

Prevalence of Dermoid Cysts in Congenital Midline Craniofacial Masses in Pediatric Population: A Case Series Study in Tertiary Care Hospital

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ABSTRACT

Background: Dermoid cyst is one of the congenital craniofacial masses, most commonly occurring in pediatric population. This study will help us to know the magnitude of dermoid cysts in midline craniofacial masses in children. More over the results of this study will be shared with other health professionals so that future research strategies may be drawn

Objective: To determine the prevalence of dermoid cysts in midline craniofacial masses in pediatric population in tertiary care hospital.

Material and Methods: This case series study was conducted at Department of Neurosurgery, Lady Reading Hospital(MTI) Peshawar, Pakistan from March 2016 to February 2019. A total of 52 patients with midline craniofacial masses were studied. After obtaining permission from hospital ethical committee and taking consent from patients included in the study, dermoid cysts in midline craniofacial masses was diagnosed on the basis of histopathology report done in our hospital laboratory. All the data like age, gender, place of residency, site of the midline craniofacial mass and dermoid cyst, its intracranial extension and its diameter (cm) were recorded in a pre designed proforma. All the data was analyzed in SPSS version 20. Mean and standard deviation was calculated for numerical variables and frequencies and percentages were calculated for qualitative variables. Chi square test for categorical variables was applied with p value of ≤ 0.05 as significant. Results were presented in the form of tables and charts.

Results: The mean and standard deviation of age was 4.15 ± 3.84 years in patients with midline congenital craniofacial masses. Out of 52 patients with midline congenital craniofacial masses, 33(63.5%) patients were from the age group 1-5 years. Out of 52 patients having midline congenital craniofacial masses, 23(44.2%) were females and 29(55.8%) were males. 33 (63.5%) out of 52 patients with midline congenital craniofacial masses resided in urban areas. Out of 52 midline congenital craniofacial masses, 16 (30.8%) were dermoid cysts on histopathological report. Out of total 52 midline congenital craniofacial masses, 5 (9.6%) were in anterior frontanale, 17 (32.7%) were in glabella, 24 (46.2%) were in nasal area, 4 (7.7%) were in vertex and 2 (3.8%) were in occiput. Dermoid cysts were statistically significant in gender, more prevalent in males.

Conclusion: Dermoid cyst is a common congenital midline craniofacial mass and is more common in males and urban population. The prevalence of dermoid cyst in congenital midline craniofacial masses is 30.8%. Dermoid cyst may present with intracranial extension and should be properly investigated before resection.

Key words: Midline craniofacial masses, dermoid cyst.

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INTRODUCTION

The midline craniofacial masses include congenital cysts, neoplasms, vascular malformations and inflammatory lesions. Dermoid cyst is one of the congenital craniofacial masses, most commonly occurring in pediatric population. It can occur in different part of the body like face, neck, brain, spine, ovary, uterus and testes. Other midline congenital craniofacial masses include gliomas and encephaloceles^{1,2}. It is estimated that about 7% of all dermoid cysts are craniofacial dermoids³. The midline craniofacial dermoid cysts may be extracranial or intracranial or both^{4,5}. Midline locations of the dermoid cysts include the anterior fontanelle, glabella, nasal area, vertex, and subocciput⁶. About 20-30% of the nasal dermoid cysts are usually associated with a sinus tract extending from the prenasal space to the foramen secum⁷.

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Dermoid cysts form as a result of abnormal embryogenesis during 4-8 weeks of gestation. They form the ectodermal sequestration in the cranial fusion lines following closure of the neural tube⁸. The diagnosis of dermoid cyst is done by imaging studies and histopathology. The histopathological examination of a dermoid cyst will show features of normal skin, its wall made of stratified squamous epithelium with dermal derivatives such as hair follicles, sebaceous glands, and, rarely, sweat glands^{9,10}.

The treatment of dermoid cyst needs a multidisciplinary team approach, consisting of neurosurgeons, otolaryngologists, neurosurgeons, plastic surgeons, neuro radiologists, and pediatric anesthesiologists. Surgery is the treatment of choice depending upon the location and approach. Extracranial or intracranial or both approaches can be done for the complete resection of the dermoid cyst depending upon its location and its extension into the brain^{11,12}.

This study will help us to know the magnitude of dermoid cysts in midline craniofacial masses in

children. More over the results of this study will be shared with other health professionals so that future research strategies may be drawn.

MATERIAL AND METHODS

This case series study was conducted at Department of Neurosurgery Lady Reading Hospital (MTI) Peshawar, Pakistan from 5th March 2016 to 5th February 2019 for duration of three year. The sample size was calculated by using WHO sample size calculator, keeping 7%³ proportions of craniofacial dermoids in all dermoid cysts, 95% confidence interval and 7 % margin error, the sample size was 52. Sampling was done by consecutive non probability sampling technique.

All the patients presenting with midline craniofacial masses with age 1 year or less to 15 years or more of both gender, were included in the study. Patients having history of known malignancy, hydrocephalus and congenital syndromes were excluded. Exclusion criteria were strictly followed to control the confounders and to exclude bias in the study results.

After obtaining permission from hospital ethical committee via ref:no 65/LRH date:02.03.2019 and taking consent from patients/attendants included in the study, dermoid cyst defined as benign cutaneous developmental anomaly that arises from the entrapment of ectodermal components along the lines of embryonic closure¹³ in the midline craniofacial masses was diagnosed on the basis of histopathological report after the resection of midline craniofacial masses in neurosurgery department by a qualified neurosurgeon with designation of assistant professor or above . Lady Reading Hospital Peshawar, Pakistan. All the patients underwent axial, sagittal, and coronal views of computed tomography to know the size and extent of the cyst before surgery. Both extra and intracranial or only extracranial approaches were done for the resection of midline craniofacial masses by our neurosurgical team having experience of more than 7 years of the operating surgeon.

Histopathology of the midline craniofacial masses was done in our hospital laboratory reported by pathologist having more than 5 years experience in the field of pathology.

All the data like age, gender, place of residency, site of the midline craniofacial mass and dermoid cyst, its intracranial extension and its diameter (cm) were recorded in a pre designed proforma.

All the data was analyzed in SPSS version 20.0. Mean and standard deviation was calculated for numerical variables and frequencies and percentages were calculated for qualitative variables. Chi square test for categorical variables was applied with p value of ≤ 0.05 as significant.

RESULTS

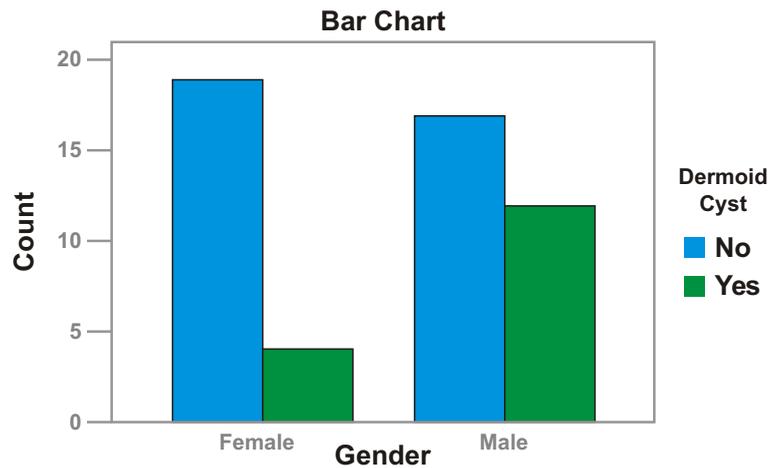
The mean and standard deviation of age was 4.15 ± 3.84 years in patients with midline congenital craniofacial masses. Out of 52 patients with midline congenital craniofacial masses, 33(63.5%) patients were from the age group 1-5 years, 9 (17.3%) were from the age group 6-10 years and 10 (19.2%) were from the age group 11-15 years. 23(44.2%) were females and 29(55.8%) were males. 33 (63.5%) patients with midline congenital craniofacial masses resided in urban areas and 19(36.5%) resided in rural areas. Out of total midline congenital craniofacial masses 5 (9.6%) were in anterior frontanale, 17 (32.7%) were in glabella, 24 (46.2%) were in nasal area, 4 (7.7%) were in vertex and 2 (3.8%) were in occiput. 16 (30.8%) were dermoid cysts on histopathological report. Whereas Out of 16 dermoid cysts, 4 (25.0) had intracranial extension on CT scan and surgery. Out of 16 dermoid cysts, 2(12.5%) had size 1-5cm (diameter), 4 (25.0) had 6-10 cm (diameter) and 10(62.5%) had >10 cm (diameter). Out of 16 patients having dermoid cysts, 10 (19.2%) patients were from age group 1-5 years, 3 (5.8%) were from age group 6-10 years and 3 (5.8%) were from age group 11-15 years. Dermoid cysts distribution in patients with midline congenital craniofacial masses among different age groups was statistically insignificant using Chi Square test with p valve = 0.05.

Table 1: Presence of dermoid cyst in relation to age group of the patient

	Dermoid cyst Absent	Dermoid cyst Present	P value
Age group 1-5 years	23(44.2%)	10(19.2%)	.983
Age group 6-10 years	6(11.5%)	3(5.8%)	
Age group 11-15 years	7(13.5%)	3(5.8%)	

Among 23 female patients 4 patients had dermoid cysts making 7.7% of all the patients having dermoid cyst. Among 29 male patients 12 had dermoid cyst making 23.1% of all the patients

having dermoid cyst. Patients having dermoid cysts were predominantly males. Gender wise dermoid cyst distribution was statistically significant.



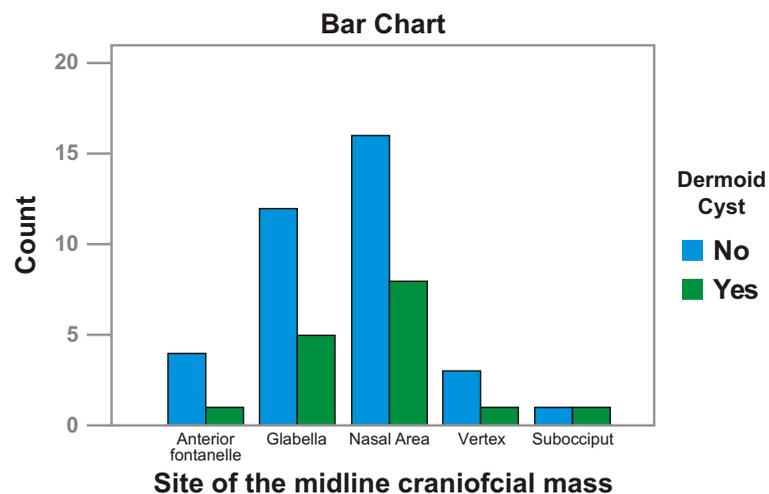
Graph 1: Midline craniofacial masses based on their type and gender

Out of 33 patients from urban areas, 11(21.2%) patients had dermoid cysts and out of 19 patients from rural areas, 5 (9.6%) had dermoid cyst. Statistically the results were insignificant. Prevalence of dermoid cyst in anterior frontanelle,

glabella, nasal area, vertex and occiput was 1(1.9%), 5(9.6%), 8(15.4%), 1(1.9%) and 1(1.9%) respectively. Statistically the results were insignificant using Chi Square test with p value = 0.05.

Table 2: Presence of midline craniofacial masses in relation to location of pathology

	Dermoid cyst Absent	Dermoid cyst Present	P value
Anterior fontanelle	4(7.7%)	1(1.9%)	.942
Glabella	12(23.1%)	5(9.6%)	
Nasal area	16(30.8%)	8(15.4%)	
Vertex	3(5.8%)	1(1.9%)	
Subocciput	1(1.9%)	1(1.9%)	



Graph 2: Number of midline craniofacial masses and their location

DISCUSSION

Congenital midline craniofacial masses are very rare lesions of head and neck including dermoid cysts, gliomas, and encephaloceles occurring in about 1 in 20,000 to 1 in 40,000 births¹⁴⁻¹⁶. Dermoid cysts are formed during abnormal embryogenesis of the. Dermoid cysts are thought to occur as a developmental anomaly in which embryonic ectoderm is trapped in the closing neural tube between the fifth and sixth weeks of gestation. They are lined by stratified squamous epithelium. Unlike epidermoid cysts, however, they contain epidermal appendages as well, such as hair follicles, sweat glands and sebaceous glands. The latter secrete the sebum that gives the characteristic appearance of these lesions on CT and MRI. In head and neck region they mostly occur in the periorbital, nasal midline and neck regions^{17,18} varying data regarding intracranial dermoid cysts is available but most commonly they occur in frontonasal area, sellar suprasellar, parasellar and posterior fossa including the vermis. Clinically dermoid cysts may be asymptomatic and found incidentally or become symptomatic over the course of time. Either because of mass effect on adjacent structures or if they rupture and cause chemical meningitis, both ways presentation can vary from simple headache, seizures to neurological deficit or at times death. When symptomatic they are treated surgically via excision but at times adhesion to surrounding structures can prevent total excision and can lead to recurrence. This study has focused on the congenital dermoid cyst in the midline in the craniofacial masses in children.

The prevalence of dermoid cyst was more common in males in our study and was statistically significant and Dutta M et al in their study also showed that males are predominantly suffered from dermoid cysts as compared to the females¹⁹. Some studies suggest that there is no significant relation between location, gender, histopathology or age of patients²⁰ but other studies have shown some significance.

The prevalence of dermoid cyst in congenital midline craniofacial masses in the present study was 30.8%, most commonly involving glabella and nasal area followed by areas such as anterior fontanelle vertex and subocciput. Rahbar et al²¹ and Hanikeri et al¹⁴ showed in their study that the prevalence of dermoid cyst in the nasoglabella area was more common i.e. 31% and 19.6% respectively. According to a study by Armon N et

al²², the prevalence of dermoid cyst in cystic lesions of head in children was 58.9%.

Dermoid cyst may be associated with cranial and spinal dysraphism, they have potential to grow over period of time and therefore intracranial involvement may be seen which needs multidisciplinary approaches for its surgery. Proper imaging is required prior to any intervention, MRI is preferred means to find intracranial extension whereas in some cases CT may be required to identify bony erosions prior to surgery^{13,23}. Some studies suggest that up to 45% nasal dermoid cysts may have intracranial extension¹³. In the present study 4 out of total 16 dermoid cysts had intracranial extension making 25% of all the dermoid cysts. A study by Pensler JM et al²⁴ showed that 19% of the dermoid cysts had intracranial extension. Dermoid cyst may present with intracranial extension and should be properly investigated before resection.

LIMITATIONS

Congenital midline craniofacial masses including gliomas and encephaloceles other than dermoid cysts, were not studied. Periorbital dermoid cyst which is very common in children were not studied.

CONCLUSION

Dermoid cyst is a common congenital midline craniofacial mass and is common in males and urban population. The prevalence of dermoid cyst in congenital midline craniofacial masses is 30.8%. Dermoid cyst may present with intracranial extension and should be properly investigated before resection.

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DATA SHARING STATEMENT: The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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AUTHOR'S CONTRIBUTION

The following authors full fill authorship criteria as per ICMJE guidelines;

Shahjehan A: Idea conception, drafting the work, final approval, agreed to be accountable for all the work.

Ullah S: Design of the work, data acquisition, critical revision, final approval, agreed to be accountable for all the work.

Haqqani U: Data analysis, Data interpretation, drafting of the work, final approval, agreed to be accountable for all the work.