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Case Report

Pediatric subcutaneous nasal glial heterotopia

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ABSTRACT

Background: Nasal glial heterotopias (NGHs) are benign lesions diagnosed at birth that are treated with complete surgical excision and have a low recurrence rate. The impact of the timing of resection on the patients' outcome remains unclear.

Case Description: We report a case of pediatric midline subcutaneous extranasal glial heterotopia over the nasal bridge in a 4-day-old female newborn. At the age of 6 months, she underwent a complete surgical excision. Follow-up magnetic resonance imaging at 3 years showed no evidence of recurrence. A summary of the 19 published cases of the specific entity of purely subcutaneous extranasal glial heterotopia among the pediatrics age group in the literature is presented, and the timing of surgery in relation to outcome is discussed.

Conclusion: Our review revealed that surgery for NGH can be safely performed when the child is 6–12 months old, and the child should be followed probably until school age.

Keywords: Nasal cerebral heterotopia, Nasal glial heterotopia, Nasal glioma, Neuroglial heterotopia

INTRODUCTION

Nasal glial heterotopia (nasal glioma) is a rare cause of congenital midline nasal masses that were first described in 1852 (Rouev *et al*., 2001).[19] It consists of the presence of mature astrocytes in an abnormal location.[11] Nasal gliomas are benign lesions that are treated with complete surgical excision and have a low recurrence rate.[16] It comprises 5% of all nasal masses with an estimated incidence of 1 in 20,000–40,000 live births (Rahbar *et al*., 2003).[18] It is extranasal in 60%, intranasal in 30%, and combined in 10% (Patterson *et al*., 1986).[16] Extranasal glial heterotopia can be located anywhere from the glabella down to the nasal tip.[1] Extremely rare locations such as the orbit, nasopharynx, hard palate, palatine tonsils, paranasal sinuses, and pterygopalatine fossa have also been reported (Amanullah *et al*., 1996;[1] Mohanty *et al*., 2003;[14] Bajaj *et al*., 2005).[2] Nasal gliomas are typically isolated lesions, and syndromic associations are exceedingly rare. Reported associations were metopic craniosynostosis (Boyer *et al*., 2015),[3] strabismus (Irkoren *et al*., 2015),[12] and cleft palate (Chandna *et al*., 2018).[4] To our knowledge, isolated subcutaneous extranasal glioma is rare, with only 19 reported cases in the English literature. Herein, we report the 20th case of pediatric isolated subcutaneous extranasal glioma; whether the timing of surgical resection affects the recurrence rate is discussed.

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CASE REPORT

A 4-day-old female newborn was noted to have a midline mass over the nasal bridge at birth. She was born at term through a cesarean section to a hypothyroid mother with an uncomplicated perinatal course. Her birth weight was 3.180 kg, and APGAR scores were 9 and 9 at the 1st and 5th min. On physical examination, there was an extranasal mass over the nasion protruding more toward the left side. It was measuring 1.5 by 2 cm, firm in consistency, and had a small purplish hue on its surface but no telangiectatic vessels [Figure 1]. It was non-pulsatile and non-expandable, with crying or straining. There was no associated intranasal mass. Magnetic resonance imaging (MRI) showed an extranasal lesion with signal intensity similar to brain tissue in T1-weighted images, while T2-weighted images revealed no fibrous stalk or intracranial communication [Figure 2]. Echocardiography and abdominopelvic ultrasonography were performed to screen for any associated congenital anomalies, and both were unremarkable. She had a normal physical and neurological development. The infant was followed up in the clinic until she became 3 years old. The ophthalmological evaluation revealed no strabismus.

At the age of 6 months, the mass had not changed in size. It was excised externally in one piece through a vertical incision, and the defect was closed primarily. There was no fibrous stalk or bony defect identified intraoperatively. The perioperative course was uneventful. Grossly, it was a single brownish rubbery mass; in the cut section, it was non-lobulated grayish-whitish in color. Microscopic examination revealed alternating dense collagenous tissue, disorganized fibrillary glial tissue, and mature astrocytes [Figures 3a and b]. Immunohistochemistry was positive for glial fibrillary acidic protein in the glial tissue and showed weak patchy staining for P53 [Figure 3c].

On 3 3-year follow-up, the wound had healed completely with an adequate nasal contour [Figure 4] and a slightly hypertrophic scar. There was no recurrence reported on follow-up MRI [Figure 5].

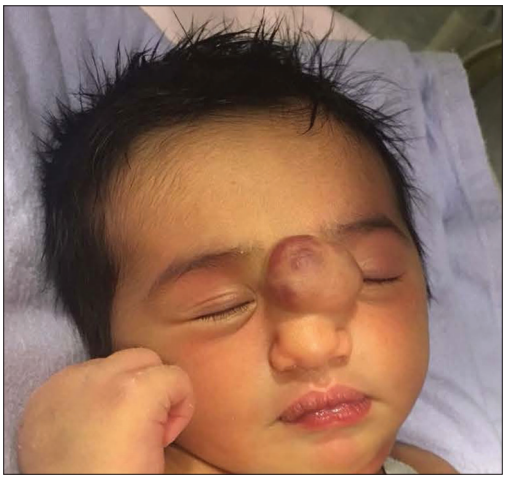

Figure 1: Extranasal mass over the nasion protruding more toward the left side. It was measuring 1.5 by 2 cm, firm in consistency, and had a small purplish hue on its surface but no telangiectatic vessels.

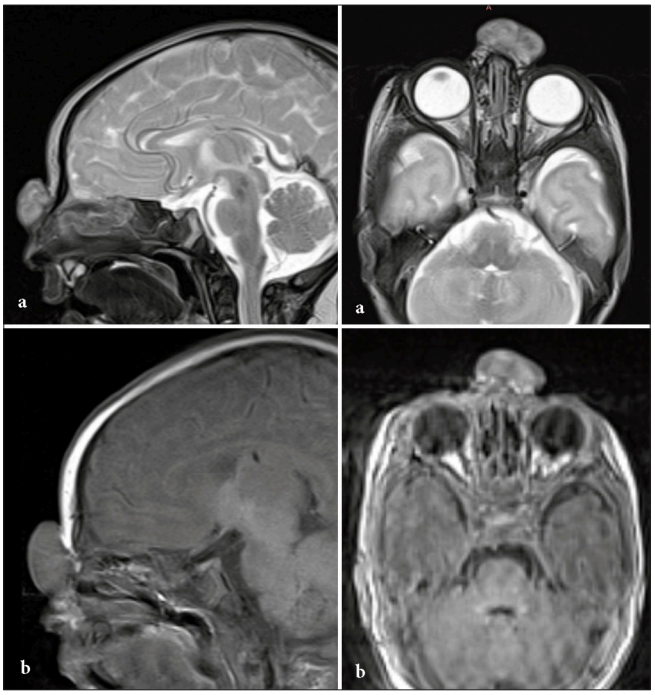

Figure 2: Magnetic resonance imaging showing extranasal lesion. (a) T2 axial and saggital images showing lesion isointense signal. (b) T1 axial and saggital images showing lesion isointense signal.

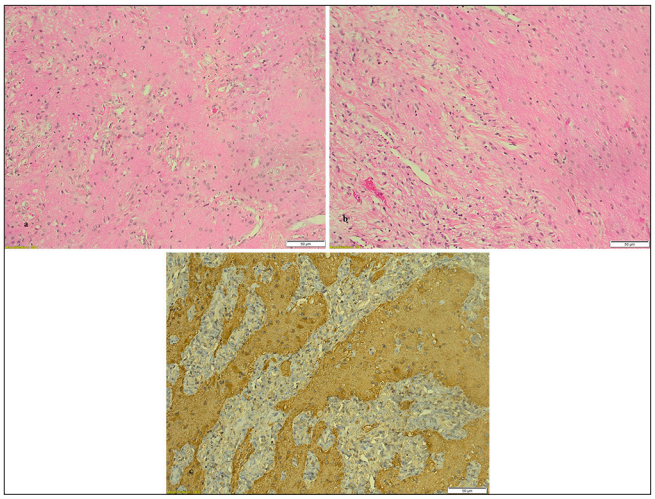

Figure 3: (a and b) Microscopic examination of hematoxylin and eosin (H&E x20) stain sections revealing fragments of alternating dense collagenous tissue and disorganized fibrillary glial tissue and mature astrocytes. (c) Immunohistochemistry (x20) study showing positive glial fibrillary acidic protein in the glial components of the lesion.

Figure 4: Postoperative picture taken at 3 years of age.

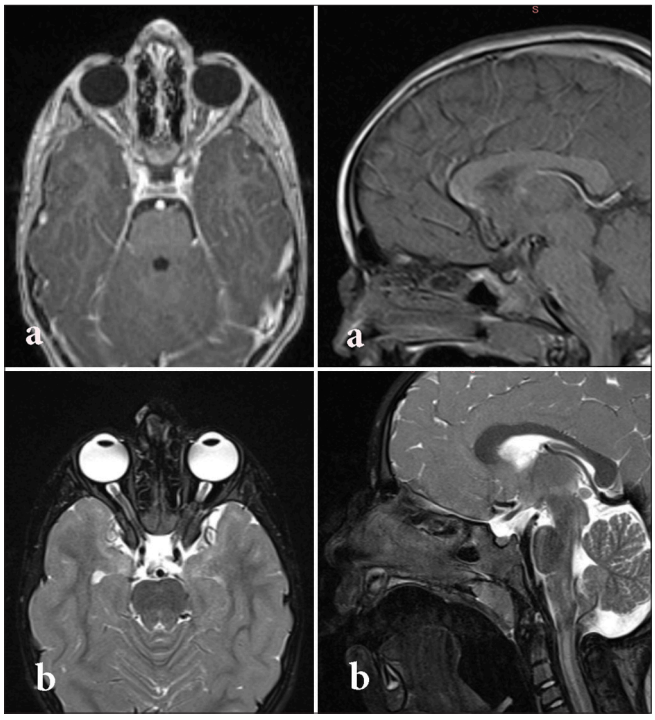

Figure 5: Postoperative magnetic resonance imaging showing no evidence of recurrunce at 3 years of age with mildy hypertropic scar. (a) T1 axial and saggital images. (b) T2 axial and saggital images.

DISCUSSION

Nasal glioma, although is a rare condition, its clinical significance lies in the potential for intracranial connection. Around 10–25% of nasal gliomas have a fibrous stalk extending to the nasal bone and down to the base of the skull (Patterson *et al*., 1986; Chau *et al*., 2005; Gallego Compte *et al*., 2022).[11,5,7] Intracranial connection is more common with intranasal lesions (Patterson, Kapur *et al*. 1986). The main differential diagnosis of nasal glioma is encephalocele. In fact, it is more accepted that nasal glioma and encephalocele lie in a spectrum rather than separate entities. Other differential diagnosis includes dermoid cyst, hemangioma, and teratoma. Nasal glial heterotopia is a rare, nonhereditary, benign congenital anomaly. Since first described by Reid in 1852, at least, 294 cases have been reported. A recently published systematic review by Compte et.al reported a review of cases of nasal glial heterotopia or the so-called "nasal glioma" in both children and adults published in the literature, with a total of 152 retrievable cases described in original publications (Gallego Compte *et al*., 2022).[9] Our review describes the specific entity of purely subcutaneous extranasal glial heterotopia among the pediatrics age group [Table 1].

The pathophysiology, clinical presentation, and surgical options for nasal glioma have been enormously discussed in the literature. However, the preferred timing of surgical excision, especially in extranasal glioma, has never been addressed. Few data exist to support or go against early versus late resection of nasal glioma and whether the timing of surgery affects long-term outcomes and recurrence rate. Nasal glioma is mostly diagnosed soon after birth, and the goal of surgery in the absence of intracranial connection or cerebrospinal fluid leakage is mainly cosmetic. The decision on when to operate was inconsistent in the literature. Some surgeons preferred to wait until the infant turns 6–12 months of age to avoid the global surgical risk in newborns (Schauer *et al*., 2018),[21] while others operated as early as the 11th day of life (Tatar *et al*., 2016).[24] A retrospective study by Rahbar *et al*. (Rahbar *et al*., 2003)[18] reviewed ten patients with nasal glioma from 1970 to 2002, and identified 2/10 recurrent cases in which primary surgical excision was performed at 2 months of age in one case and at 6 months in the other. The recurrence happened at 10 months and 2.5 years after surgery, respectively. None of the reviewed cases developed an intracranial infection with a median follow-up of 2 years postoperatively. A recently published case in 2016 reported a case of an extranasal glioma confined to the right alar wing that had an incomplete excision for cosmesis at the age of 3 months, and recurrence was evident 6–9 months after surgery (Harttrampf *et al*., 2016).[11] This goes in line with a review by Peter Lamesch who studied 166 published cases during 1890–1987 and found 18 documented recurrences among 166 cases (11.5%) mainly due to incomplete surgical excision.

It was hypothesized that dermal involvement could be associated with recurrence in extranasal gliomas and total excision of the skin overlying the mass if the skin is adherent and prevents recurrence (Thomson *et al*., 1995).[25] In our review of all published cases of extranasal gliomas from 1949 until the present case [Table 1], we found that

Table 1: Table summarizing the previously reported 19 cases of pediatric subcutaneous nasal glioma.

Author and year	Country	Age at diagnosis	Gender	Site of the lesion	Clinical associations	Intracranial connection or skull base defect in MRI/CT	Pathology findings	Age at surgical intervention, approach	Recurrence/ follow up
Yan Y.,2020[28] (Yan YY, Zhou ZY, Bi J, *et al*.2020)	China	At birth	F	nasal dorsum	Not known	No	fibrous muscle tissue and had a rich blood supply, In addition, glial tissue was found within the tumor	6 months, Extranasally	No/ 2yr
Gan W., 2018[10] (Gan W, Xiang Y, Tang Y, *et al* 2018)	United states	At birth	F	Right nasal dorsum	No	No	prominent astrocytes and oligodendrocytes with occasional neurons intermixed with a fibrous connective tissue stroma, GFAP was strongly positive	17 mo, externally	No/6 mo
Charles NC., 2018[5] (Charles NC, Lisman RD, Patel P, Callahan AB 2018)	United states	2 y	F	Superomedial aspect of the right eye	Nasal encephalocele that was surgically repaired right after birth	No intracranial connection but CT showed a small defect in the inferior portion of the left frontal bone above the lesion	Astrocytes making glial islets enmeshed in eosinophilic collagenous tissue, exhibiting an architecture that resembles optic nerve. GFAP, S-100, AE1/ AE3 (cytokeratin) and vimentin positive. It was negative for CD68, CD163, (histiocytic markers) and smooth muscle actin. Ki67 showed a very low proliferative rate	2 y, externally	N/A
Chandna S., 2018[4] (Chandna S, Mehta MA, Kulkarni AK 2018)	India	At birth	M	Midline of nasal dorsum	Type II cleft palate extending up to incisive foramen	No	Mature neuroglial tissue and leptomeningeal membrane with dilated and congested vascular channels	25 days, externally through an elliptical incision	N/A
Schauer, An., 2018 (Schauer A, Harvey NT, Vijayasekaran S, Wood BA 2018)	Australia	At birth		Midline of nasal dorsum	No	No	central core of fibrous and fatty connective tissue containing both glial and meningothelial elements formed in whorled arrangements with associated psammomatous calcification. GFAP positive and epithelial membrane antigen positive	6 months, externally	No/
Tatar., 2016 (Tatar EC, Yıldırım GA, Keseroğlu K, Özdek A, Saylam G, Korkmaz MH, Polat R 2016)	Turkey	At birth	F	Right nasal dorsum	No	No	Glial tissue GFAP positive	11 days, externally through a subciliary incision	No/18 mo
Harttrampf AC., 2016 (Harttrampf AC, Schupp W, Timme S, Niemeyer CM, Otten JE, Rossler J 2016)	Germany	#1 At birth #2 at birth	F	#1 Left nasal dorsum #2 right nasal dorsum	#1 No #2 No	No	Glial tissue involving the dermis. Stained strongly positive for GFAP and S-100	5 w, externally	#1 No/6mo #2 yes/6-9 mo
Irkoren S., 2015 (Irkoren S, Selman Ozkan H, Karaca H 2015)	Turkey	At birth	F	Right nasal dorsum and inferomedial orbital border	Strabismus	No	Nonmalignant gliomatous cells with low proliferative activity. No meningeal or dural tissue seen	12 mo, externally	No/6mo
Hye, R., 2015[15] (On HR, Seo J, Chung KY 2015)	Korea	1 y	M	upper part of the right side of the nose	NA	No	prominent dermal and subcutaneous neural proliferation composed of astrocytes, neurons, and neuroglial fibers intermixed with a fibrous, connective tissue stroma	1 y	NA
Locke R., 2011[13] (Locke R, Kubba H 2011)	United Kingdom	1y	F	Nasal dorsum	No	No	Glioma	1y/externally	No/3mo
Vilarinho C., 2009[26] (Vilarinho C, Ventura F, Vieira AP, Bastos MJ, Teixeira M, Brito C 2009)	Portugal	At birth	F	Left nasal dorsum	No	No	Skin, overlying glial tissue positive for glial fibrillary acid protein,(GFAP) and enlarged neurons positive for synaptophysin. (Fig. 4), consistent with neuroglial heterotopia.	18 mo	No/ 2.5 yr
Sharma JK., 2006[22] (Sharma JK, Pippal SK, Sethi Y, Arora S, Raghuwanshi SK 2006)	India	At birth	F	Left nasal dorsum	No	No	Nasal glioma	11mo/externally	No/6mo
Cheung D., 2005 (Cheung, D., Woodruff, G., Brown, L. *et al* 2005)	United Kingdom	At birth	F	Right nasal dorsum	No	No	Nasal glioma	there was attachment via a thin stalk to a small depression on the anterior surface of the frontal process of the maxilla	N/A
Taege C., 2001[23] (Taege C, Musil A, Klohs G, Rath FW 2001)	Germany	At birth	F	Right upper lip	No	No	Fibromyxoid connective tissue with a central area of disorganized cerebral tissue lacking surrounding meningeal tissue or any sort of capsule	1mo/externally	N/A
Hoeger PH., 2001 (Hoeger PH, Schaefer H, Ussmueller J, Helmke K 2001)	Gemany	At birth	F	Midline nasal dorsum	No	No	Non-malignant gliomatos cells GFAP and S-100 positive	7mo/externally	N/A
Sanjuán Rodríguez S., 1998[20] (Sanjuán Rodríguez S, Díaz Pino P, Ortiz Barquero MC, Fernández Portales I, Cabezudo Artero JM 1998)	Spain	prenatally		Nasion	No	CT/RI were not performed preoperatively	Nasal glioma	Newborn/externally	N/A
Pensler JM., 1996[17] (Pensler JM, Ivescu AS, Ciletti SJ, Yokoo KM, Byrd SE 1996)	United states	10 patients were reported, 6 are extranasal : mostly diagnosed at birth	3 M/3 F	Ranges from glabella to nasal dorsum	No	1/6 had intracranial extension	Variable proportions of glial and fibrous tissue	3mo- 12y/ externally	No/8.9±3.7y
Fletcher CD., 1986[8] (Fletcher CD, Carpenter G, McKee PH 1986)		3 cases: diagnosed at birth		Over the nasion	No data	No data	No data	No data	No data
Whitaker SR., 1981[27] (Whitaker S. R., Sprinkle P. M., Chou S. M 1981)		At birth	F	Nasion	No	No	fibrocollagenous septae separating interspersed glial cell islets and scattered hypoplastic skeletal muscle cells	11mo/externally	N/A

despite the variability of the timing of surgery, there was no difference in outcome with regard to local tissue destruction or intracranial infection. The mass had remained the same size and shape until the time of excision, suggesting a very slow, if any, growth rate. The presence of a fibrous stalk extending the skull base was reported (Cheung et al., 2005),[4] but none of the reviewed cases developed cerebrospinal fluid leakage or meningitis before or after surgery during the follow-up period (Cheung, Woodruff *et al*. 2005). Moreover, none of the cases had a local tissue destruction or invasion. Recurrence was reported in three cases due to incomplete excision. However, it is worth noting that most of the cases had a short follow-up duration and given the extremely slow growth rate of nasal glioma, longer follow-ups are needed.

We believe that in surgery for extranasal glioma, the approach should provide adequate exposure for complete excision, allow for exploration of a fibrous stalk or a bony defect or intracranial communication, and provide a good cosmetic result. It can be safely performed when the child is 6–12-month-old and the child should be followed probably until school age.

CONCLUSION

Our review revealed that the timing of surgery in extranasal glioma does not make a difference in outcome with regard to local tissue destruction, infection risk, or recurrence. The recurrence was mostly due to incomplete resection. We believe that in surgery for extranasal glioma, the approach should provide adequate exposure for complete excision, allow for exploration of a fibrous stalk or a bony defect or intracranial communication, and provide a good cosmetic result. The clinical course of nasal glioma is static, which gives more flexibility in choosing the timing of surgery. It can be safely performed when the child is 6–12 months old. Recurrence was reported up to 2.5 years after excision so the authors suggest that the child should be followed probably until the school age.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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