

Submandibular Cavernous Hemangiomas with Multiple Phleboliths Masquerading as Sialolithiasis

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Vasoformative tumors (almost exclusively hemangiomas) are the most common lesions of the major salivary glands during infancy and early childhood. They are more common in the parotid gland but are particularly rare in the submandibular gland. Changes in blood flow dynamics within hemangiomas result in thrombus formation and phleboliths. Hemangiomas of the salivary glands in adults are histologically unlike those in infants, the former being characteristically of the cavernous variant. Most cavernous hemangiomas require surgery since they do not show a tendency to regress. A case of an adult man with cavernous hemangioma affecting the submandibular salivary gland that clinically simulated sialolithiasis is presented to alert surgeons to the possibility of such a lesion. We describe the clinical course and review the literature. [*J Chin Med Assoc* 2005;68(9):441–443]

Key Words: cavernous hemangioma, phlebolith, submandibular gland

Introduction

Hemangiomas are the most common lesions of the major salivary gland during infancy and early childhood.^{1,2} They occur predominantly in the parotid gland or adjacent structures, but they are very rarely reported in the submandibular area. These hemangiomas are frequently found in association with a lesion of the overlying skin.³ Changes in blood flow dynamics within hemangiomas result in thrombus formation and phleboliths.⁴ Although hemangioma is a relatively common lesion in the head and neck, it is rarely associated with phlebolith in the submandibular region. It is strikingly predominant in females,⁵ and has an observed left-sided preference.⁶ Due to the rarity of these tumors and unfamiliar presentation, inaccurate preoperative diagnosis and inappropriate treatment planning are common. We present an unusual case of cavernous hemangioma with multiple phleboliths affecting the submandibular salivary gland, and review the literature.

Case Report

A 65-year-old male visited the otolaryngological service at Taipei Municipal Yang-Ming Hospital, complaining of intermittent slightly painful swelling in his left neck for 5 years. The swelling was not exacerbated during eating. Physical examination revealed a 6.5 × 5.5-cm soft mass situated in the left submandibular region. The overlying skin was elevated, flesh-colored, and soft. The remainder of the examination was unremarkable. Computed tomography (CT) showed calcified foci within the enlarged left submandibular gland with minimal enhancement (Figure 1). Submandibular sialolithiasis was suspected. The full blood count, electrolytes, liver function tests, and blood glucose were all within normal limits.

The submandibular gland was surgically removed via an external approach and was sent for histopathologic examination. There was no unusual bleeding during surgery. The patient left the hospital on the fourth postoperative day after an uneventful recovery.

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E-mail: ccc0216@yahoo.com.tw • Received: December 13, 2004 • Accepted: May 31, 2005

Macroscopically, the specimen showed a submandibular gland intimately attached to a blood-filled spongy vascular lesion. The extraglandular lesion had 2 grayish-white nodules and an intact submandibular capsule. Microscopically, the submandibular gland itself was normal. The vascular lesion showed histopathologic features of a cavernous hemangioma: large irregular spaces lined by thin-walled endothelial cells. The lumina were filled with blood cells (Figure 2).

During the 3-month follow-up, the functional and cosmetic results were excellent.

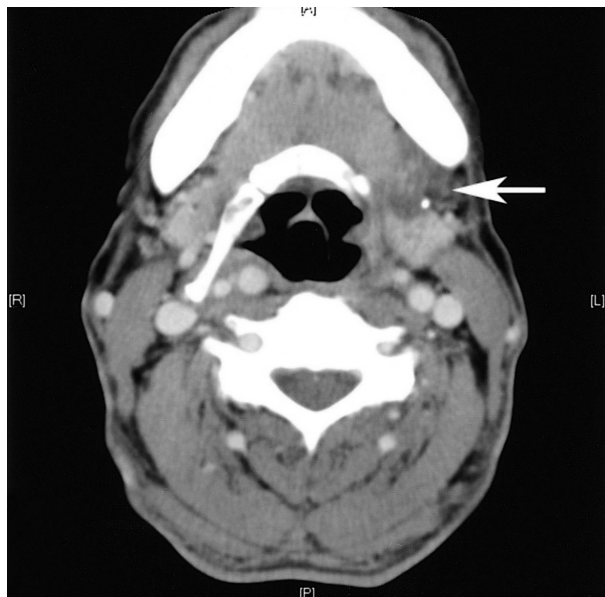


Figure 1. Axial contrast-enhanced computed tomography demonstrates the calcified foci and the left heterogeneous enlarged submandibular gland (arrow).

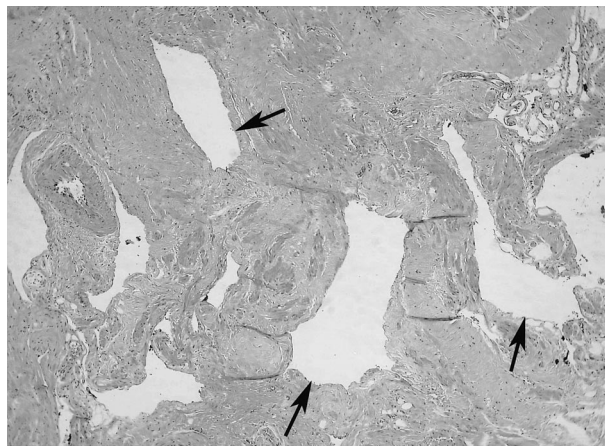


Figure 2. Microscopic views of the cavernous hemangioma reveal a dilated, thin-walled vascular space filled with blood and lined by endothelial cells (arrows) (hematoxylin & eosin, $\times 100$).

Discussion

Benign tumors of mesenchymal origin involving the major salivary glands often pose a difficulty in diagnosis. If the diagnosis is merely based on clinical features and history, the preoperative assessment may suggest a benign epithelial tumor of the salivary gland or sialolithiasis.⁵ An analysis of all the major salivary gland lesions recorded at the Armed Forces Institute of Pathology (AFIP) found that 1.4% were benign mesenchymal tumors.⁷ Of this group, 30% were hemangiomas. They were more common in the parotid gland (87.5% of all salivary gland hemangiomas) than in the submandibular gland (12%). Hemangiomas may arise from the gland proper,⁸ or by invasion of subcutaneous blood vessels into the gland structure. Hemangiomas occur twice as often in females as in males, and may fluctuate in size with pregnancy and menarche. These phenomena suggest that the endothelial cells may be quite responsive to circulating hormones.⁵ An observed left-sided preference has been noted.⁶ Of the cases registered with the AFIP, 8 hemangiomas occurred in the submandibular gland; 4 of these were in adults. Review of the literature revealed only 5 other cases of hemangiomas in the submandibular gland.⁹

Adult salivary gland hemangiomas are of the cavernous type, while infantile hemangiomas are usually capillary.¹⁰ The cavernous variant, as seen in this case, is characterized by dilated, thin-walled vascular spaces filled with blood and lined with flattened endothelial cells.

The submandibular triangle normally contains the submandibular gland, lymph nodes, and fat.¹¹ If a radiologic opacity is found in this area, sialolithiasis should be considered first. Cavernous hemangiomas with multiple phleboliths are very rare.¹²

It is currently believed that hemangiomas are benign and congenital neoplasms, which are usually undetected for long periods of time until sudden growth induces pain or cosmetic deformity. More than 90% of cases present before the fourth decade of life.¹³ The average age of patients presenting with hemangiomas is 10 years (range, 2 months to 74 years). However, cavernous hemangiomas typically occur in adolescents and adults.

Some patients may complain of a tender "cord" in the floor of the mouth. Frequently, all symptoms disappear spontaneously but recur a few months or years later. Presumably, the obstruction is relieved due to the calculus movement, partial disintegration, or the resolution of local inflammation.

Plain film X-ray demonstrates multiple calcified phleboliths in 2–3% of cases.⁵ On ultrasound, hemangiomas are heterogeneous hypoechoic lesions in which calcified phleboliths are identifiable.¹⁴ To differentiate parotid from extraparotid lesions, ultrasound has been largely supplanted by CT with contrast enhancement or in combination with sialography.¹⁵ CT dynamic scanning may show a tumor with enhancing quality similar to that of blood vessels, depending on the rate of blood flow through the hemangioma.¹⁶ With magnetic resonance imaging, lesions with isointensity to muscle on T1-weighted images and characteristic hyperintensity on T2-weighted images have been reported.¹⁶ Enlarged vessels may be seen as signal voids within and around the lesions. Nuclear medicine imaging with ^{99m}Tc red blood cell scintigraphy has also been reported to be useful in differentiating sialadenitis from hemangiomas.¹⁴

Cavernous hemangiomas tend to be larger and less well circumscribed than capillary and juvenile hemangiomas. They do not show a tendency to regress. Most cases require surgery. Other treatment methods include cryotherapy, laser surgery, vascular ligation, and corticosteroids.

In conclusion, there are 2 interesting points in this case. First, the submandibular area is a very uncommon site for a cavernous hemangioma. Second, the clinical and radiologic features often mislead physicians to make an erroneous diagnosis of sialolithiasis. Cavernous hemangioma with phleboliths should be included in the differential diagnosis of a swelling in the submandibular region.

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