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Sinonasal Schwannoma in a Six-Year-Old: An Unusual Presentation

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Abstract

Schwannomas commonly occur in the head and neck region and extremities, but nasal schwannoma is extremely rare. It accounts for less than 4% of benign solitary schwannomas of the head and neck region. Few cases and very few case series have been reported till date. We report the case of a six-year-old female who presented with mouth breathing of two years with persistent and progressive right nasal obstruction of three months. There was recurrent watery ipsilateral nasal discharge, but no epistaxis, no otologic or throat symptoms, no headaches, facial pain, proptosis or visual loss, no facial hypoesthesia, fever or weight loss. She also had right sided epiphora. She had used topical steroid and decongestant without improvement. Ten months prior to presentation, she had right dacrocystorhinostomy. Examination revealed mucosa mass completely filling the right nasal cavity with an ipsilateral deviation of nasal septum—it was painless with no contact bleeds. Nasal patency was clinically absent bilaterally. Computed tomogram revealed expansile isodense soft tissue mass in the right nasal cavity with heterogeneous enhancement on contrast administration which measured 2.5 cm by 3.7 cm by 5.2 cm. She had an intranasal excision of the tumour and histology revealed Antoni A and B patterns, which is diagnostic of schwannoma. She was fully relieved of symptoms after surgery. Conclusion: Solitary nasal schwannoma is rare especially in childhood. There are various causes of epiphora; definitive investigation of the cause in the index case was not established before dacrocystorhinostomy. Thorough nasal evaluation of epiphora or related ocular symptoms should be sought before definitive procedures.

Keywords

Schwannoma, Dacrocystorhinostomy, Nasolacrimal Duct, Antoni A and B Patterns

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1. Introduction

Schwannoma (also known as neurilemoma, Neuroma, neurolemoma and Schwann cell tumour) is the most common benign tumour of peripheral nerves [1]. They are rare encapsulated tumours that are derived from the Schwann cells of the sheath of myelinated nerves. They are usually solitary but can be multiple (Schwannomatosis), are universally S-100 positive and can also occur as lesions associated with Neurofibromatosis. They rarely cause neurological deficits and are usually extirpable [2]. Less than one percent becomes malignant.

Head and neck schwannomas constitute about 25% to 45% of all cases of schwannomas [3].

Only 4% of the lesions involve the sinonasal tract [3]: ethmoid sinus, maxillary sinus, nasal fossa, and rarely sphenoid [4]. Involvement of the nasal cavity or ethmoidal is said to be the commonest [5] [6].

Schwannomas of the sinonasal tract present with nonspecific symptoms but the majority are nasal (nasal obstruction, epistaxis and anosmia) [5].

Epiphora as the only presenting symptom has not been reported hence we report this unusual presentation.

2. Case

A six-year-old girl was referred to the Ear, Nose and Throat outpatient clinic by the ophthalmologist with persistent and progressive right nasal cavity obstruction with associated mouth breathing of three months. There was recurrent watery-to-mucoid ipsilateral nasal discharge, but no epistaxis. There was no history of snoring, excessive sneezing or symptoms suggestive of an allergy.

She had no otologic or throat symptoms, no history of headache, facial paraesthesia or pain, proptosis, visual loss or weight loss. Topical decongestants and subsequently topical steroids had been prescribed without improvement afterwards.

Ten months prior to presentation, she had right external dacrocystorhinostomy for right-sided epiphora of 3 years. No associated visual loss, itching, ocular pain, previous trauma, previously treated ocular disease or any of the aforementioned nasal symptoms at that time.

Examination revealed mouth breathing in a young girl, not chronically ill-looking or febrile, no clinical features of anaemia, jaundice or cyanosis. There was a mass completely filling the right nasal cavity with a deviation of the nasal septum to the left. The nasal cavity mucosa over the mass appeared normal—it was painless with no contact bleeding. Both nasal cavities were obstructed clinically, but worse on the right.

There was no extension of the mass into the nasopharynx or oropharynx clinically and no dental anarchy. Examination of the ear, throat and the other systems revealed essentially normal findings.

A Computerized tomogram of the paranasal sinuses revealed a well-defined expansile isodense mass appearing to arise from the lateral wall of the right nasal cavity causing deviation of the septum to the lateral wall of the left nasal cavity. It extended superiorly to involve the lamina papyracea of the ipsilateral ethmoidal sinus and the floor of the left sphenoid sinus. There was pressure remodelling of the adjacent bony walls but no intracranial extension (**Figures 1-3**).

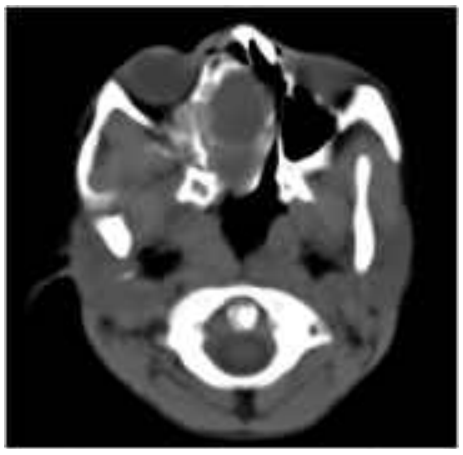

Figure 1. (Axial CT image). Mass completely filling the right nasal cavity and maxillary sinus.

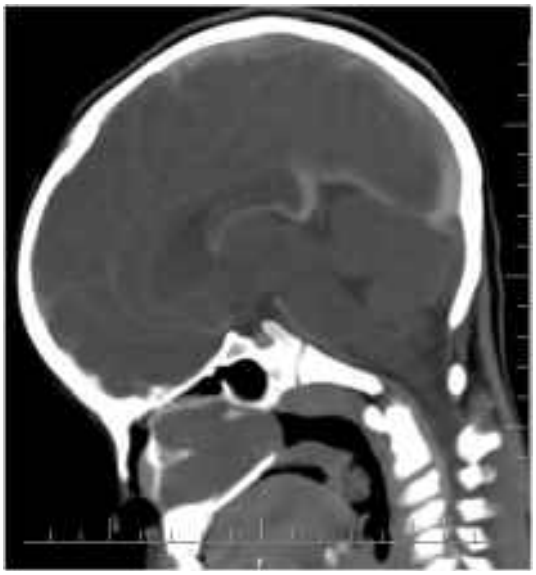

Figure 2. (Sagittal CT image). No evidence of intracranial extension.

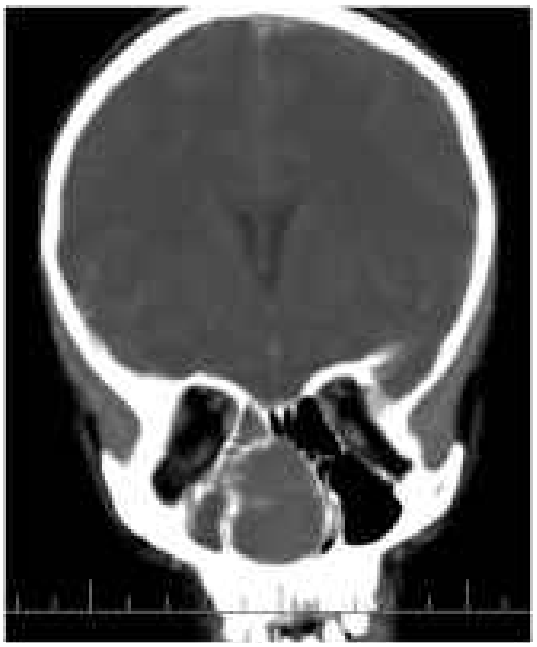

Figure 3. (Coronal CT image). Deviation of nasal septum.

The aforementioned sinuses were also filled with isodense lesions, possibly retained secretions. The mass had heterogeneous enhancement on contrast administration and measured 2.5 cm by 3.7 cm by 5.2 cm. Haematological and Biochemical parameters (Full Blood Count and Electrolyte, Urea and Creatinine) were within physiologic ranges respectively. Plain radiograph of the chest showed no abnormality.

She had complete intranasal excision of the mass. The mass having completely filled the nasal cavity prevented the introduction of the Hopkins' telescope but after the bulk of the mass was removed, remnants were completely resected endoscopically.

She was fully relieved of the symptoms after surgery. Follow-up in the outpatients' clinic has been satisfactory with no evidence of recurrence in one year.

Histology of the excised specimen revealed Antoni A and B patterns, which was suggestive of schwannoma. Tissue specimen was strongly immunoreactive to S-100 (**Figure 4**), had Verocay bodies (**Figure 5**), it was Epithelial Membrane Antigen (EMA) negative (**Figure 6**), Neuron Specific Enolase (NSE) negative (**Figure 7**), desmin negative (**Figure 8**) and vimentin positive (**Figure 9**).

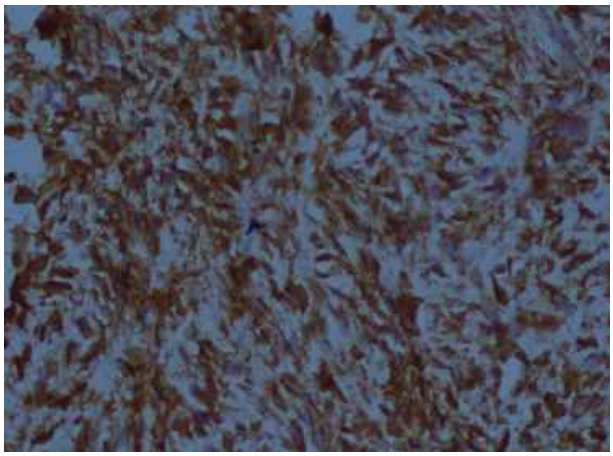

Figure 4. S-100 positive ×400.

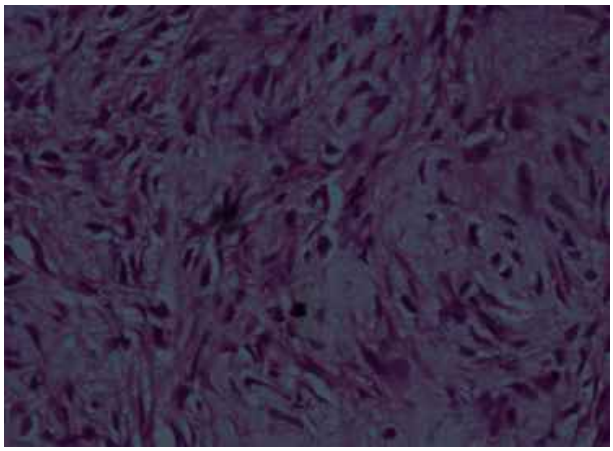

Figure 5. Presence of verocay bodies ×100.

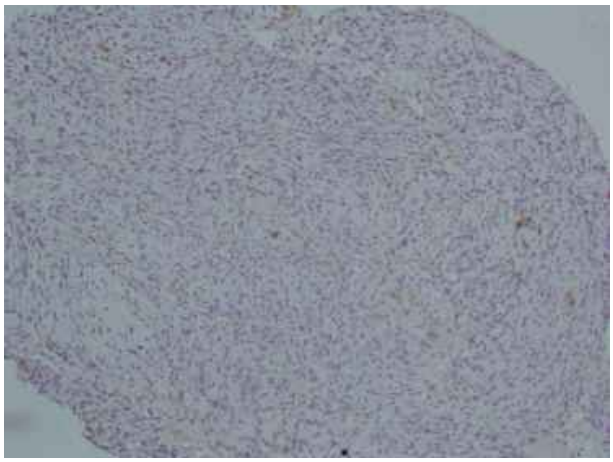

Figure 6. EMA negative ×100.

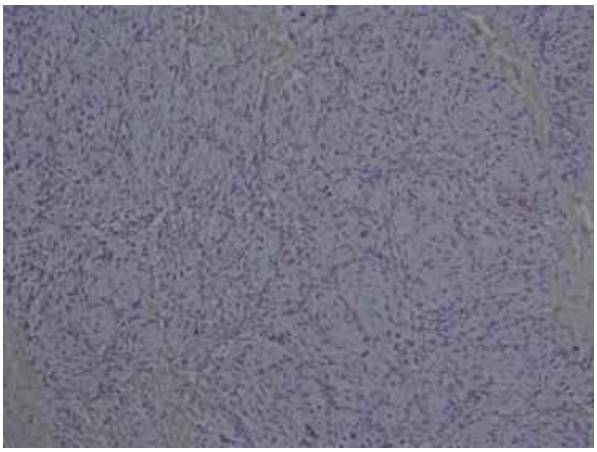

Figure 7. NSE negative ×40.

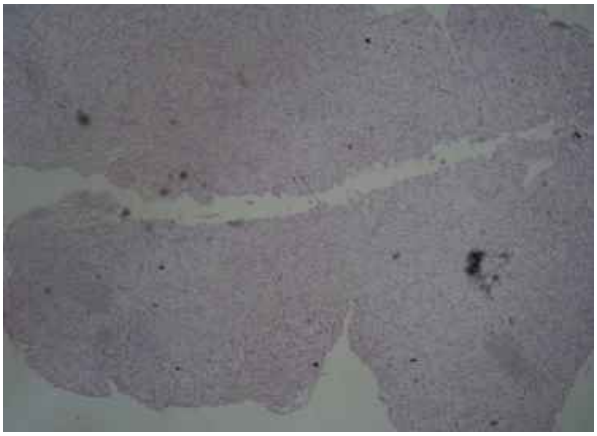

Figure 8. Desmin negative ×40.

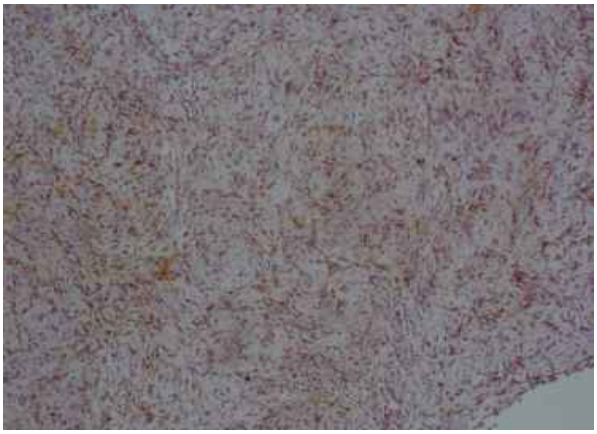

Figure 9. Vimentin positive ×100.

3. Discussion

Obstructive epiphora has not been reported as the only presenting complaint in patients with sinonasal schwannoma, this was the only symptom the patient had before dacrocystorhinostomy. This is a retrospective diagnosis as no initial imaging was done prior to surgery. It is thought that the tumour would have obstructed the nasolacrimal duct along its course and/or at its opening at the inferior meatus. However, the precise origin of the tumour

on the lateral wall of the nasal cavity is not certain.

Nasal obstructive symptoms are the most common presentations of sinonasal schwannoma. However, other rare presentations have also been described—right orbital proptosis and right-sided headache, with intracranial extension [5] [7].

The characteristic features of sinonasal schwannoma on computed tomography are; a well-defined iso-attenuating soft-tissue mass, most frequently occurring in the nasal cavity and ethmoid sinus with pressure remodelling of the adjacent bony wall (medial wall of right maxillary sinus). These are similar to the CT findings in our patient. There was no intracranial extension.

Mild contrast enhancement, cystic or haemorrhagic changes are features that have been noted in some cases. Magnetic Resonance images of sinonasal Schwannomas appears as predominantly high signal intensity mass on T_2 weighted. There is also strong enhancement on contrast [8].

Schwannomas show biphasic growth patterns (Antoni A and Antoni B). In the Antoni A pattern of growth, elongated cells with cytoplasmic processes, are arranged in fascicles in areas of moderate to high cellularity with little stromal matrix; the "nuclear-free zones" of processes that lie between the regions of nuclear palisading are termed Verocay bodies.

In the Antoni B pattern of growth, the tumor is less densely cellular thus otherwise termed hypocellular area. It has microcysts and myxoid changes [9]. In addition to these, sinonasal schwannomas have woven bone and remnants of pseudostratified ciliated epithelium and seromucinous glands—perhaps in keeping with the location in the nasal cavity.

Their immune reactivity to S-100 is the basis for their staining with it—thus differentiating the tissue from a neurofibroma. Other stains described are; Epithelial Membrane Antigen (EMA), S-100 (Schwann cells), calcinurin, laminin, type 4 collagen, vimentin, CD68, GFAP [9].

The patient's tissue specimen was strongly immunoreactive to S-100, this confirmed a schwannoma and differentiated it from a neurofibroma, it also had Verocay bodies, it was EMA negative and this ruled out a malignant transformation of the tissue or a meningioma, Vimentin was also positive, histological evidence that the tissue was of mesenchymal origin.

Schwannomas are largely radio-resistant, so surgery is the best treatment option [3] [10]. Complete excision of the mass is rarely followed by recurrence. As stated above, a close differential of nasal schwannoma is a benign neurofibroma. This has the possibility of recurrence after excision as well as malignant transformation. However, the histological characteristics, as well as immunohistochemistry techniques, will help to differentiate the two lesions [11]. Various approaches have been described but the endoscopic approach has been described to be the best approach with reduced morbidity and mortality [12]. It could also be combined with an external approach.

Prognosis is excellent and malignant change is extremely rare in Schwannomas, although local recurrence can follow incomplete resection, it is low in cases where the endoscopic approach was used [12] [13]. This was done in this patient. She has remained free of symptoms and examination showed no evidence of recurrence one year after surgical excision.

4. Conclusion

There are numerous obstructive causes of epiphora. A nasal evaluation, including diagnostic nasal endoscopy and radiologic imaging, cannot be overemphasized in the evaluation of the lacrimal system in obstructive epiphora. This patient had an external dacryocystorhinostomy done without identifying any possible nasal aetiology. The opening of the nasolacrimal duct relates anteriorly to the inferior meatus on the lateral wall of the nasal cavity where it could be obstructed. Solitary nasal schwannoma is indeed rare in children and it is, therefore, worthy of note that meticulous nasal evaluation in obstructive epiphora, or in related ocular symptoms should be sought before definitive surgical procedures are carried out.

Consent

Full consent to report this case including the use of the Computed Tomogram and histological images was obtained from the patient's parents.

Conflict of Interest

The author has no conflict of interest.

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