Glomus tumors are rare vascular neoplasms, typically occurring in the subungual region. Although the occurrence of these tumors in the head and neck area is extremely rare, they may occur in the trachea and the sinonasal tract. These tumors are often of a benign nature and can be cured through simple resection. They may be locally aggressive, so complete resection is important. Glomus tumors are thought to emanate from the normal glomus apparatus, a thermoregulator which is found in the dermis throughout the body. In this paper we present a rare case of an intranasal glomus tumor found in the nasal septum, which caused nasal obstruction and intermittent epistaxis. The tumor was surgically excised with resolution of symptoms.

**Key words:** glomus tumors, immunohistochemistry

**Introduction**

Glomus tumors are relatively uncommon benign mesenchymal neoplasms emanating from the normal glomus apparatus. They most commonly occur at distal extremities, particularly in the subungual region [1]. These tumors may also develop in sites where the normal glomus body may be sparse or even absent. Unusual locations [2,3,4,5] for the occurrence of glomus tumors include the chest wall, bone, breast, tongue, esophagus, stomach, liver, genitourinary tract, trachea and nose [6,7,8,9]. Typically diagnosed in adults, but may occur at any age, glomus tumors represent only about 1.6% of all soft tissue tumors. There is also an exceedingly rare malignant variant of glomus tumor which constitutes less than 1% of all glomus tumors. Glomus tumor of the nasal cavity is very rare and may be either asymptomatic or present with nasal obstruction, epistaxis and pain.

**Case report**

A 67 year-old woman was referred to our medical center with a complaint of a left nostril mass which has been gradually growing for approximately two years, causing nasal obstruction with occasional epistaxis but without any pain. Her medical history included hypertension, hypercholesterolemia and ductal carcinoma in situ of the breast followed by lumpectomy and Tamoxifen treatment.

Physical examination of the nose revealed a soft, blue-red, vascular mass originating in the left Kiesselbach...
plexus, measuring 0.5 cm in diameter and located 2 cm proximally to the nasal vestibule. Excisional biopsy was collected with minimal bleeding managed by local tamponade.

The histopathology sections demonstrated a portion of mucosal tissue covered by squamous epithelium with submucosal proliferation of monomorphic, mononuclear, round-to-cuboidal cells arranged in sheets surrounding capillary-sized vascular spaces in a slightly myxomatous stroma. No necrosis or mitotic figures were seen, Figure 1 and 2.

These cells stained positively with immunohistochemical stains for vimentin (Figure 3), smooth muscle actin (SMA) (Figure 4), muscle specific actin (MSA) (Figure 5) and were negative for desmin, keratin, S-100, melan A, chromogranin, synaptophysin, epithelial membrane antigen (EMA) and glial fibrillar acid protein (GFAP).
Case Presentation

The patient was under follow up for 2 years post-surgery and no recurrence of the tumor was detected.

Discussion

The glomus body is a unique form of dermal arteriovenous anastomosis contributing to thermal regulation, found in the reticular dermis throughout the body, mostly in the digits. Glomus tumors emanate from the arterial portion of the glomus body, or the Sucquet-Hoyer canal, which is the arteriovenous shunt that contributes to temperature regulation [4]. Although glomus tumors are thought to emanate from glomus cells, these tumors have been observed in extracutaneous locations that are not known to contain glomus cells. A possible explanation for this finding is that these tumors emanate from perivascular cells that can differentiate into glomus cells, which are modified smooth muscle cells. Clinically, the glomus tumor is a small red-blue nodule, usually less than 1 cm in diameter, associated with a long history of paroxysmal pain exacerbated by tactile stimulation and cold. Glomus tumors are divided into three histological sub-groups according to the relative part of glomus cells, vascular structures and smooth muscle tissue: solid glomus tumor (glomus tumor proper), glomangioma and glomangiomyoma [5]. The most common sub-group is the solid glomus tumor.

Glomus tumors are extremely rare and are defined as a large (less than 1 cm), deep lesion with very few mitotic figures and nuclear atypia. Microscopically, solid glomus tumors are well-bounded lesions consisting of capillary-sized vessels surrounded by sheets of glomus cells set in a hyalinized or myxoid stroma. Glomus cells are small, uniform and round with a globular nucleus at the center and a slightly eosinophilic cytoplasm. Each cell is surrounded by a basal lamina seen best on PAS or with Toluidin blue staining [5]. As to immunohistochemistry dyes, vimentin and muscle actin isoforms can be identified in nearly all glomus tumors. Desmin is much more varied.

Tumors of the nasal septum are rare [10]. There have been reported cases of primary olfactory neuroblastoma [11], primary meningoima [12], synovial sarcoma [13], chondrosarcoma [14], pleomorphic adenoma, lymphoma, squamous cell carcinoma and adenocarcinoma [10]. The clinical manifestations of these tumors are nasal obstruction with or without episodes of epistaxis. Hence a thorough investigation involving the collection of a biopsy should be carried out in all cases where a patient presents with a septal mass.

Glomus tumors at uncommon sites are often diagnosed late, causing patients to go undiagnosed or misdiagnosed for many years. Awareness of this diagnosis is important for the prevention of unnecessary delay in the treatment of asymptomatic patients suffering from epistaxis and pain.

References