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Case Report *Volume 5 Issue 1*

A Rare Presentation of Spindle Cell Tumour Presenting as an External Nasal Mass: A Case Report

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Abstract

Congenital nasal lesions (such as nasal dermoid, encephalocele, and gliomas) are rare and result from abnormal embryonic development. It is necessary to distinguish them and understand their full pathology to avoid unnecessary complications. Nasal dermoid cyst-like swelling is a rare ectodermal and mesodermal origin condition, with a connecting tube or tail extending intra-cranially in only 10% of congenital diseases. The intracranial component of the mass predisposes children to meningitis and abscess formation. Another swelling, such as craniofacial neurofibromatosis, is a benign but devastating disease often accompanied by significant mid-face deformity. Recurrence can be seen if inadequately removed, so all patients' long-term follow-up should be done.Here we present a case of midline nasal swelling in a 1-year-old child and share our experience about the successful management of midline nasal mass.

Keywords: Congenital Midline Mass; Nasal Dermoid Cysts; Neurofibromatosis

Abbreviations

CEMRI: Contrast-Enhanced Magnetic Resonance Imaging; NF: Neurofibromatosis.

Introduction

There are many types of nasal midline swellings, the most common is the nasal dermoid cyst, which accounts for 1% to 3% of all dermoid cysts and 4% to 12% of head and neck dermoid cysts. It may appear as a cystic mass or sinus opening on the midline nasal dorsum at birth, or during early childhood [1]. The embryological origin of nasal dermoid cysts is different from dermoid cysts elsewhere on the face and requires special treatment care. Histologically, dermoid cysts or sinuses are composed of a fibrous capsule of squamous epithelium and contain adnexal structures as hair follicles, sebaceous glands and sweat glands [2]. The differential diagnosis needs to be carefully considered during assessment and includes ectopic neuroglia, encephaloceles, and teratomas [3]. All connecting stalk must be removed to prevent cerebrospinal fluid leakage and reduce the likelihood of recurrence.

The roles of CT and MRI appear well established in congenital midline nasal masses so accordingly surgeon will plan for the requirement of craniotomy and rhinoplasty [4]. Proper clinical history and radiological investigations are important before planning the surgical management [5]. The face, especially the nose and lips, is another area of the face that

is frequently affects in craniofacial neurofibromatosis and overall impact on aesthetic results of these patients [6]. Neurofibromatosis (NF) is a neuro-cutaneous syndrome characterized by the development of tumours of the central or peripheral nervous system including the brain, spinal cord, organs, skin, and bones [7]. Here we are presenting a case of midline nasal swelling in which we completely removed the midline nasal mass.

Case Report

A 1-year-old male child was reported in the OPD with the chief complaint of swelling over the dorsum of the nose since birth. It was insidious in onset with slowly increasing in size. There was no history of nasal discharge, nasal bleeding, mouth breathing, fever, trauma, or any discharge from the swelling. The child was fully vaccinated till age and had no developmental delay. On local examination of the nose, there was an external swelling of around 2 x 2 cm over the dorsum of the nose extending from the glabella to the midpoint of the dorsum of the nose and laterally up to the naso-facial groove. There was no previous scar mark or any blood/pus discharge. On palpation, the swelling was firm in consistency, non-tender, mobile, non-fluctuating, non-compressible and non-reducible. On anterior rhinoscopy, there was a central septum with congested nasal mucosa bilaterally. The rest of the ear and throat examination was normal and no abnormality was detected. Earlier the patient was investigated at some other hospital and was referred here due to the non-availability of dates for early operation.

CT brain (at the previous center) suggested a well-defined soft tissue density lesion involving the subcutaneous plane anterior to the nasal cavity predominantly in the midline. There was no evidence of intracystic calcification or communication with bone. The suggested diagnoses were sebaceous cyst/dermoid cyst or fibroma (Figure 1).

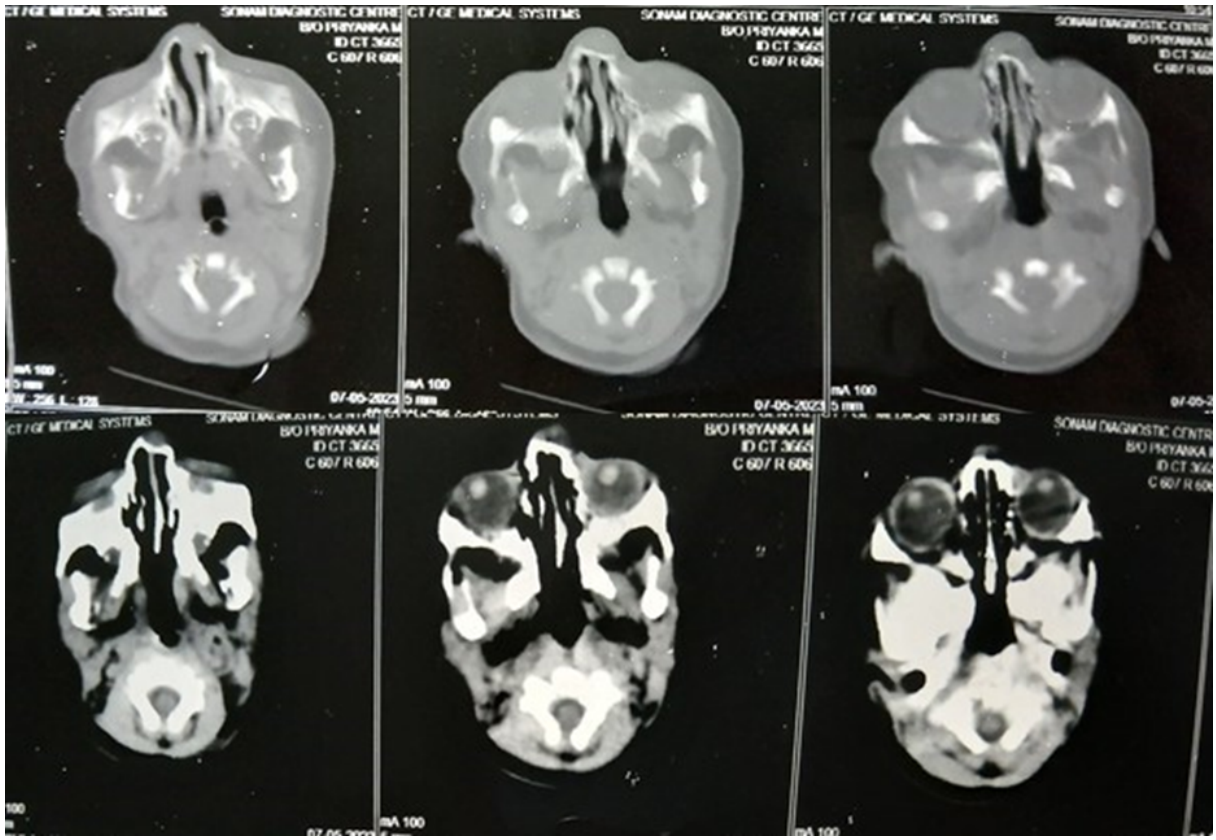

Figure 1: CT brain and nose revealed a well-defined soft tissue density lesion 6*10mm involving the subcutaneous plane anterior to the nasal cavity.

CEMRI brain (at previous center) revealed a well-defined hyperintense lesion (1.5*2*2.4cm) in the subcutaneous plane anterior to the nasal cavity predominantly in the midline and slightly towards the left side. No significant neuro-parenchymal abnormality was detected and the most likely diagnosis suggested there was a dermoid cyst (Figure 2). Therefore surgical excision was planned according to this diagnosis at the previous center.

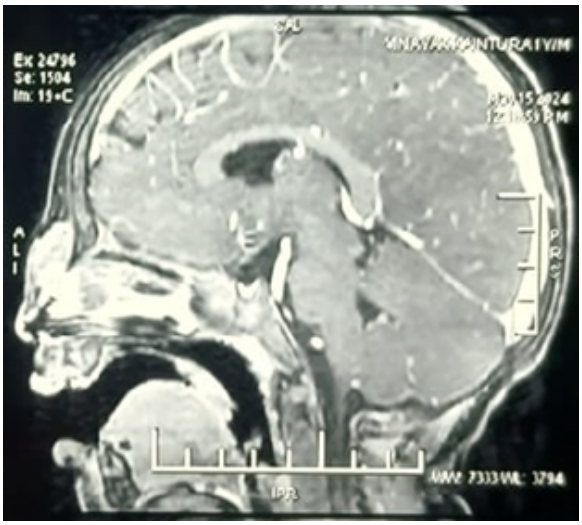

Figure 2: CEMRI brain [a (axial) & b (sagittal) cuts] showing a well-defined hyperintense lesion in the subcutaneous plane anterior to the nasal cavity.

However, at our center, after taking the radiologist's opinion sonography was done as the mass was hyperintense on MRI. USG soft tissue revealed a 2.25*1.10*1.54 cm heterogeneously hypoechoic soft tissue lesion along the root of the nose with internal vascularity suggestive of nasal glioma (nasal glial heterotopia) (Figure 3).

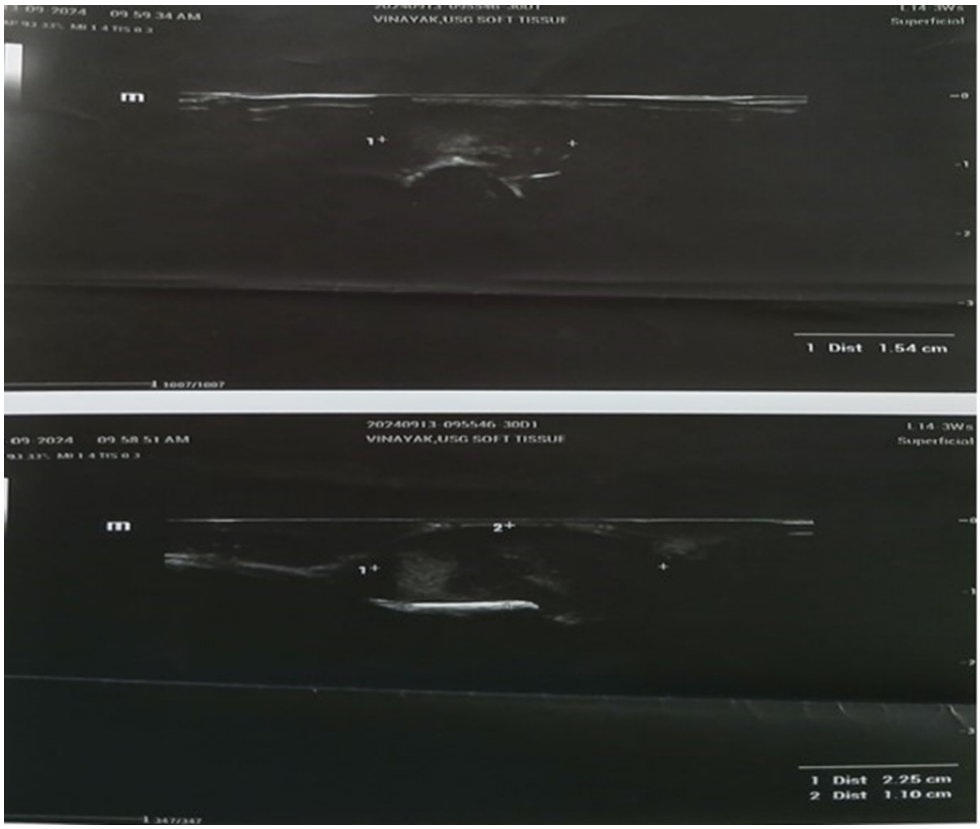

Figure 3: USG revealed a soft tissue lesion along the root of the nose with internal vascularity, possibly a nasal glioma.

Fine needle aspiration cytology revealed a paucicellular hemorrhage smear with few clusters of epithelial cells in a proteinaceous background and no evidence of malignancy. Based on the above findings surgical excision of the nasal cyst was planned under general anaesthesia. 2% local anaesthesia with adrenaline was infiltrated around the swelling. A horizontal incision was given over the cyst. Dissection started and the cyst was visualized and further dissection was done around the cyst. Cyst was found adhered to the left side upper lateral cartilage of the nose and left side of septum. Cyst freed from all adhesions- removed in toto and sent for histopathology. Merocel was put in the left nasal cavity. Suturing was done in 2 layers (Figure 4).Sutures were removed on the 8th postoperative day (Figure 5). On histological analysis, it showed a spindle cell tumour, possibly neurofibroma (Figure 6).

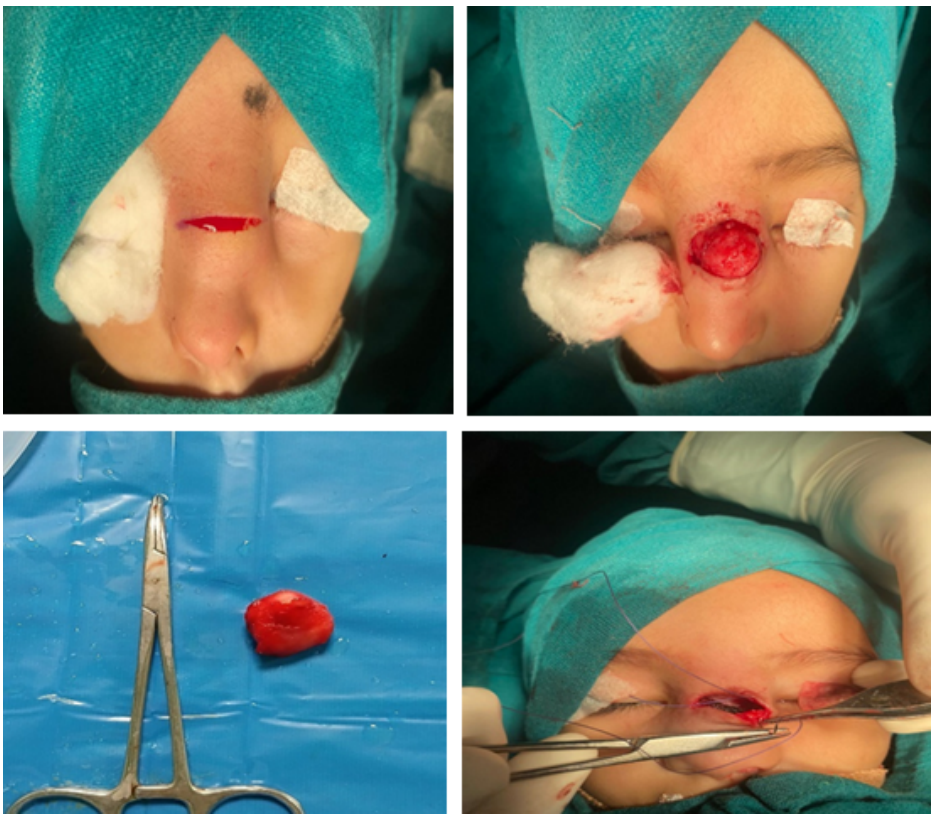

Figure 4: Intraoperative image showing **4A:** Horizontal incision **4B:** Dissection around cyst **4C:** Excised nasal cyst **4D:** Suturing in 2 layers.

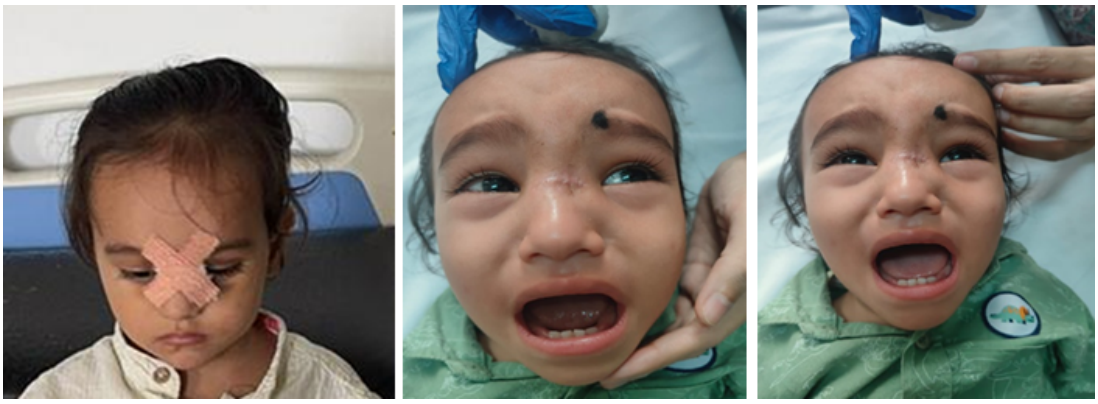

Figure 5: Photograph on follow-up of a case, on 8th postoperative day after suture removal.

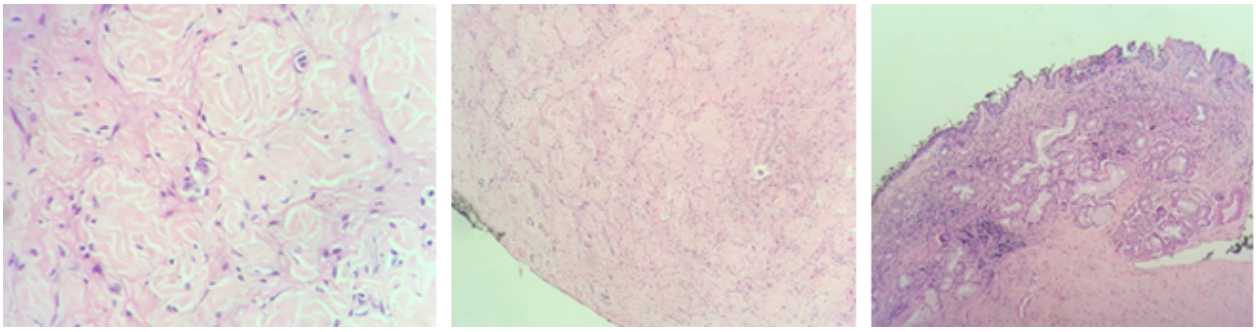

Figure 6: Imageof the histological analysis showed **6A:** Encapsulated neoplasm with interlacing bundles of elongated cells with wavy darkly stained nuclei with interspersed variably sized collagen bundles (H and E, X40). **6B:** Encapsulated neoplasm with interlacing bundles of elongated cells with wavy darkly stained nuclei with interspersed variably sized collagen bundles (H and E, X10). **6C:** Section shows respiratory epithelium with underlying submucosal gland and uncircumscribed lesion (H and E, X40).

The postoperative period was uneventful and the patient maintained a regular follow-up.

Discussion

There are many types of swellings in the midline dorsal part of the nose, such as nasal tumours, nasal gliomas, encephalocele, teratomas, lipomas, vascular anomalies, neuro fibromas and epidermoid cysts. Detection of intracranial extensions is of utmost importance to avoid complications that could result in irreversible brain damage [4]. In our case there is attachment to the left upper lateral cartilage of the nose but no intracranial extension. The gliomas, encephaloceles, and meningoceles can be differentiated from dermoid cysts by the fact that they are most commonly external, are located in the midline near the base of the nose, and are frequently associated with cranial defects. The glioma is firm and solid. The hemangioma is bluish-gray and distends with crying. Sebaceous cysts are superficial and show dimpling, and are attached to the skin, whereas lipomas are not cystic and are freely movable, whether superficial or deep. Inclusion and implantation cysts usually indicate a history of trauma and scarring [8]. In our case, the swelling was spherical, firm, painless, solitary and movable.

Nasal dermoid cysts are the most common congenital midline nasal lesions, accounting for 1% to 3% of all dermoid cysts [1]. The most common presentation is progressive swelling of the nasal dorsum, causing deformity and causing psychological distress to the patient or parents. In our case, the most common symptom was progressive swelling of the nasal dorsum without nasal obstruction.

NF1 affects 1 in 3000–4000 individuals worldwide. Although NF1 is usually inherited from autosomal chromosomes, 50% of mutations detected are de novo. The condition usually occurs in childhood [7]. Gliomas are the least common congenital midline masses. The male-to-female ratio is 3:2 and there is no concept of familial predisposition. Despite their names, these diseases do not represent true neoplasm. Their embryologic origin is also similar to encephalocele, except that they lack an intracranial connection in 70% of cases [9]. Similarly, in our case also there is no intracranial extension was seen.

Bony details of these midline nasal masses are best defined by CT, while MRI images soft tissues more precisely. The objective of imaging studies is to confirm the clinical diagnosis and to delineate any intracranial involvement if present [3]. The recurrence may occur in midline nasal masses but it occurs several years after the initial surgery. Therefore, the long-term follow-up of all patients with a history of midline nasal mass is essential [10].

A simple biopsy can be dangerous because it can cause cerebrospinal fluid leakage and meningitis in the event of intracranial dissemination. Excision and direct closure have the advantage of removing the cyst or abnormal skin overlying the sinus ostium. We also perform similar technique to excise a midline nasal mass. On the contrary open rhinoplasty, either alone or combined with direct excision, offers the opportunity to correct the position of the alar cartilages, which is often splayed by a lesion at the nasal tip [5].

The disease in the upper part of the nose is important and causes the pseudo-hypertelorism effect. Elimination of these lesions can improve the aesthetic appearance of the nose. Simultaneous medial canthal repositioning should be considered in these cases to reposition the canthus and prevent post-operative ectropion [6]. As of now, the patient is in a month post-operative

period with good wound healing and the external scarring of the patient has improved with normal facial symmetry.

Conclusion

Early intervention and comprehensive treatment of midline nasal masses with or without intracranial extension may help to achieve a definitive and aesthetic outcome. All patients require preoperative radiographic evaluation to detect deeper nasal or intracranial involvement. Long-term follow-up is required. Customized, precise surgery that emphasizes defined pathology and anatomy may lead to effective treatment with a lower recurrence rate and fewer complications. We hope this study will provide a sincere reading to torhinolaryngology surgeons and help improve their understanding of the management of midline nasal swellings in children.

Conflict of Interest

All authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethics Approval

All procedures performed in the study involving human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Consent to participate Informed consent was obtained from all individual participants in the study

Author's Contribution

A K Pandey concept, literature search, manuscript editing and review, Saurabh Nautiyal- manuscript writing, editing Shefali Chandel - data acquisition Saqib Ahmed, Aparna Bhardwaj- pathologic correlate, manuscript review. All authors read and approved the final manuscript.

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