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AV MALFORMATION DISGUISED AS SUBMANDIBULAR GLAND TUMOR: AN INTRAOPERATIVE IDENTIFICATION AND MANAGEMENT

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Abstract:

Submandibular gland swellings that present as a painless neck mass have extensive differential diagnoses. In this report we discuss case a that was taken up for submandibular gland excision and was incidentally found to be an arteriovenous malformation. We discuss our intraoperative management by including identifying features and surgical procedure for the benefit of other surgeons, who might find them in a similar position by encountering AV malformations intraoperatively in cases diagnosed as neoplasms.

Keywords: Arteriovenous Malformation, Submandibular gland, Surgical Excision, Salivary gland neoplasm

Introduction:

Slow-growing soft and fluctuant swelling in the submandibular region warrants a clinical diagnosis of benign lesion. Yet, painless neck swellings have a plethora of differential diagnosis as given below.

- Benign salivary gland tumours: pleomorphic adenoma, Warthins tumour

- Malignant salivary gland tumours: Adenoid Cystic Carcinoma, Mucoepidermoid Carcinoma
- Systemic Diseases: Sjogren's Syndrome, Mikulicz's Disease
- Malignant neck node: carcinoma metastases, lymphoma
- Granulomatous disease: Tuberculosis, Actinomycosis[1]
- Developmental: branchial cysts, laryngoceles, pharyngeal pouches
- Congenital: lymphangiomas, AV malformations, dermoid cyst, thyroglossal cysts
- Skin and subcutaneous tissue: sebaceous cyst, lipoma
- Thyroid swellings: multinodular goitre, solitary thyroid nodule
- Tumours of the parapharyngeal space: deep lobe parotid, chemodectoma[2]
- Reactive neck lymphadenopathy: tonsillitis, glandular fever, HIV

Ruling out each diagnosis requires careful history taking, understanding the age of onset and progression of disease. Diligent examination and imaging follow. Yet, one must be open minded when coming to the provisional diagnosis of any neck mass as the list is exhaustive.Arteriovenous Malformations (AVMs) are a congenital abnormality of blood vessels, characterized by an unorganized tangle of arteries and veins without capillaries between them. Accounting to a small percentage of 1.5% of all vascular anomalies, their occurrence in the oral and maxillofacial region is strikingly high (50%)[3]. We report a rare case of AVM within the right submandibular salivary gland of a 43-year-old female, with numerous phleboliths. This adds challenges to the already technique sensitive procedure of submandibular gland excision.

Case Report:

A 43 year old female patient reported to our OPD with swelling in right submandibular region since 2 months. The swelling was earlier inconspicuous and gradually increased in size over time. The patient complained that the swelling increased in size in the morning while brushing her teeth and on consuming sour and spicy food. The extent of the swelling was from parasymphysis to 1cm anterior to the angle of the mandible and was of 5 x 3 x 2 cm^3 approximately. There were no signs of any intraoral infection, lesion or ulcer. On palpation the swelling was firm with palpable calculi and submandibular lymph nodes and non pulsatile. The overlying skin was unaffected. USG reported calcification of right submandibular gland with reactive sub centimetric lymph nodes in bilateral mid cervical region.

Provisional diagnosis was pleomorphic adenoma or sialadenitis secondary to sialolith.

Patient was taken for Right Submandibular Gland Excision under General Anesthesia. Horizontal incision 2 cm below the inferior border of mandible along relaxed skin tension line was taken. Marginal mandibular branch of facial nerve was isolated and continuity preserved. Following flap elevation multiple phleboliths could be visualized in the surgical site along with profuse bleeding that required meticulous dissection and intermittent hemostasis. One of the most common complication of arteriovenous malformation is the formation of phleboliths within it. Anterior aspect of gland is mobilized posteriorly, exposing the mylohyoid muscle

and lingual nerve. Submandibular Ganglion is separated and ligated. Submandibular duct is traced anteriorly, ligated and transected. Posteriorly the feeder vessel was recognized branching from facial artery which was ligated permitting the complete removal of the gland from the submandibular space. Negative suction drain was secured and closure was done after ascertaining hemostasis. Healing and recovery of patient was uneventful. Histopathology report of the excised mass revealed thick and thin-walled vessels with organizational changes that was discrete from the parenchyma of submandibular gland and was thus confirmed as arteriovenous malformation.

Figure 1: Extraoral presentation: Submandibular swelling on right side as seen in [A] frontal and [B] worms view photographs.

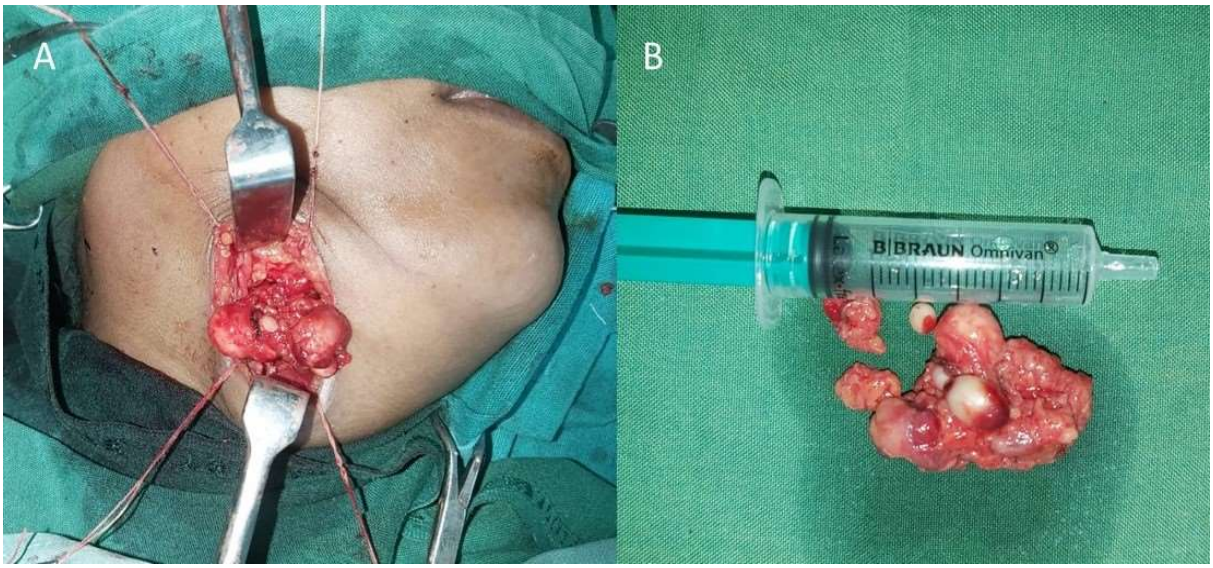

Figure 2: Intraoperative photographs [A] Right submandibular salivary gland superficial lobe in situ associated with multiple phleboliths [B] Primary resected specimen of size 5cm in greatest dimension along with multiple phleboliths.

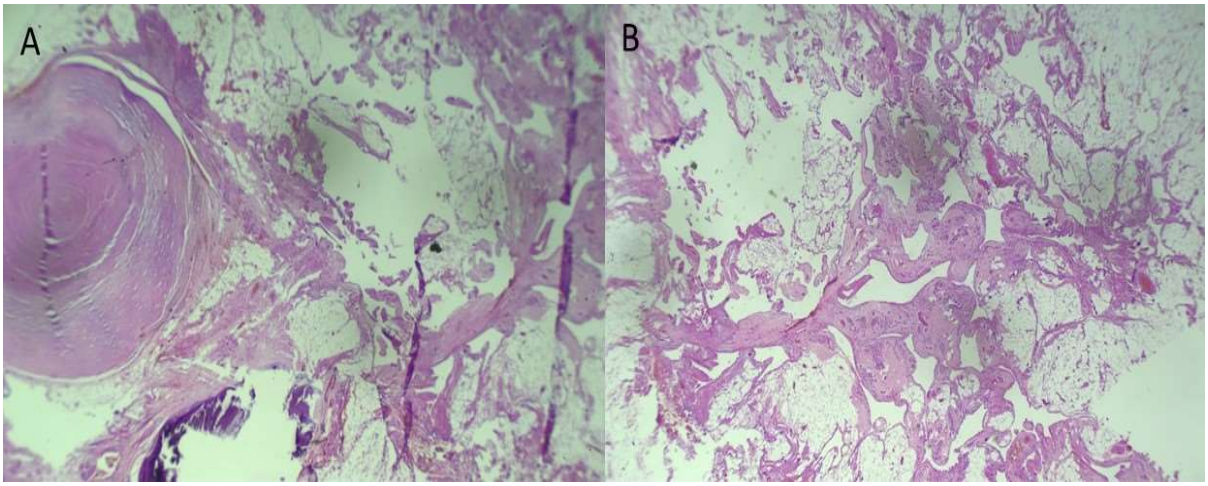

Figure 3: Microscopic Photographs of slides of resected specimen with H & E stained 2X magnification [A] biopsy showing calcified body in association with thin walled vessels (Phlebolith) [B] biopsy showing thick and thin walled vessels with organizational changes.

Discussion:

Arteriovenous malformations (AVMs) are a result of anomalous angiogenesis and are composed of a feeder arteriole and a draining vein. Between them lies a network devoid of the normal capillary framework, that are unorganized and hyperemic[4]. These vessels can vary in size, fragility and can often be pulsatile[3]. Vascular anomalies lead to local complications (including bleeding, infection, obstruction, pain, thrombosis, ulceration, and destroyed anatomic structures) and can also cause general complications such as congestive heart failure, disseminated intravascular coagulation, pulmonary embolism, thrombocytopenia, and sepsis[5]. The International Society for the Study of Vascular Anomalies (ISSVA) recently adopted a classification scheme, clearly separating vascular tumours (hemangiomas of different types) which result from active cell proliferation, from vascular malformations, which are inborn defects in vascular morphogenesis. Vascular malformations can be classified according to the vessel(s) types they are composed of, separating the malformations of vascular trunks from tissular malformations which are more intimately embedded in the surrounding tissues[6]. AVMs in the submandibular region are relatively rare, but they can cause significant symptoms such as pain, swelling, and inadvertent bleeding. They cause significant haemorrhage and cosmetic defect. The most common problem associated with vascular anomalies is psychological distress related to disfigurement as well as functional defects, as many lesions affect the head and neck. Submandibular salivary gland as a subsite for AVMs is so rare that their exact incidence and prevalence are not well established[7]. When planning surgery for any vascular malformation to be present within submandibular gland, a doppler test is necessary to ascertain the arteriovenous shunt and differentiate between low or high resistance and low or high velocity of flow. Arteriovenous malformations (AVM) of the head

and neck are quite rare in contrast to low-flow vascular anomalies. Treatment of these high-flow vascular anomalies is hazardous and has a predictably high incidence of recurrence if not managed correctly[8]. In cases where AVMs masquerade as other tumors this is the biggest drawback. Hence, being prepared for bleeding and hemorrhage must be practiced universally for all surgical excisions. Electrocautery with bipolar pens, ligaclips and ties along with arrangements for volume replacement and blood transfusion are necessary.

As mentioned earlier, another finding that is occasionally seen in the salivary gland region is phleboliths, which are small calcified deposits within the veins. The presence of phleboliths is often an incidental finding on imaging studies, and they are generally considered benign and asymptomatic. However, several studies have reported a higher incidence of phleboliths in patients with AVMs of the salivary gland region, suggesting a potential link between the two conditions[9].

Arteriovenous malformation (AVM) of the submandibular salivary gland can lead to complications including hemorrhage, infection, and nerve damage. Treatment options include embolization and surgical excision. The common risks associated with submandibular gland excision are facial nerve palsy, salivary fistula, and recurrence[10]. Due to the added risk of uncontrolled bleeding owing to the element of vascular malformation all hemostatic measures must be in place as a prophylactic precaution. The functional and cosmetic outcome of such cases must outweigh the risk of surgery.

Conclusion:

Arteriovenous malformation is a rare cause of submandibular gland swelling but most not be ruled out. Timely assessment of incidental findings such as phleboliths and being prepared to manage bleeding intraoperatively and hemorrhage associated with vascular lesions is a deciding factor in such cases. Decision-making regarding management should involve careful consideration of the risks and benefits of each option, taking into account the specific characteristics of the malformation and the patient. Complications and risks of treating this rare disease should be weighed against the benefit of symptom relief for the patient.

References

[1] S. Saxena, S. Kumar, and J. Kharbanda, “Swelling of the submandibular gland region: Cystic or neoplastic entity?,” J. Oral Maxillofac. Pathol. JOMFP, vol. 23, no. 3, pp. 468–471, 2019, doi: 10.4103/jomfp.JOMFP_155_19.

[2] N. Roland and P. J. Bradley, “Neck Swellings,” BMJ, vol. 348, p. bmj.g1078, Jan. 2014, doi: 10.1136/bmj.g1078.

[3] C. Fowell, R. Jones, H. Nishikawa, and A. Monaghan, “Arteriovenous malformations of the head and neck: current concepts in management,” Br. J. Oral Maxillofac. Surg., vol. 54, no. 5, pp. 482–487, Jun. 2016, doi: 10.1016/j.bjoms.2016.01.034.

[4] K. Schimmel et al., "Arteriovenous Malformations—Current Understanding of the Pathogenesis with Implications for Treatment," Int. J. Mol. Sci., vol. 22, no. 16, p. 9037, Aug. 2021, doi: 10.3390/ijms22169037.

[5] A. K. Greene, "Vascular anomalies: current overview of the field," Clin. Plast. Surg., vol. 38, no. 1, pp. 1–5, Jan. 2011, doi: 10.1016/j.cps.2010.08.004.

[6] M. Wassef, R. Vanwijck, P. Clapuyt, L. Boon, and G. Magalon, "[Vascular tumours and malformations, classification, pathology and imaging]," Ann. Chir. Plast. Esthet., vol. 51, no. 4–5, pp. 263–281, 2006, doi: 10.1016/j.anplas.2006.07.017.

[7] J. C. Burns, J. Julian, and J. Alexander, "Arteriovenous malformation of the submandibular gland," J. Oral Maxillofac. Surg., vol. 43, no. 4, pp. 294–296, Apr. 1985, doi: 10.1016/0278-2391(85)90292-7.

[8] M. W. Erdmann, J. E. Jackson, D. M. Davies, and D. J. Allison, "Multidisciplinary approach to the management of head and neck arteriovenous malformations.," Ann. R. Coll. Surg. Engl., vol. 77, no. 1, pp. 53–59, Jan. 1995.

[9] S. Hammer, F. Zeman, C. Fellner, W. A. Wohlgemuth, and W. Uller, "Venous Malformations: Phleboliths Correlate With the Presence of Arteriovenous Microshunts," Am. J. Roentgenol., vol. 211, no. 6, pp. 1390–1396, Dec. 2018, doi: 10.2214/AJR.18.19703.

[10] H. Boyes, A. Jones, and L. Cheng, "Arteriovenous vascular malformation of the submandibular gland masquerading as Küttner's tumour," BMJ Case Rep., vol. 12, no. 12, p. e233476, Dec. 2019, doi: 10.1136/bcr-2019-233476.