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Case Report

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Schwannoma nasal cavity: a clinicopathological case report

Fathima Seles M.*, Padmakantha Srinivasan, Narmadha Ramadoss

Institute of Pathology, Madras Medical College, Chennai, Tamil Nadu, India

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***Correspondence:**
Dr. Fathima Seles M,
E-mail: dr.fathimaseles@gmail.com

ABSTRACT

Nasal cavity schwannomas are uncommon lesions, representing less than 4% of all head and neck schwannomas. They give rise to nonspecific symptoms such as nasal obstruction, epistaxis, and anosmia. Here we report a 18year old girl who presented with nasal discharge and recurrent epistaxis for four months.

Keywords: Schwannoma, Neurilemmoma, Neurioma, Nasal cavity, Benign nerve sheath tumor, Peripheral nerve sheath tumor, Histopathology, Clinicopathological

INTRODUCTION

Schwannomas are lesions that arise from the neural sheath of peripheral nerves, autonomic nerves, or cranial nerves.[1] Any nerve with a Schwann cell sheath may give origin to a schwannoma and so this neoplasm may develop in almost any part of the body.[2] Schwannoma commonly occur on the flexor aspect of extremities, posterior spinal nerve roots, mediastinum, retroperitoneum, cerebellopontine angle and rarely in head and neck region (25%).[3]

Nerve sheath tumors of the head and neck region mainly involve the eighth cranial nerve with only 4% occurring in the nasal cavity. Schwannomas of the nose and paranasal sinuses are discovered late in their clinical course because of their obscure anatomic location.[4]

CASE REPORT

A 18 year old girl presented with foul smelling nasal discharge for four months, nasal obstruction for two months associated with intermittent nasal bleeds. She also had post nasal drip and snoring. Anterior rhinoscopy showed septal deviation to the right and congested nasal mucosa. Posterior rhinoscopy and nasal endoscopy showed well defined globular mass with prominent vascular markings seen filling the entire right side of choana with right eustachian tube obstruction.

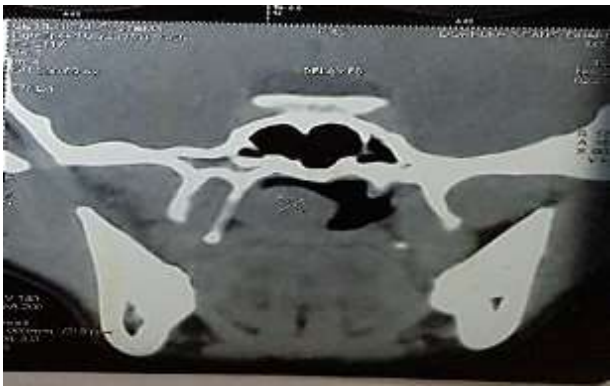

Figure 1: CT PNS showed a soft tissue mass lesion in the right Fossa of Rosenmuller, filling almost the entire right choana.

Paranasal sinus computed tomography (CT) scan showed mild deviation of the nasal septum to right with right concha bullosa polypoidal soft tissue mass lesion measuring 24×17 mm in the right nasopharynx arising

from fossa of Rosenmuller (Figure 1). On contrast showed minimal enhancement (Figure 2).

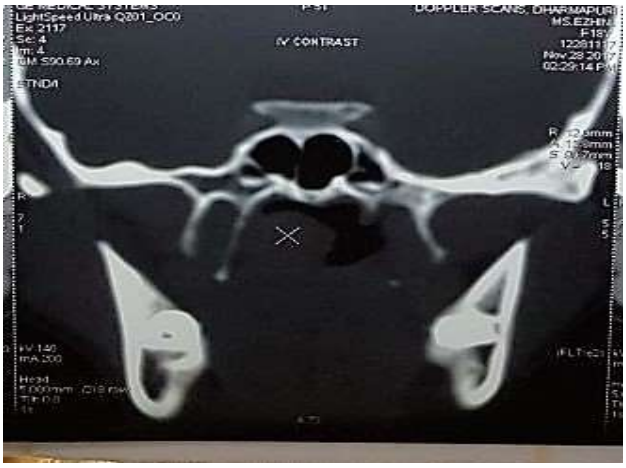

Figure 2: CT contrast showed minimal enhancement of the mass lesion.

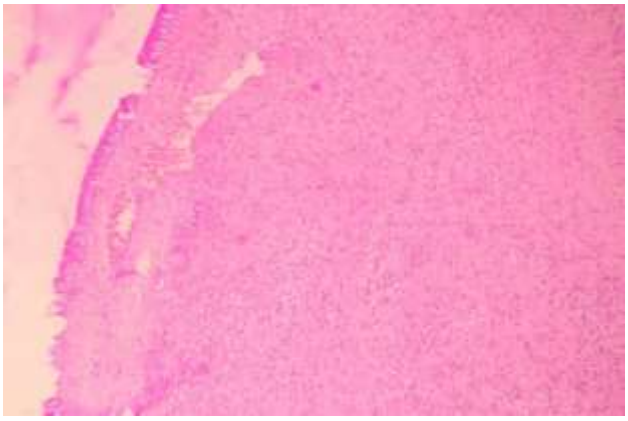

Figure 3: Respiratory epithelium with hypercellular (ANTONI A) and hypocellular (ANTONI B) areas (under 4X, H&E).

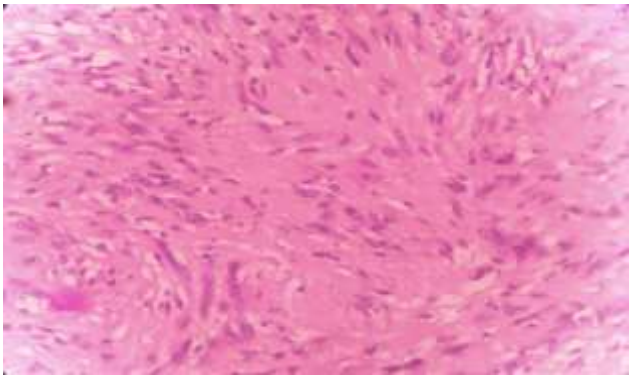

Figure 4: Spindle cells with nuclear palisading and Verocay body formation (under 40X, H&E).

Grossly grey brown grey white glistening soft tissue mass, firm in consistency, measuring 1×1 cm. Microscopically showed pseudostratified ciliated columnar epithelium with squamous metplasia and underlying unencapsulated lesion disposed in sheets and bundles with hypercellular (ANTONI A) and hypocellular (ANTONI B) areas (Figure 3). The cells are spindle shaped with moderate amount of eosinophilic cytoplasm, indistinct cell border, elongated vesicular nuclei. Many foci shows peripheral palisading with Verocay body formation (Figure 4). There are numerous thick and thin walled blood vessels, some showing hyalinization. Also seen are scattered histiocytes, lymphocytes and hemosiderin laden macrophages. Immunohistochemical staining revealed diffuse strong positivity for S100 (Figure 5).

DISCUSSION

Schwannomas are benign nerve sheath tumors arising from the Schwann cells in the peripheral nerve sheath that are derived from the neuroectoderm, first described by Verocay in 1908. In the year 1935, Stout coined the term Neurilemmoma believing that this tumor arose from cells of sheath of Schwann[5]. Since these tumors are schwannian in origin, it is suggested that the term schwannoma be used more freuquently.

Nasal schwannomas originate from the ophthalmic and maxillary branches of trigeminal nerve, may also arise from sphenopalatine ganglion, sensory inferior orbital nerve and other nerves in the nasal mucosa.[6]

Schwannomas are usually described as being encapsulated, the capsule is derived from the perineurium of the nerve of origin. Nasal schwannoma that derive from autonomic nervous system fibres, are devoid of perineural cells and therefore lack encapsulation.[7] Unencapsulation in nasal schwannoma probably explains the aggressive pattern compared to the schwannomas in other locations. The lack of encapsulation does not imply malignancy, but for the clinician the lack of encapsulation might make the tumor more difficult to define and extract completely.[7]

Symptoms are non specific and are the result of the mass effect. Patients may present with nasal obstruction, rhinorrhea or recurrent epistaxis. Facial swelling and pain are associated with paranasal sinus involvement.[8]

Computeristed tomography (CT scan) delineates the soft tissue tumor and bony invasion. After contrast administration, demonstrates central nonenhancing necrotic or cystic areas and peripheral enhancing neovascular areas of the tumor. The areas of inhomogeneity on CT may relate pathologically to confluent areas of hypocellularity adjacent to the hypercellular areas, therefore Schwannomas may have areas of mixed attenuation on CT.[8] MRI is used to better assess intracranial extension and showed the mass lesion to be T1-weighted and heterogenous increased T2-weighted signal intensity.[9] Although magnetic resonance imaging (MRI) is superior in defining soft tissue tumors,

CT offers better resolution of bony invasion. However as benign schwannoma can erode bone by pressure, bony erosion is not a criterion for malignancy.

Macroscopically, tumor is solitary, well demarcated with a oval, round or fusiform shape, grayish to yellowish in color, fleshy and shiny on cut surface.[10] Microscopically, Nasal schwannomas exhibit two architectural patterns, ANTONI A and ANTONI B in various proportions.[11] ANTONI A area is composed of an organized compact cellular stroma with elongated spindle cells and parallel rows of palisading nuclei forming verocay bodies. ANTONI B area is composed of disorganized loose myxoid stroma with few spindle cells and hyalinised blood vessels. Additionally shows intense nuclear and cytoplasmic immunostaining for S100 protein in both areas which differentiates schwannoma from other neural tumors.[12]

Differential diagnosis of schwannoma nasal cavity should include juvenile nasopharyngeal angiofibroma, solitary fibrous tumor, neurofibroma, fibromyxoma.[13]

Nasopharyngeal angiofibroma is common in male child, with intricate mixture of blood vessels and fibrous stroma.[14]

Solitary fibrous tumor has hemangiopericytoma-like pattern and alternation of cellular and fibrous areas. The tumor cells are strongly stained by CD34, CD8 and CD23.[15]

Neurofibroma is multiple, non encapsulated, formed by combined proliferation of all elements of peripheral nerve: axons, Schwann cells, fibroblasts and perineural cells, with axons traversing the tumor mass and more malignant potential.[16] Immunohistochemistry with S100 shows focal positivity.

Schwannomas do not recur if they are completely removed. In contrast to multiple neurofibromas (Von Recklingshausen's disease) schwannomas almost never undergo malignant transformation.[17]

CONCLUSION

Schwannoma arising from the nasal cavity is very rare and conclusive diagnosis is made only with histopathological examination. The possibility of schwannomas should be kept in mind when facing with soft tissue mass in nasal cavity. The autonomic nerve fibres of sinonasal mucosa are devoid of perineural cells, therefore they lack encapsulation which explains the aggressive growth pattern, local bone erosion, destruction and recurrence. The lack of encapsulation thus does not imply malignancy.

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