Submandibular sialoangiolipoma: A rare hamartomatous lesion causing diagnostic dialemma

Sir,

An eighteen years male presented with swelling in right side of neck of three years duration. The swelling was gradually increasing in size associated with localized discomfort. General physical examination was unremarkable. Local examination revealed a large 4 × 2 cm, soft, euthermic, non-tender swelling in right submandibular region. The swelling was non-compressible, non-pulsatile with fluctuation and transillumination tests being negative. Routine laboratory investigations were within normal limits. Ultrasound neck showed well defined hypoechoic mass measuring 2.8 × 1.6 cm in submandibular region abutting the medial border of right submandibular gland. With a preoperative diagnosis of AV malformation in right submandibular region, the patient was taken up for excision of the swelling under local anesthesia. Grossly, we received an irregular, rough, brownish tissue partly covered with skin measuring 4.5 × 2 × 1.5 cm. On microscopic examination, the tissue showed varied histomorphological features Composed of haphazardly arranged fibrous tissue, skeletal muscle fibers, mature adipose tissue, blood vessels of varying calibers and ducts of salivary gland. The tissue also showed a focus

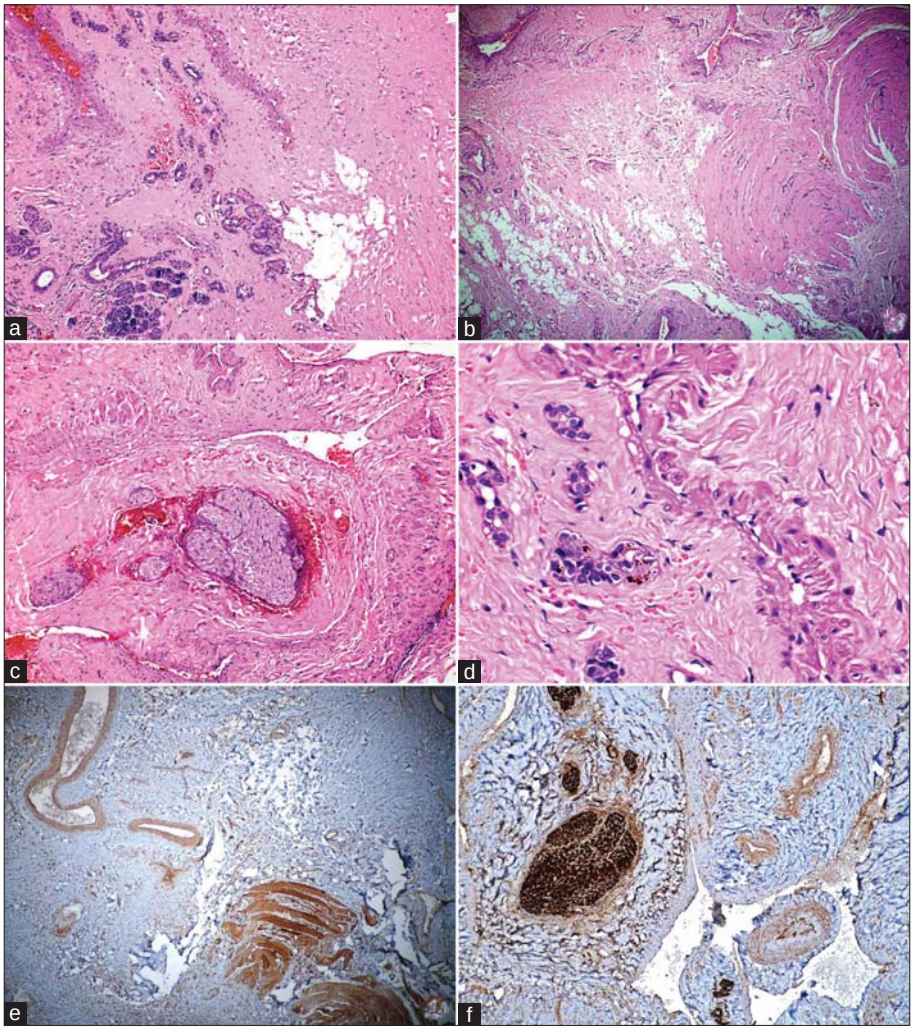

Figure 1: (a) Low power photomicrograph showing myriad structures like abnormal blood vessels, adipose tissue and ducts of salivary glands in a fibrous background (H and E, ×100). (b) Low power photomicrograph demonstrating the skeletal muscle fibres in addition to fibroadipose tissue and blood vessels (H and E, ×100). (c) High power photomicrograph showing a nerve bundle within the vessel wall (H and E, ×40). (d) High power photomicrograph showing salivary gland ducts adjacent to a large blood vessel (H and E, ×40). (e) Photomicrograph demonstrating myogenin immunostain highlighting the skeletal muscle fibers as well as smooth muscle fibers in the vessel wall (Myogenin immunostain, ×100). (f) Photomicrograph demonstrating S-100 positivity in the nerve bundle within the vessel wall (S-100 immunostain, ×40)

of nerve bundle within the vessel wall [Figure 1a-d]. Immunostains for myogenin [Figure 1e] highlighted the skeletal muscle bundle whereas S-100 [Figure 1f] highlighted the nerve bundle and adipocytes. CD34 immunostain highlighted the endothelial cells of the vascular component of the tumor and cytokeratin (CK) immunostain stained the salivary gland ducts. With the above histomorphological features, a diagnosis of mesenchymal hamartoma of the submandibular gland was offered. Six months later, the patient did not report any adverse outcome.

Mesenchymal tumors of the salivary glands occur most frequently in parotid gland and account for 2-5% of all salivary gland tumors.[1] These comprise the angiomas, schwannomas, neurofibromas, lipomas and rarely hibernomas, lipoblastoma and pleomorphic/spindle cell lipomas.[1] Combined mesenchymal tumors like sialolipoma, lipoadenomas, angiolipomas have been described only exceptionally in salivary glands with most of the cases being reported in parotid gland.[1-4] Only four cases of sialolipomas have been described till now in the submandibular gland with three occurring in adults and one in a child.[4] However, to the best of our knowledge, no case of submandibular gland hamartoma comprising such myriad structures including salivary gland ducts, skeletal muscle fibers and nerve within a vessel wall has been described in literature. One similar case has been described recently in the parotid gland and has been designated as sialoangiolipoma. [1] It was a tumor composed of normal looking acinar and ductul structures along with adipocytes and a prominent vascular component. Another report described similar lesions in the tongue containing salivary gland structures along with mesenchymal components.[5] However, the current case differs in that it lacks a lobular arrangement and apart from the above mentioned components, it had skeletal muscle fibers, a nerve bundle and only a minor component of salivary glands tissue in the form of a few ducts in fibrous stroma. Also it has occurred in a young adult male whereas the above mentioned cases occurred in children of female gender with some occurring even in newborns.[1,5] Hamartomas are tumor like malformations characterized by abnormal mixture of tissue indigenous to the part which grows with the body until maturity of the tissue is attained. Accordingly, our case presented at a young age with a very slowly growing mass (over 3 years) and with characteristic histological picture comprising admixtures of tissue indigenous to the part. The biological behavior in this case is anticipated to be completely benign in accordance to other hamartomatous condition elsewhere in the body and surgical excision seems to be curative. [1,5,6] Since, common recurrent cytogenetic abnormalities involving chromosome 12 at multiple aberration region (MAR) have been reported in hamartomas of the breast, it may be worthwhile to investigate this genetic aberration in the present tumor.[6] To conclude, we recommend the consideration of hamartomatous lesion in differential diagnosis of mass lesion of salivary gland and propose a larger study to evaluate the role of recurrent genetic abnormalities in hamartomatous lesion of the salivary gland.

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