Case Report

Schwannoma in the nasal cavity: A benign nerve sheath tumor at an odd location

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ABSTRACT

Schwannomas are neurogenic tumors arising from the Schwann cells present in the neural sheath of the myelinated nerves. These are benign tumors that can arise anywhere in the body. Schwannomas are rarely seen in the nose and paranasal sinuses representing <4% of all head-and-neck schwannomas. Surgical excision is the treatment of choice in these cases. Nasal polyps are common nasal cavity lesions which are usually inflammatory polyps. Schwannoma of the nasal cavity usually presented with headache, unilateral nasal obstruction due to mass obliterating the nasal cavity, and epistaxis. Here, we report the case of a 47-year-old female who presented with a mass in the left nasal cavity causing the nasal blockage.

Key words: Benign, Histopathological, Nasal cavity, Schwannoma, Soft tissue

Schwannomas, also known as Neurilemmoma or Neurinoma, comprise dedifferentiated Schwann cells [1]. Schwann cells are present in the nerve sheath of myelinated nerves such as the cranial, peripheral, or autonomic nerves, and therefore, schwannoma can arise in any part of the body that has a myelinated nerve fiber with Schwann cell sheath or neural sheath [2]. The most common sites of presentation of Schwannoma include extremities, retroperitoneum, cerebellopontine angle, mediastinum, and rarely head-and-neck region [2]. Schwannomas of the head-and-neck region constitute only 25% and among them, <4% of cases are confined to the sinonasal cavity. The most frequent site of these tumors in the head-and-neck is the branches of the trigeminal and acoustic nerves [1,3]. Cranial nerves such as optic and olfactory nerves do not have Schwann cell sheath; hence, schwannomas arising from these locations are extremely rare. In the past literature, head-and-neck schwannomas have been reported in the oral cavity, larynx, pharynx, middle ear, parotid gland, scalp, and sinonasal tract [3,4]. Nasal polyps are common nasal cavity lesions which are usually inflammatory polyps. Schwannoma of the nasal cavity usually presented with headache, unilateral nasal obstruction due to mass obliterating the nasal cavity, and epistaxis [1,4]. Since there are no characteristic features of schwannoma in radiology, histopathology is the gold standard for the diagnosis in this case.

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CASE REPORT

A 47-year-old female came with complaints of left-sided nasal blockage for 1 month. The patient also had a history of frequent headaches, bleeding from the nose twice, and a reduced sense of smell for 10 days. She was a known hypertensive for 3 years, on regular medications and her blood pressure was under control.

General examination revealed a normal appearance with no generalized lymphadenopathy or pedal edema. Vital Signs like pulse rate, temperature, respiratory rate, and blood pressure were normal. On examination, the left nasal cavity revealed a mass arising from the left inferior turbinate and occupying the nasal cavity.

Positron emission tomography-computed tomography showed a heterogeneously enhancing soft-tissue density, ovoid lesion measures 2.3 × 1.3 cm in bilateral ethmoid, sphenoid, and left frontal sinusitis.

Endoscopic removal of nasal mass with midfacial degloving was done. Intraoperatively, an endoscopic examination was done with a 4-mm 0° telescope which revealed a 2 cm × 1 cm mass in the left nasal cavity arising from the left inferior turbinate. The mass was soft and fleshy with a broad-based attachment to the anterior end of the inferior turbinate. There was no significant bleeding during excision. The mass was excised completely and removed transnasally. Hemostasis was secured intraoperatively using Neuray packing which was removed after 5 min. Post-operative

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packing was done. The next day post-operative nasal pack was removed and there was no evidence of bleeding. The patient was discharged home the next day. The patient was seen at the outpatient clinic 2 weeks post-surgery with symptom-free and no post-operative complications

Grossly, we received a small nodule measuring 2 cm × 1.5 cm × 1 cm with attached cartilage at the base of the lesion. The cut surface was tan-yellow and soft. Also received an irregular piece of yellow-brown soft tissue measuring 2 cm × 1 cm × 1 cm.

Microscopy revealed sinonasal mucosa lined by stratified squamous epithelium without significant nuclear atypia. Hyperplastic mucous glands were noted. Subepithelial tissue showed a circumscribed benign spindle cell tumor arranged in intersecting fascicles and bundles. They showed elongated cells with tapering ends interspersed with collagen fibrosis, ill-defined cytoplasm, dense chromatin, and inconspicuous to prominent nucleoli. Few areas showed degenerative atypia. Rare mitotic figures of 0–1/10 high power field were noted. Focal nuclear palisading was noted. No definite necrosis was seen. Thick hyalinized blood vessels and variable lymphoplasmacytic infiltrate were noted (Fig. 1a and b).

Immunohistochemical studies were done and the tumor cells expressed S-100 protein (Fig. 1c), SOX-10 (SRY-related HMG-box 10), and glial fibrillary acid protein (focal); while they were immunonegative for smooth muscle actin (Fig. 1d), cytokeratin, desmin, cluster differentiation 34 (CD34), human melanoma black-45, melan-A, and microphthalmia associated transcription factor. H3K27me3 protein showed diffuse intact nuclear expression. The Mib-1 labeling index was 6–8% in areas of the highest proliferative activity.

Hence, a diagnosis of benign nerve sheath tumor in favor of Schwannoma was made. The patient is under regular follow-up every 6 months with no recurrence to date.

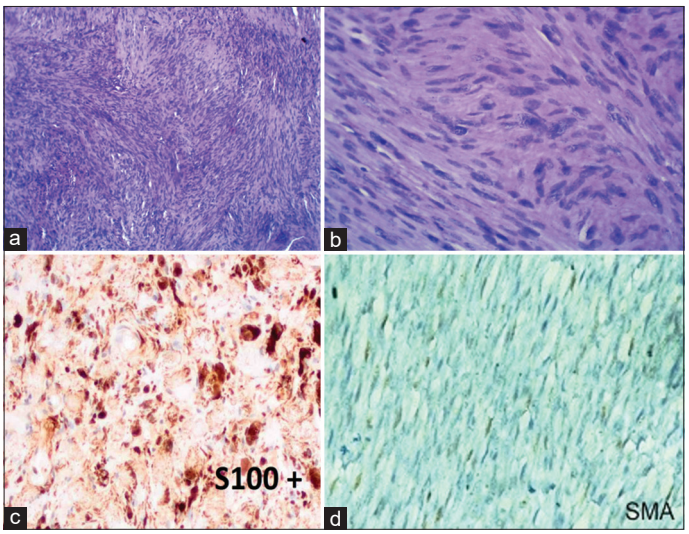

Figure 1: Photomicrograph showing (a) benign spindle shaped tumor cells in intersecting fascicles and bundles (hematoxylin and eosin stain, ×10); (b) spindled tumor cells in the hypercellular area (hematoxylin and eosin stain, ×40); (c) S100 protein immunoreactive spindle tumor cells (S100 protein positive stain, ×40); and (d) immunonegative staining for smooth muscle actin (SMA) of the tumor cells (SMA negative stain, ×40)

DISCUSSION

Schwannomas are benign tumors that rarely undergo malignant transformation [1]. These tumors arise from the differentiated Schwann cells present in the peripheral nerve sheath derived from the neuroectoderm [2]. Stout termed these tumors neurilemmoma as they originated from the cells of the sheath of Schwann. Since the originating cells are Schwann cells, these tumors are often termed schwannomas. Schwannomas usually involve the peripheral nerves of the skin in the head-and-neck region and are frequently seen to involve the flexor aspects of the extremities [1,2]. Head-and-neck schwannomas constitute about 25–45% of the total cases, among these <4% involve the sinonasal tract. In our literature search, we found that very few cases of schwannomas were reported in sites such as the nasal cavity, maxilla, frontal sinus, ethmoid sinus, sphenoid sinus, and alae nasi [1,5].

Acoustic neuroma, which is the most common head-and-neck schwannoma, has been found to arise from the vestibulocochlear nerve [6]. On the other hand, schwannomas occurring in the nasal cavity have been most commonly seen arising from the trigeminal nerve branches like the ophthalmic and maxillary branches [2]. These tumors are also seen arising from the inferior orbital nerve, parasympathetic fibers of the sphenopalatine ganglion, sympathetic fibers of the carotid plexus, and nerves of the nasal mucosa [2,4]. Patients usually presented from 6 to 78 years of age with nasal cavity schwannoma and there was no racial or gender predilection in these cases [5,7,8]. Schwannomas generally present as solitary lesions without any associated genetic syndrome but when there are multiple schwannomas in the same individual, neurofibromatosis Type 2 should be ruled out in these patients. Schwannomas are benign and slow-growing tumors that rarely undergo malignant transformation [1-3,8,9].

Patients with schwannomas of the nasal cavity present with nasal obstruction, external nasal deformity, and epistaxis, other symptoms may include headache, exophthalmos, facial swelling and pain, visual disturbances, rhinorrhea, and cranial nerve palsies [1]. Nasal cavity masses have a list of differential diagnoses that include antrochoanal polyp (most common), inverted papilloma, fibrous dysplasia, concha bullosa, capillary hemangioma, mucocele, septal dermoid, neurofibroma, fibro myxoma, neurilemmoma, chondrosarcoma, squamous cell carcinoma, meningioma, esthesioneuroblastoma, and nasopharyngeal angiofibroma extending into the nasal cavity. The most common differential diagnosis of schwannomas includes leiomyoma, leiomyosarcomas, malignant melanomas, and hemangiopericytoma. [3,5].

Contrast-enhanced computed tomography of the paranasal sinuses is helpful in evaluating the size of the mass, origin, localization of the tumor, and involvement of surrounding structures. When the schwannoma is present in the nasal cavity, the important differential diagnoses are solitary fibrous tumor, fibro myxoma, juvenile nasopharyngeal angiofibroma, and neurofibroma. The solitary fibrous tumor has alternating cellular and fibrous areas along with hemangiopericytoma-like blood vessels and the tumor cells show strong positivity for CD34, CD23, and CD8. Nasopharyngeal angiofibroma comprises a

mixture of fibrous stroma and blood vessels. Neurofibromas are non-encapsulated tumors composed of Schwann cells, axons, perineural cells, and fibroblasts, these tumors show increased chances of malignant transformation. On immunohistochemical studies, they show focal S-100 protein positivity [2,5].

Complete surgical excision of the tumor is the treatment of choice along with histopathological confirmation. Recurrence has been very rarely reported. On macroscopic examination, these tumors are often well encapsulated and appear cystic or gelatinous [2,5,7,8]. The tumor appears well-demarcated, round to oval in shape, grayish in color, and soft with a tan yellow cut surface. The capsule of these tumors has been found to be derived from the perineurium of the originating nerve. Schwannomas arising from the autonomic nerve fibers lack capsule as they are devoid of perineural cells. Unencapsulation does not mean the tumor is malignant, it implies the aggressive nature of the tumor and makes it difficult for the surgeon to extract the tumor in Toto. Schwannomas can also undergo degenerative changes such as lipidization, cystic degeneration, necrosis, and angiomatous change with thrombi formation [2,8,10].

Microscopically, Schwannomas comprise two major architectural patterns, Antoni A and Antoni B areas in varying proportions [2,5,7,8]. Antoni A has two types of patterns – Type A fasciculated pattern and Type B reticular pattern. These areas are hypercellular and composed of spindle-shaped cells arranged in a compact stroma in an interlacing fascicular pattern along with palisading nuclei arranged in parallel rows which are known as the Verocay bodies. Antoni B areas are hypocellular composed of very few spindle cells arranged in a disorganized loose fibrillary myxoid stroma along with few hyalinized blood vessels [2,4,5,7]. There are two major differences between schwannoma of the nasal cavity and schwannoma from other sites (a) nasal schwannomas do not have a fibrous capsule, as the autonomic nerve fibers of the nasal mucosa do not have perineural sheaths; and (b) nasal schwannomas are usually hypercellular.

Immunohistochemical staining for S-100 protein shows intense/strong nuclear and cytoplasmic staining, particularly in the Antoni A areas, which helps in differentiating Schwannomas from other neural tumors and malignant peripheral nerve sheath tumors [1,2,4,7]. Recurrence of schwannoma after complete surgical excision is rare but longstanding benign schwannomas undergoing malignant transformation have been reported, hence regular follow-up is always recommended [1,2,4,8].

CONCLUSION

Nasal cavity schwannomas are very rare, benign, and slow-growing tumors, and a confirmatory diagnosis is made only by histopathological examination. Schwannomas should be considered in the differential diagnosis when a patient presents with a cystic mass in the nasal cavity before considering it a mucocele. The nasal mucosa does not have perineural cells because of the autonomic nerve supply; hence, the tumor lacks an intact capsule explaining the aggressive growth, destruction, local bone erosion, and recurrence of these tumors but this does not imply that the tumor is malignant.

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