Recurrent Epistaxis from an Intranasal Glomus Tumor: the 22nd Case Report

Somboon Keelawat MD*,
Prakobkiat Hirunwiwatkul MD**, Voranuch Thanakit MD*

** Department of Pathology, Faculty of Medicine, Chulalongkorn University*
*** Department of Otolaryngology, Faculty of Medicine, Chulalongkorn University*

The case of a 66-year-old Thai woman who was treated at King Chulalongkorn Memorial Hospital due to recurrent epistaxis for two years is presented. Her physical examination revealed a 0.7-cm tumor mass on the right nasal septum. A biopsy was subsequently done and the tissue was sent for pathological examination. The lesion was then diagnosed as "glomus tumor of the nasal septum". Histologically, the tumor was rather well-circumscribed and located in the submucosal region. The neoplastic cells were uniform, round to ovoid in shape and contained bland-looking, finely chromatic nuclei and moderate amount of cytoplasm with ill-defined cell borders. These cells were intervened by tortuous vascular structures. Immunohistochemically, the tumor cells were strongly reactive to smooth muscle actin and negative to cytokeratin and S-100 protein. A glomus tumor is rare in the sinonasal region. So far, there have been only 21 reported cases in the literature. Almost all cases of sinonasal glomus tumor are benign and are usually cured by complete excision.

Keywords : *Glomus tumor, Glomangioma and sinonasal tract*

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A glomus tumor is a benign neoplasm which derives from modified muscle cells of special arteriolovenular anastomoses, the so-called cutaneous glomus bodies[1], that functions in thermal regulation[2]. These tumors are common in the subungual regions of fingers and the lateral areas of the digits and palmar anastomosis where normal glomus bodies are located[2]. Less frequent sites in which the tumors have been described include lung, trachea, mediastinum, bone, stomach, oral mucosa, as well as cervix, labia and vagina[3]. Sinonasal glomus tumors are extremely rare. To our knowledge, only 21 cases [1,2,4-12] have been reported. The authors present herein the 22nd sinonasal glomus tumor.

Case Report

A 66-year-old Thai woman complained of recurrent epistaxis for two years but the condition was left untreated. One day before coming to the hospital, she experienced severe bleeding from her right nostril. Her physical examination revealed a 0.7-cm rubbery, smooth-surfaced mass on the right nasal septum without active bleeding. A biopsy was performed and suggested a vascular tumor. Excision of the mass was subsequently done.

Operative findings revealed a well-circumscribed exophytic reddish mass with contact bleeding, size 5x8 mm. The mass was found on the cartilagenous part of the right nasal septum.

Grossly, the mass was described as firm, grayish-white with multiple hemorrhagic spots. Histological examination revealed a rather well-circumscribed submucosal lesion(Fig. 1A) which was composed of uniform, round to ovoid cells with benign-looking, finely chromatic nuclei and moderate amphophilic or eosinophilic cytoplasms which showed ill-defined borders (Fig. 1B). Variable-sized tortuous vascular structures (Fig. 1C) with focal staghorn appearances resembling hemangiopericytoma were present among the aforementioned cellular component. The background stroma showed myxoid degeneration in some areas reminiscent of a mixed tumor or myoepithelioma of the salivary gland. Immu-

Correspondence to : Keelawat S, Department of Pathology, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand.

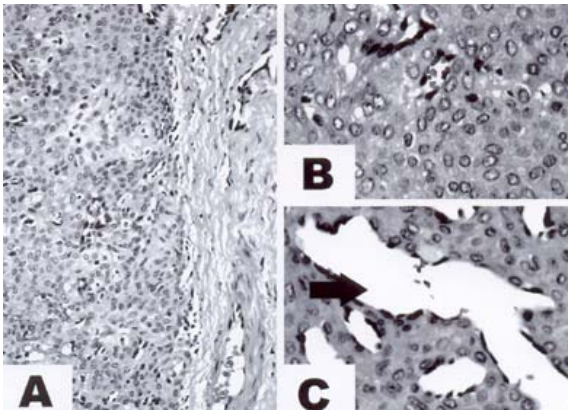

Fig. 1 Photomicrographs of the lesion
(A) The neoplasm shows a well circumscribed border (H&Ex200)
(B) The neoplastic cells appear uniform and contain round to ovoid, bland-looking and finely chromatic nuclei with ill-defined cytoplasm (H&E x 400).
(C) The tumor cells are intervened by tortuous vascular spaces (arrows) (H&E x 400)

nohistochemical studies showed tumor cells to be strongly reactive to smooth muscle actin and negative to S-100 protein and cytokeratin.

With the above features (morphological and immunohistochemical), "glomus tumor" was the most appropriate diagnosis.

Postoperatively, there was no bleeding or infection. The raw surface was healed completely in 2 weeks. No recurrence was found within 6 months after the excision.

Discussion

To many otolaryngologists, the term "glomus tumor" indicates a tumor of the paraganglion system which is commonly located in the middle ear. In fact, it should be noted that the term "glomus tumor of the middle ear" is a misnomer. Genuine glomus tumors derive from glomus bodies which histologically are completely different from the former. To avoid any possible confusion, glomus tumor of the middle ear should be called "paraganglioma"[13].

Due to the rarity of a glomus tumor in the nasal cavity, the lesion might not be missed when it is encountered at this location. Also, it may be histologically confused with other conditions such as olfactory neuroblastoma, hemangiopericytoma[2,3], myoepithelial tumor of salivary gland or lymphoid aggregate (especially in a small biopsy)[4].

Diagnosis of a glomus tumor is based primarily on its unique histological features plus some immunohistochemical findings. Classical histological appearance of the tumor shows a well-circumscribed lesion consisting of uniformly round, ovoid, polyhedral, or fusiform cells[14] with scant eosinophilic cytoplasm[5]. These cells tend to proliferate about tortuous and convoluted, narrow, or dilated vessels[14]. The background stromal component often shows myxoid change[15]. Histological variants include glomangioma and glomangiomyoma[15]. Immunohistochemically, glomus tumor cells are positive to vimentin and actin but negative to cytokeratin[15,16]. Desmin staining gives variable results[16]. They are focally positive for CD 34[2]. In addition, the neoplasm is also reactive to immunostaining for laminin and type IV collagen, the two constituents of basal lamina[16]. These materials outline each tumor cell resulting in chicken wire pattern of staining[16] Electron microscopic studies show typical smooth-muscle myofibrils that course into typical smooth-muscle "dense bodies" in the cytoplasm and at the plasma membrane[14,17].

Glomus tumor is distinguished from esthesioneuroblastoma by the absence of Homer-Wright rosettes. Furthermore, the cells of a glomus tumor contain more eosinophilic granular cytoplasms in contrast to esthesioneuroblastoma which is composed of densely packed small round cells with a scant amount of cytoplasms[2].

A hemangiopericytoma contains irregular staghorn-shaped vascular spaces and spindled tumor cells[2] which are uncommon in a glomus tumor. In contrast to a glomus tumor, a hemangiopericytoma usually does not stain to actin[2].

Myoepithelioma of the salivary gland, may be confused with a glomus tumor especially when focus is given on solid areas of the neoplasm (Fig. 1A). Immunohistochemical studies are helpful to distinguish between these two entities. Both tumors are positive for actin[2,4,15,18]. However, myoepithelioma is also reactive to cytokeratin and S-100 [17] while a glomus tumor stains negatively to them[1,2,4,15].

The clinical presentations for an intranasal glomus tumor include recurrent epistaxis, nasal obstruction, local pain or complaint of nasal mass[1,2,4-10]. Usually, the lesions are slow-growing with the duration of symptoms ranging from several months to years[1, 2, 4-10]. From all the cases with available details[1,2,4-10] described in the literature plus the current case, female patients outnumber males with the ratio of approximately 2:1. The patients' ages range from 9-81 years, with the average of about 53 years.

The tumors are located on the nasal septum in about half of the cases[1,2,4-10].

Most glomus tumors of the sinonasal region are benign[1,5,6,8,9]. They are usually cured by complete excision. However, local recurrences have been described in patients whose tumors have been incompletely excised[8]. On review of the literature, so far the authors found that there has been only one case of aggressive glomus tumor of the nasal region, reported by Hayes et al[4], which presented with six local recurrences within 18 years after complete removal on all occasions. The morphological appearances described in that report are not different from other cases. It seems that morphology alone is not helpful to predict the biological behavior of the tumor. There is evidence from a few studies[3, 4] suggesting that the absence of actin in a glomus tumor is correlated with a more aggressive biological behavior[3, 4]. In the case reported by Hayes et al[4], the neoplasm was also negative to smooth muscle actin.

The treatment of choice for a glomus tumor is complete surgical removal either by endonasal access or lateral rhinotomy depending on the size and the localization of the tumor mass[1]. Radiotherapy should only be employed if the tumor is inoperable[1].

In summary, the authors report a case of intranasal glomus tumor which is the 22nd case described in the literature. Although rare in this region, the neoplasm should be recognized and not to be confused with other tumors that show some histological similarities. Also, the authors would like to emphasize herein that a the glomus tumor of the middle ear is in fact a paraganglioma. To avoid any possible confusion, the term "glomus tumor of the middle ear" should be abandoned and replaced by "paraganglioma of the middle ear". Almost all glomus tumors in the sinonasal region are benign and they are usually cured by complete excision.

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อาการเลือดกำเดาไหลกลับเป็นซ้ำเนื่องจากเนื้องอกโกลมัสของผนังกั้นโพรงจมูก

สมบูรณ์ คีลาวัฒน์, ประกอบเกียรติ หิรัญวิวัฒน์กุล, วรนุช ธนากิจ

ได้รายงานผู้ป่วยหญิงไทย อายุ 66 ปี มารักษาที่โรงพยาบาลจุฬาลงกรณ์ด้วยอาการเลือดกำเดาไหลกลับเป็นซ้ำมาสองปี จากการตรวจร่างกายพบก้อนเนื้องอกขนาด 0.7 เซนติเมตรที่ผนังกั้นโพรงจมูกด้านขวา จึงได้ส่งชิ้นเนื้อตรวจทางพยาธิวิทยา ผลการตรวจพบว่าเป็นเนื้องอกโกลมัสของผนังกั้นโพรงจมูก จากการดูด้วยกล้องจุลทรรศน์ก้อนเนื้องอกมีขอบเขตชัดเจน อยู่ใต้ต่อชั้นเยื่อบุผิวโพรงจมูก เซลล์เนื้องอกมีรูปร่าง และขนาดใกล้เคียงกัน นิวเคลียสกลมหรือ เป็นรูปไข่ ภายในนิวเคลียสมีเม็ดโครมาตินละเอียด ซัยโตพลาสซึมมีปริมาณปานกลาง ขอบเขตของเซลล์เห็นไม่ค่อยชัด ในระหว่างเซลล์เนื้องอกเหล่านี้ มีโครงสร้างหลอดเลือดรูปร่างคดเคี้ยวแทรกอยู่ด้วย การย้อมพิเศษโดยวิธีอิมมูโนฮิสโตเคมี พบว่าเซลล์เนื้องอกเหล่านี้ ให้ผลบวกต่อ smooth muscle actin และให้ผลลบต่อโปรตีน S-100 และ cytokeratin เนื้องอกโกลมัสพบได้น้อยมากที่โพรงจมูก จากการทบทวนวารสารทางการแพทย์พบว่าทั่วโลกมีรายงานเพียง 21 รายเท่านั้น เนื้องอกโกลมัสเกือบทุกรายของโพรงจมูกเป็นเนื้องอกชนิดที่ไม่ร้ายแรง ส่วนใหญ่รักษาหายด้วยการผ่าตัดเอาก้อนเนื้องอกออก