic resonance imaging (MRI) can provide supportive evidence.

We present a case of a 16-month old boy with an extracerebral lytic lesion in the craniofacial region investigated with CT and MRI, which was surgically excised and confirmed as a pure dermoid cyst.

CASE REPORT

A 16 month old boy presented to local hospital with a soft tissue swelling in his forehead and glabella (shown in Figure 1). MRI demonstrated a well-defined lesion over the inferior aspect of the forehead in the midline (Figure 2) that passed through the frontal bones and extended into the anterior cranial fossa. Incidentally, the scan also revealed a Chiari I malformation with the cerebellar tonsils extending to the level of the posterior arch of C1, but no other abnormalities. The CT scan confirmed a defect in the frontal bone and a bifid crista gali, in addition to the soft tissue lesion. The clinical and imaging appearances were suggestive of a dermoid cyst with intracranial extension.

The cyst became infected pre-operatively and was managed by aspiration and drainage (Figure 3). The microbiology sample grew *Staphylococcus epidermidis* and the patient was started on intravenous cefotaxime and metronidazole and discharged home with oral linezolid to await his elective surgery.

Once the infection had resolved, definitive management was undertaken. A vertical elliptical incision was made and the cyst capsule dissected down to the defect in the frontal bone. The tract was traced inferiorly, in a

subcutaneous plane, down to the nasal pit, which was also excised. A coronal flap was raised to expose the frontal bone and the dissected cystic lesion was left attached to the margins of the bone defect. A bifrontal craniotomy was performed and the intracranial component of the cyst exposed. A tract was traced inferiorly between the bifid crista gali and was found to run parallel to the dura, but not entering it. The intra-cranial component was removed en bloc with the previously dissected extracranial component. The bone defects were filled with bone dust and the wound closed in layers. The patient made an uneventful recovery.

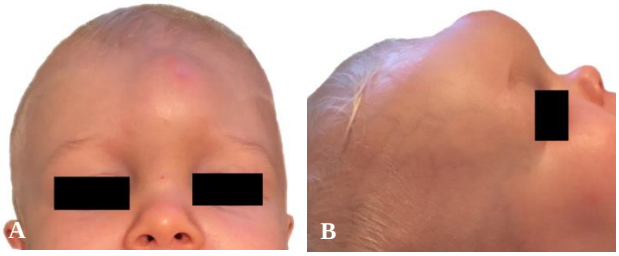

Figure 1 (A and B): 18-month-old male with a palpable mobile and painless mass on the midline frontal forehead.

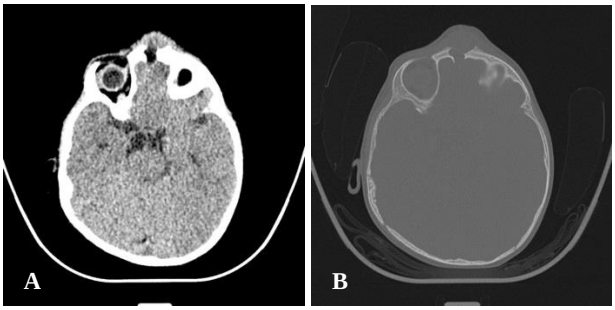

Figure 2 (A and B): Axial non-contrast CT shows fat density of the mass at the anterior aspect of the glabella with a bony defect.

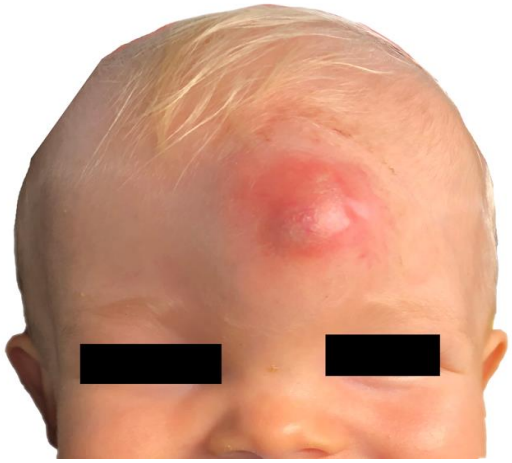

Figure 3: Infected palpable mobile mass on the midline frontal forehead.

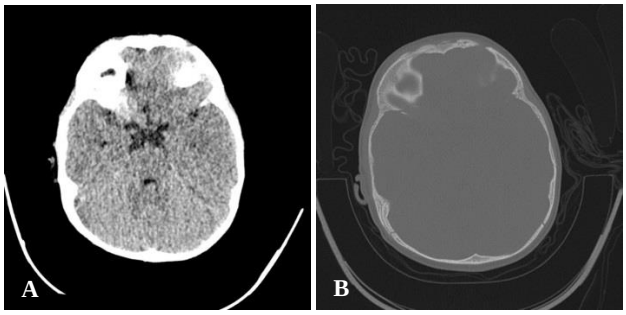

Figure 4 (A and B): Post-operative non-contrast axial CT with excision of dermoid cyst and repair of bony defect.

Histopathological examination of the excised specimen confirmed it to be a dermoid cyst. The patient had an interval post-operative CT scan (Figure 4), which demonstrated satisfactory bony infill of the frontal bone defect and no evidence of recurrence.

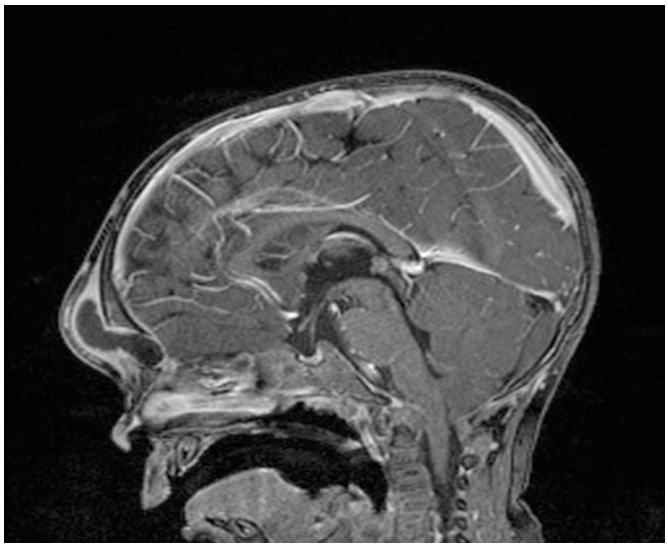

Figure 5: T1 sagittal MPR post-contrast sequence showing a well-defined cystic structure passing through the frontal bones and abutting the frontal lobes of the brain.

DISCUSSION

Dermoid cysts are rare, benign heterotopic neoplasms termed choristomas. Etiologically, they are thought to be derived from the dermal and epidermal tissues trapped in the cranial fusion lines as the neural tube closes in embryogenesis during 3-5 weeks of foetal life.[2]

They can occur in three primary locations in the head and neck: the frontotemporal region, the periorbital region and the nasoglabellar region.[6] Lesions in the nasoglabellar region are also termed nasal dermoid sinus cysts. Pryor et al found, in their review of 49 cases of paediatric dermoid cysts, that the periorbital region was most commonly involved (61%) and the frontotemporal

and nasoglabellar regions accounted for only 16% of cases.[7] However, Rahbar et al in their series of 42 cases found the most common site of presentation was the nasoglabellar region (31%).[8] Depending on the location, management of these cysts require a multidisciplinary approach including the surgical expertise of otolaryngologists, neurosurgeons, oral and maxillofacial surgeons, plastic surgeons and ophthalmologists.

The prenasal theory, based on observations by Grunwald in 1910 and refined by Pratt in 1965 is a popular explanation for the development of nasoglabellar dermoids.[3,4] As the dura recedes from the prenasal space, if it is still attached to the dermis, it may pull nasal ectoderm upwards and inwards which results in trapped cutaneous elements along the path of the diverticulum.[4,5] Proliferation of entrapped epithelium produces a dermoid, which exists as an epithelial-lined sac containing adnexal structures including glands and hair follicles. Dermoids are definitively diagnosed with histology, which shows a lining of squamous epithelium and dermal elements such as hair follicles, sebaceous and sweat glands.

Accurate pre-operative assessment is critical to evaluate these lesions, especially any intra-cranial extension. Progressive enlargement of a glabellar dermoid can cause soft tissue and skeletal deformity, local infection, meningitis and brain abscess. A meta-analysis of multiple series by Hanikeri et al found the frequency of intracranial extension for nasoglabellar dermoids to be of 19.6%.[3]

Nevertheless, all patients with a midline dermoid should be considered to have an intracranial extension until proven otherwise. Cross sectional imaging, including CT and MRI are important in identifying the extent and location of the lesion and associated tracts and bony defects.

Infections are not uncommon and should be controlled prior to undertaking definitive management. These infections may present as erythema, as noted in this case, and/or intermittent drainage. Rahbar et al has found that 24% of patients in their case series had a history of location infection, however none had a history of intracranial infection or meningitis.[8]

Surgery is the only effective treatment, with the goal of complete removal of the lesion to prevent recurrence. Which surgical strategy is employed depends on the location of the cyst and the extension of the tract and weighing these up with a focus on the best cosmetic results. Complete resection can be complicated by fibrous adhesions of the tumour capsule with adjacent neural and or vascular structures. Subtotal excision will fail to eradicate the lining of the cyst, resulting in recurrence rates ranging from 30-100%.[8] Dermoid cysts can elicit an inflammatory response and the risk of infection should be anticipated. Thus, patients should be administered antibiotics in the peri-operative phase and during surgery.

There is a difference of opinion in the literature regarding the surgical strategy when a preoperative imaging demonstrates a sinus tract with a bifid and/or widened crista galli but without an intracranial mass. Sessions et al. advocates a biopsy of the stalk of the cranial base before craniotomy, while the tract may appear to extend intracranially it is often fibrous and does not need formal excision.[9,10] Histopathology can confirm if the tract is indeed fibrous or holds epithelial content. However Posnick et al. have suggested that various epidermal and adnexal elements may appear staggered along the sinus tract at presentation, therefore a single biopsy might lead to a false negative.[11] The case presented here had a bifid crista galli and a bony defect which warranted a craniotomy, and thus full excision of the dermoid cyst along with the tract.

CONCLUSION

A rare case of a craniofacial dermoid cyst with intra-cranial extension is reported. The tumour was completely removed through a bifrontal craniotomy. Dermoid cysts may arise anywhere in the intra- and extracranial space with the majority in the frontotemporal, nasoglabellar and periorbital regions. Although they are benign tumours that expand slowly, they may cause a marked inflammatory response and destroy surrounding tissue structures, and can be a conduit for recurrent infections, particularly intracranially. Complete removal of the cyst and its tracts is required and is best managed in a multidisciplinary approach.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Demir MK, Yapıcıer O, Onat E, Toktaş ZO, Akakın A, Urgun K, et al. Rare and challenging extra-axial brain lesions: CT and MRI findings with clinico-radiological differential diagnosis and pathological correlation. Diagn Interv Radiol. 2014;20(5):448-52.
2. Yeola M, Joharapurkar SR, Bhole AM, Chawla M, Chopra S, Paliwal A. Orbital floor dermoid: an unusual presentation. Indian J Ophthalmol. 2009;57(1):51-2.
3. Hanikeri M, Waterhouse N, Kirkpatrick N, Peterson D, Macleod I. The management of midline transcranial nasal dermoid sinus cysts. Br J Plast Surg. 2005;58(8):1043-50.
4. Pratt LW. Midline cysts of the nasal dorsum: embryologic origin and treatment. Laryngoscope. 1965;75(6):968-80.
5. Hughes GB, Sharpino G, Hunt W, Tucker HM. Management of the congenital midline nasal mass: a review. Head Neck Surg. 1980;2(3):222-33.
6. Bartlett SP, Lin KY, Grossman R, Katowitz J. The surgical management of orbitofacial dermoids in the pediatric patient. Plastic Reconstructive Surg. 1993;91(7):1208-15.

7. Pryor SG, Lewis JE, Weaver AL, Orvidas LJ. Pediatric dermoid cysts of the head and neck. Otolaryngol Head Neck Surg. 2005;132(6):938-42.
8. Rahbar R, Shah P, Mulliken JB, Robson CD, Perez-Atayde AR, Proctor MR, et al. The presentation and management of nasal dermoid: a 30-year experience. Arch Otolaryngol Head Neck Surg. 2003;129(4):464-71.
9. Sessions RB. Nasal dermal sinuses--new concepts and explanations. Laryngoscope. 1982;92(29):1-28.
10. Pensler JM, Bauer BS, Naidich TP. Craniofacial dermoids. Plastic Reconstructive Surg. 1988;82(6):953-8.
11. Posnick JC, Bortoluzzi P, Armstrong DC, Drake JM. Intracranial nasal dermoid sinus cysts: computed tomographic scan findings and surgical results. Plastic Reconstructive Surg. 1994;93(4):745–6.

Cite this article as: Faoury M, Mitrasinovic S, Hellier W, Mathad N, Ethunandan M. Craniofacial dermoid cyst: a case report. Int J Otorhinolaryngol Head Neck Surg 2019;5:785-8.