Case Report

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Dermoid cyst over the left sphenoid in a child: a rare case report

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ABSTRACT

In this case report a bony swelling was noticed clinically which had a cystic presentation in CT imaging. After surgical removal it was sent for histopathological examination and was diagnosed as dermoid cyst. Dermoid cyst is rarely encountered lesions of head and neck region so most frequently misdiagnosed. Though this lesion is very rare but should be considered as a differential diagnosis while evaluation cystic lesions of head and neck region.

Keywords: Dermoid cyst, Congenital dermoid cyst, Maxillofacial region

INTRODUCTION

Dermoid cyst of the maxillofacial region usually occurs during childhood as rare solitary lesions. Dermoid and epidermoid cysts of the orbit are described as both superficial and deep formations with slow intermittent growth. Other than aesthetic effects, during their growth, dermoid and epidermoid cysts can cause disturbances in the eye motility, and optical nerve compression syndrome.^{1,2}

It usually presents as an asymptomatic soft mobile cystic mass covered by normal skin. Diagnostic imaging such as X-rays and computer tomography (CT) scan are done for identifying and diagnosing the lesion. Here we report a rare case of dermoid cyst over the left sphenoid bone in a $4\frac{1}{2}$ year old girl which was excised surgically.

CASE REPORT

A 4½ year old female child was referred to us with a hard mass over the left side of the head. The mass was seen at birth as a soft tissue mass around the left side of temple region, and presented as soft, non-tender, almond shaped, non-mobile mass with progressive growth but without any neurological findings. Overlying skin was normal and no fixity was observed. On examination it was measuring approximately $1 \text{ cm} \times 0.5 \text{ cm}$ in diameter over the left sphenoid region. Systemic examination was within normal limit. Radiograph of the skull revealed a cystic lesion involving left sphenoid bone (Figure 1).



Figure 1: Posterior-anterior view of skull.

CT scan showed a well-defined, round cystic mass measuring $1 \text{ cm} \times 0.5 \text{ cm}$ in diameter located extracranially over the left sphenoid bone (Figure 2).



Figure 2: Preoperative CT scan.

No active intervention was done initially and the case was followed up for six months. The lesion was found to be slowing growing and expansible in nature, with no other finding. Repeat CT scan revealed the dimension to be of $1.5 \text{ cm} \times 1 \text{ cm}$. FNAC was attempted but margin being bony, was inconclusive. The lesion was completely curetted out through an incision bordering the mass under general anaesthesia (Figure 3). The postoperative period was uneventful.



Figure 3: Curettage under general anesthesia.

DISCUSSION

Dermoid cysts have been classified as true dermoid cysts, epidermoid cysts and teratoid cysts. Their prevalence is 7% in head and neck patients and 1.6% within the oral cavity. There is no sex predilection. Several theories have been proposed to explain the development of dermoid cysts: they may result from entrapment of ectodermal tissue of the first and second brachial arches during fetal development; they could represent a variant form of the thyroglossal duct cyst; finally, previous surgical or accidental events could lead to traumatic implantation of epithelial cells into deeper tissues.³⁻⁵ Dermoid cysts of the head and neck usually occur during childhood and relatively rare lesions over the left sphenoid bone. Dermoid cyst is a pathologic term for a cyst lined by

squamous epithelium containing hair follicles, sebaceous, and sweat glands. Pathologically dermoid cysts, have been classified in three groups: congenital dermoid cyst of the teratoma type which is confined to the ovaries and testis, acquired implantation dermoid cyst formed by cells implanted traumatically into deeper structures and congenital dermoid inclusion cyst (CDIC) resulting from the inclusion of displaced dermal cells along the embryonic fusion line. CDIC over the sphenoid bone is an uncommon cystic lesion. It is a developmental lesion due to inclusion of dermal elements within the neuroaxis between the third and fifth week of the embryogenesis when the ectoderm folds into the neural tube. It is a soft, mobile or fixed, cystic mass covered by normal skin and does not cause discomfort, pulsation or throbbing. CDIC is described as a slow growing, non-tender, soft lump covered with intact skin. These are usually observed at birth and develop gradually through the accumulation of secretions and internal desquamation. They may be associated with a flattening or depression of the external table of the calvarium but have no intracranial extension or cutaneous involvement. The diagnosis can be made at birth, although adult cases had been also reported. Dermoid cysts are generally seen in the midline, in areas of embryonic fusion, either following sequestration of ectodermal tissue, or due to failure of separation of the ectoderm from the mesoderm during 3rd-5th weeks of gestation.^{6,7} However, their most common location in the head-neck region is around the eyes, followed by dorsum of the nose and anterior aspect of neck.8 The cystic fluid can be clear or yellow. CT scan and MRI are considered the best investigation, to confirm its extracranial position. Encephalocele, hemangioma, meningocele, lipoma, sebaceous cyst and cephalohematoma are important differential diagnosis. Dermoid cysts may increase in size and cause inflammation, thus, it is recommended that even asymptomatic cysts should be removed.9 Early excision is usually planned to prevent secondary infection. The recurrence rate after excision of an infected cyst is 20%.⁷ These cysts carry relatively higher risk of recurrence compared to lipomas and certain other benign lesions that mimic these cysts. Therefore, these cysts require thorough histopathological examination and close

follow-up due to their potential for malignant transformation. Dermoid cysts of the head and neck region are generally excised surgically, which was done in our case. The best procedure is a complete resection of the cystic mass with removal of the wall by blunt dissection of the lesion.¹⁰⁻¹²

CONCLUSION

Dermoid cyst of are uncommon lesions of head and neck region, and involvement of sphenoid and mimic as bony lesion was unusual and rare. They present complex diagnostic challenge and may be frequently misdiagnosed because of their rare and varied clinical presentation. In our case report, CT scan played a pivotal role in diagnosing, surgical planning and post-surgical follow up of the lesion. Hence, while diagnosing bony cystic lesion of cranium and maxillofacial region dermoid cyst should be kept as a differential diagnosis while evaluation.

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