Subcutaneous Nasal Schwannoma in a Pediatric Patient: A Rare Case Report with Emphasis on Histopathology’s Role in Differential Diagnosis

**Patient:** Female, 9-year-old

**Final Diagnosis:** Subcutaneous nasal schwannoma

**Symptoms:** Painless nasal swelling

**Clinical Procedure:** Excisional biopsy

**Specialty:** Otolaryngology

**Objective:** Rare disease

**Background:** Schwannomas are rare and benign tumors of the nerve sheath, composed of Schwann cells, and they are extremely rare in the nasal area. Here, we report a case that presented to our clinic as a growing nasal mass and was found to be a unilateral subcutaneous schwannoma. There have been a few previous cases reported of such patients having nasal obstruction, epistaxis, or other symptoms, but our patient did not. We stress the importance of considering schwannoma in the differential diagnosis of nasal masses, even in pediatric patients, and the role of histopathology differentiating it from other diagnoses such as neurofibroma.

**Case Report:** Our patient was a 9-year-old girl with a painless nasal swelling on the nasal bridge that she first noticed 2 years ago, which started growing gradually and began to become firm. She was otherwise asymptomatic and had no relevant family history. Histopathology revealed an encapsulated spindle cell tumor with both hypo- and hyper-cellular areas, and immunohistochemistry showed that the tumor was strongly positive for S-100 and negative for both desmin and CD34, with blood vessels marking. A final diagnosis of schwannoma was made.

**Conclusions:** We presented a case of nasal septal schwannoma, emphasizing the importance of considering schwannoma in the differential diagnosis of nasal masses, and the role of histopathology to rule out other possible diagnoses.

**Keywords:** Surgical Oncology • Nasal Cavity • Neoplasms
Background

Schwannomas, also called neurilemmomas, are benign nerve sheath tumors that originate from Schwann cells. These tumors commonly emerge in middle-aged adults, displaying equal occurrence across sexes and racial groups. Schwannomas can occur in various locations, with the most common sites being the head and neck region, which account for 25-45% of schwannomas [1]. Within the head and neck region, the most common site is the cerebellopontine angle [2].

Nasal schwannomas are extremely rare, accounting for less than 4% of all sinonasal tumors. They are typically solitary and slow-growing, and their clinical presentation varies depending on the location and size of the tumor. Common reported symptoms include nasal obstruction, epistaxis, and rhinorrhea, but some patients also present with facial pain, anosmia, and proptosis [3], although imaging modalities can be useful to assess the origin and extent of the tumor. The final diagnosis of schwannoma must be confirmed by histopathology [4].

In this report, we present a rare case of subcutaneous nasal schwannoma, discussing its clinical presentation and providing insight into the histopathological features, how they differ from other possible diagnoses, and the management of this rare tumor.

Case Report

A previously healthy 9-year-old girl presented to our Ear Nose and Throat (ENT) clinic with painless nasal swelling of 2 years duration. The swelling was first noted 2 years ago following a facial trauma. It was mobile and growing gradually on the nasal bridge. It had been soft but was recently noticed to become firm. The patient was otherwise asymptomatic with no constitutional symptoms, pain, headache, epistaxis, rhinorrhea, smelling problems, or breathing difficulties reported (Figure 1A). The patient had no medical or surgical history and was not on any medication. No other family member had ever reported a similar problem or neurofibromatosis. Her general physical examination was normal, without any café-au-lait spots, Lisch nodules, or other symptom suggesting neurofibromatosis.

On examination, the mass was mobile, located on the mid-line of the anterior aspect of the nasal bridge, approximately 1×2 cm, oval, with a smooth surface, and did not fluctuate. The mass was firm and non-tender. No remarkable findings were observed on rhinoscopy. The remaining ENT and head and neck examinations were normal.

Computed tomography (CT) and magnetic resonance imaging (MRI) without contrast (Figure 2A-2C) showed a dense lesion on the left anterior aspect of the nose, measuring 2×1.24×2.2

Figure 1. (A) Patient with unilateral nasal swelling. (B) Patient post-operatively.
cm. It was well-defined with some lobulations. The imaging studies were initially suggestive of subcutaneous lipoma. An excisional biopsy using open rhinoplasty general anesthesia was made. Infiltration of diluted adrenaline and transcolumellar skin incision and rim incision was performed. Then, deskeltonization was done. Dissection around the mass was done with complete removal of the mass. Hemostasis was ensured using pressure packs and bipolar cautery. The wound was sutured, and a Steri-Strip was put on the wound, which was removed 1 week later in the clinic (Figure 1B).

On gross examination the specimen revealed multiple grayish white pieces of tissue.

Multiple tissue sections were stained with hematoxylin and eosin (H&E).

On histopathology the examined section showed an encapsulated spindle cell tumor with both hypo- and hyper-cellular areas. A confirmatory immunohistochemical examination showed the tumor was strongly positive for S-100, negative for both desmin and CD34, with blood vessels marking, and a final diagnosis of schwannoma was made (Figure 3A-3D).

Discussion

Schwannomas are peripheral nerve tumors that are most common in young adults [1]. Our case report is one of the few schwannoma in a pediatric patient [5,6], and while our patient had an asymptomatic, growing mass, other cases of naso-schal schwannoma in the literature presented with isolated nasal obstruction [2,7,8], accompanied by anosmia or voice changes [4] and epistaxis [9,10].

The potential differential diagnoses for nasal cavity schwannomas encompass neurofibromas, meningiomas, angiofibroma, glomangiopericytomas, leiomyomas, inflammatory polyps, inverted papilloma, melanomas, and neuroblastomas [11]. It is not unusual for schwannomas to be misdiagnosed. The differentiation of schwannomas presents significant challenges when relying solely on symptoms, endoscopy, and imaging. An accurate diagnosis is contingent upon histopathological and immunohistochemical analyses, and clinicians should remain watchful for signs and
symptoms of NF1, such as café-au-lait spots, multiple neurofibromas, and Lisch nodules [12], which were negative in our case.

Under microscopic examination, schwannomas exhibit 2 distinct patterns. The Antoni A region displays densely packed spindle cells forming a palisading nucleus around a non-cellular central area, known as Verocay bodies. In contrast, the Antoni B region shows loosely arranged tissue comprising diverse cells separated by a rich myxoid matrix. Due to challenges in diagnosis based solely on morphology, immunohistochemical stains are frequently utilized. Presence of the S-100 protein can help confirm the diagnosis. Although both schwannomas and neurofibromas react to the S-100 protein in immunostaining, research indicates that this reaction is more pronounced in schwannomas compared to neurofibromas. Additionally, Calretinin and CD56 are notably specific to schwannomas, while CD34 and factor XIIIa are more sensitive indicators for neurofibromas. These markers play a crucial role in facilitating

Figure 3. (A) Microscopic appearance showing a spindle cell tumor with both hypo and hypercellular areas. (B) S100 protein showing a diffuse staining as a characteristic feature for schwannoma. (C) Immunohistochemistry showing CD34 negativity in tumor cells with blood vessels marking. (D) Immunohistochemistry showing desmin negativity.
the differentiation between these conditions [3]. Our case revealed positive S-100 protein and negative CD34 and desmin. Peripheral nerve tumors also rarely present simultaneously, as in a previous report [13].

Conservative surgical excision is the standard treatment for nasal cavity schwannomas. The surgical procedure depends on the tumor size, location, and invasion of adjacent structures [10]. Several surgical techniques are discussed in the literature, including open septorhinoplasty [7], lateral rhinotomy [10], and endonasal endoscopic excision [2,4], with latter considered the standard surgical approach [1].

A special aspect of our case is that the mass appeared after facial trauma, which was not found in our literature review of nasal schwannoma, except for 1 case in Brazil [14]. Interestingly, a previous paper suggested that schwannomas tend to occur more frequently in areas susceptible to nerve compression or physical trauma, often referred to as “predilection sites”. Within the cranial nerves, the vestibular, trigeminal, and hypoglossal nerves share a confined anatomical pathway, making them susceptible to physical strain and injury and more frequently impacted by schwannoma, thus supporting the nerve regeneration failure theory [15], but this needs to be verified in further research.

Conclusions

Despite being rare, subcutaneous schwannomas should be considered in the differential diagnosis of nasal masses. The clinical presentation can be variable and non-specific, making pre-operative diagnosis difficult. However, histopathological examination is the only accurate way to confirm a diagnosis of schwannoma and differentiate it from other possible diagnoses that may have a similar presentation. Complete surgical excision is the treatment of choice, and endoscopic surgery has emerged as a safe and effective option. Further studies are needed to better understand the pathogenesis and optimal treatment strategies for this rare tumor.

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References: