Case Report

Schwannoma of the Nasal Septum: A Rare Clinicopathological Case Report in an 18-Year-Old Female

Abstract

Schwannoma, a benign tumor, arise from schwann cells of myelin sheath; occur anywhere in the body but commonly occur on flexor aspect of extremities. Nasal septum being the rarer site. We report a case of nasal septum schwannoma in an 18-year-old female presented with intermittent epistaxis and progressively increasing nasal obstruction for 2-year duration. The differential diagnosis of juvenile angiofibroma, pyogenic granuloma, and pleomorphic adenoma was made and complete surgical excision was done. Histopathological examination revealed ciliated stratified columnar epithelium, underlying tumor area with two distinct patterns, mainly hypercellular and few hypocellular areas. The cells have spindle shaped pointed basophilic nuclei with abundant eosinophilic cytoplasm. Overall feature was suggestive of nasal septum schwannoma. For confirmation, immunohistochemical staining with S-100 was done and tumor was found positive. Herein, we report the clinicopathological features of nasal septum schwannoma in an 18-year-old female.

Keywords: *Juvenile angiofibroma, S-100, Schwann cell, Verocay bodies*

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Introduction

Schwannoma, a rare slow-growing benign tumors, arise from Schwann cells of myelin sheath of peripheral nerve, autonomic nerves, and/or cranial nerves.[1,2] Mostly, they occur in middle-aged adults, without gender and race predilection. Schwannoma may occur anywhere in the body commonly occurring on flexor aspect of extremities, posterior spinal nerve roots, mediastinum, retroperitoneum, cerebellopontine angle, and head and neck region.[1,2] Of all the schwannoma, 25 to 45% are confined to the head and neck region, in which nasal cavity and paranasal sinuses contribute about 4% and nasal septum the rarest site.[1,2]

Case Report

An 18 year old female presented to the Outpatient Department of Otorhinolaryngology with complaints of intermittent epistaxis and progressively increasing nasal obstruction for two year with foul smelling discharge and snoring. Local examination revealed distorted nasal pyramid. Anterior rhinoscopy shows right sided nasal septum deviation and a mass in left nostril attached to nasal septum with almost completely filling the left nostril. Posterior rhinoscopy showed a lobulated mass completely obliterating the left side nasal choana. Paranasal sinus computed tomography (CT) scan showed deviation of nasal septum to the right site and polypoidal soft mass in the left nasal cavity. On the basis of history, clinical examination and imaging finding the differential diagnosis of juvenile angiofibroma, pyogenic granuloma and pleomorphic adenoma was made. The patient was planned for surgery. The patient underwent surgical excision of the mass via lateral rhinotomy under general anesthesia. The excised tissue was sent for histopathological examination. On gross examination, the specimen revealed multiple grayish white pieces of tissue all together measuring 5.8 cm × 4.3 cm × 2.2 cm [Figure 1]. Multiple tissue sections were passed and processed to obtain 3–4 µm thick section and stain with H and E. On histopathological examination, the multiple sections examined revealed ciliated stratified columnar epithelium lined tissue with underlying tumor area with two distinct patterns mainly hypercellular and

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few hypocellular. In hypercellular areas, the cells were arranged in fascicles in a collagenous stroma. The cells have spindle shaped pointed basophilic nuclei and poorly defined eosinophilic cytoplasm. Some of these cells show nuclear palisading, surrounding an eosinophilic cytoplasm known as Verocay bodies. In hypocellular areas, these spindle cells are separated by an abundant myxoid stroma and also show presence of thick walled hyalinised blood vessels [Figure 2a-c]. Above histological features were those of schwannoma. For confirmation, immunohistochemical examination with S-100 was done and the tumor cells were diffusely positive for it [Figure 3a and b]. The diagnosis of nasal schwannoma was confirmed.

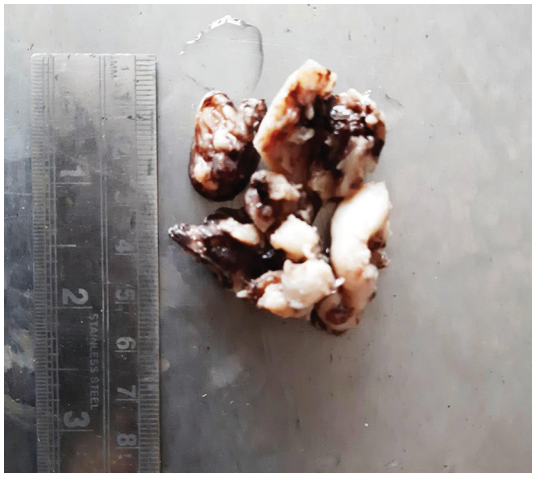

Figure 1: Multiple grayish white pieces of tissue showing myxoid changes at places

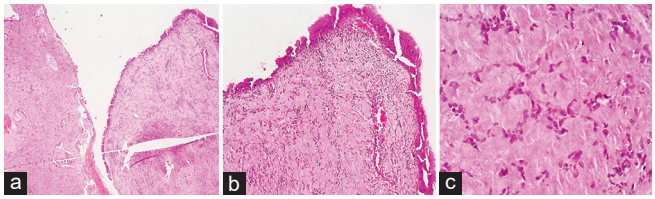

Figure 2: (a) Stratified epithelium with underlying tumor consisting of hypercellular and few hypocellular areas (×4), (b) ciliated stratified epithelium with underlying tumor composed of organized stroma with spindle shaped cells (×10), (c) spindle cells with parallel rows of palisading nuclei surrounding an acellular central material, known as Verrocay bodies (×40)

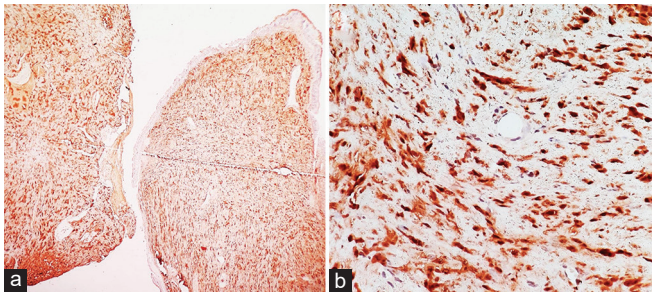

Figure 3: (a and b) Tumor showing immunopositivity with S-100 (IHC, ×4 and × 40). IHC: Immunohistochemistry

Discussion

Verocay described schwannoma in 1910 first time. Schwannoma, benign encapsulated nerve sheath neoplasm, originates from the neuroectodermal schwann cells of sheaths of the cranial, peripheral, and autonomic nerve.[1] In 1935, the term neurilemmoma was given by Stout believing that tumor arose from cells of sheath of Schwann.[2,3] The first case of nasal septum schwannoma was described by Bogdasarian and Stout in 1943.[4,5]

Schwannoma may arise from multiple sites. In head and neck region mostly arise from 8th cranial nerve (vestibulocochlear), never arise from optic and olfactory nerve as these nerve lack nerve sheaths which contain schwann cells.[5] On the basis of location, schwannoma are divided into nonvestibular schwannoma, extracranial head and neck schwannoma and intracranial acoustic schwannoma. Sinonasal schwannomas are rare and account only about 4% of all head and neck schwannomas.[1] In sinonasal tract, the most common site involved is the ethmoid sinus followed by maxillary sinus, nasal cavity and sphenoid sinus. In nasal cavity, the nasal septum is rarer site. The origin of sinonasal tract schwannomas is difficult to determine because of the thin nerve innervation of the nasal cavity. Sympathetic nerve from the stellate ganglion, parasympathetic nerve from sphenopalatine ganglion and sensory nerves from branches of the ophthalmic nerve (anterior ethmoid nerve) and maxillary nerves (nasopalatine nerve) are considered as the possible nerves of their origin.[1,6]

Usually, schwannomas are encapsulated; the capsule is derived from the perineurium of the nerve of origin. However, nasal schwannoma arising from autonomic nervous system fibers lack encapsulation due to devoid of perineural cells.[2,4] Unencapsulation in nasal schwannoma probably explains the aggressive pattern compared to that of other locations. However, lack of encapsulation does not imply malignancy, but lack of encapsulation might make the tumor more difficult to define and extract completely for clinician.[2,7]

The most common symptoms of nasal cavity schwannomas are nasal obstruction, epistaxis, rhinorrhea, and deformity of the nasal pyramids. Headache, hyposmia, nasal swelling and pain, exophthalmos, diplopia, and facial distortion were less recorded and usually implied due to extended lesions with adjacent structure involvement.[1,2,4,5,8,9]

Contrast-enhanced CT scan demonstrates peripheral enhancing neovascular areas and nonenhancing central necrotic or cystic area of the tumor.[1,2,5] Magnetic resonance imaging (MRI) is superior to CT in differentiating tumors from inflammatory changes and in the evaluation of their extranasal extensions.[1,2] Fascicular sign, seen at MRI, implies a lesion of neurogenic origin.[10]

Schwannomas, perineuromas, neurofibromas, myxomas, and granular cell tumors are called as peripheral nerve

sheath tumors which develop from Schwann cells, perineural cells, and neural fibroblasts. Differentiations of these tumors are made by immunohistochemical staining. S-100 protein expression is diagnostic for schwannoma.[10]

Histological examination is still the gold standard for diagnosis because of its atypical clinical symptoms and lack of specific image manifestation. Macroscopically, tumor is solitary, well demarcated with an oval, round, or fusiform shape, grayish to yellowish in color, fleshy and shiny on cut surface.[2] On microscopic examination, schwannoma consist of two distinct areas. Antoni-A area composed of compact organized cellular stroma with elongated spindle cells and dense spindle cells with parallel rows of palisading nuclei surrounding an acellular central area, known as Verocay bodies. While Antoni-B area shows a loose disorganized tissue composed of few spindle cells separated by an abundant myxoid matrix with hyalinised blood vessels. Immunohistochemical stains are often employed in difficult diagnostic cases purely based on its morphology. Intense S-100 protein positivity aid in the diagnosis.[1,2,5,9] There are several subtypes of schwannoma: cellular, pigmented, plexiform, and epithelioid.

The histological differential diagnosis of sinonasal schwannomas include lesions characterized by spindle cells that include the following: juvenile nasopharyngeal angiofibroma, neurofibroma, fibromyxoma, ectopic or secondary meningioma, glomangiopericytoma, leiomyomas, and solitary fibrous tumor.[2,8]

Juvenile nasopharyngeal angiofibroma, common in young male, composed of hyalinised vascular stroma consist of stellate and spindled fibroblasts with numerous mast cells. The vascular spaces are characterized by staghorn appearance with variable wall thickness and lack of elastic fibers. The cells show positive immunostaining with vimentin and beta-catenin with focal smooth muscle actin (SMA) positivity.[2,8]

Neurofibroma is characterized by proliferation of schwann cells, perineural cells, fibroblasts and myxoid matrix with scattered mast cells. Showing EMA and NF immunostaining positivity and focal positivity with S-100.[2,8]

Solitary fibrous tumor has hemangiopericytoma-like pattern, ovoid to fusiform spindle cells with indistinct cell borders, with haphazard arrangement or pattern less pattern along with proliferation of variable sized blood vessels and tumor cells show immunohistochemical positivity for CD34 and CD99.[2]

In ectopic or secondary meningioma, the tumor cells have a syncytial arrangement with a whorled pattern, pseudonuclear inclusions and psammoma bodies. EMA immunoreactivity can be helpful to exclude the entity.[8]

Fibromyxoma composed of bland spindle to stellate shape cells arranged in random to loose storiform and fascicular pattern with myxoid or collagenous stroma, prominent blood vessels and mast cells.

Glomangiopericytoma, distinctive spindle cell lesion of the sinonasal cavity, originate from modified perivascular myoid cells. It is comprised of densely packed uniform spindled to oval cells, little intervening collagen and staghorn capillary vessels with occasional hyalinised walls. The tumor cells show SMA, FXIIIa, vimentin positivity.[8,11]

Leiomyomas composed of spindled cells in intersecting fascicles, cigar-shaped nuclei, and perinuclear halos when the fascicles are visualized in cross-section. The cells show desmin, SMA, calponin, and H-caldesmon immunopositivity.[8]

Ancient change in schwannoma may be misdiagnosed as malignant peripheral nerve sheath tumor. The malignant peripheral nerve sheath tumor can be excluded by absence of fascicular growth, high mitotic activity and hypercellularity.

The treatment is complete surgical excision. Usually, head and neck schwannomas are encapsulated; however, some sinonasal schwannomas are not encapsulated, making *en bloc* tumor resection difficult. The surgical procedure depends on the tumor size, location, and invasion of adjacent structures.[1,5]

Conclusion

Schwannoma, a benign tumor, arise from schwann cells of myelin sheath; occur anywhere in the body. A well-known entity but rare in nasal and paranasal sinus area. Nasal septum is extremely rare site. Due to multiple differential diagnosis of soft tissue nasal mass and lack of definite imaging finding of schwannoma; the conclusive diagnosis is made only by histopathological examination. It is successfully treated by transnasal endoscopic excision.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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