CASE REPORT

Myxoglobulosis of lower lip: Report of two cases

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ABSTRACT

In this paper, we present two case reports of myxoglobulosis, in a 24-year-old female and a 40-year-old male patient who came to our hospital with a chief complaint of painless swelling of the lower lip of approximately 6 months duration. A study of two case reports has been given here. In these patients, histological examination of the surgically excised tissue was carried out. Histopathological examination showed an extravasation mucocele with the lumen exhibiting unique globular organizations of mucin surrounded by granulation tissue capsule and lacking an epithelial lining. Our two cases are possibly an analogue of myxoglobulosis, a rare variant of the appendical mucocele. Thus, though rare, the possibility of occurrence of myxoglobulosis in cystic lesions of the lip should be considered. The prognosis is regarded better as compared to the other types of mucoceles with low recurrence rate due to good host response and globular organizations of mucin. However, follow- up of these cases and more such cases is required to confirm the prognosis. The need for study of many such cases to confirm the etiology, pathogenesis and biologic nature of this variant is being felt.

Key words: Extravasation mucocele, lower lip, myxoglobulosis

INTRODUCTION

Myxoglobulosis, a term coined by von Hansemann in 1914, is described as a variant of mucocele. It is associated with the formation of opaque globules varying in diameter from 0.1 cm - 1.0 cm and lying in a clear mucous.[1] It was first described by Lapham in 1897. In the oral cavity, two types of mucoceles are recognized, mucous retention cyst and extravasation mucoceles. Mucous retention cyst is a true cyst, pathogenesis of which can be due to ductal dilatation, secondary to ductal obstruction, leading to increased intraluminal pressure.[2]

Extravasation mucocele is a common lesion of the oral mucosa that results from rupture of a salivary gland duct, often the result of local trauma, and spillage of mucin into the surrounding connective tissue.[2] It is most frequently seen in association with the minor salivary glands with a site predilection for the lower lip.[3] The lumen contains extravasated mucin and is surrounded by granulation tissue wall with no epithelial lining. Our present case report reveals extravasation mucocele with unique intraluminal globules resembling those usually seen in appendix – Myxoglobulosis, a rare variant of mucocele, of the lower lip.

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CASE REPORTS

Case 1

A 24-year-old female patient presented an asymptomatic swelling of lower lip which had gradually increased over a period of 6 months and measured about 1 cm × 2 cm. The swelling was dome-shaped, smooth-surfaced, soft in consistency and without discharge of any fluid. Her medical records did not show any relevant systemic findings. A provisional diagnosis of mucocele was done. The lesion was surgically excised under local anesthesia. Histopathological examination of the excised tissue revealed a well demarcated cystic lumen in the sub-mucosa, surrounded by compressed granulation tissue forming the wall of the lumen with overlying atrophic epithelium [Figure 1]. The cystic lumen showed a few muciphages and numerous globules (approximately 100 in number). The size of each globule varied, ranging from small to large (88 µm to 416 µm). The shape of most globules was predominantly polygonal while a few were round. Most of the globules were detached from each other due to their globular nature. Each globule exhibited a central, eosinophilic, amorphous and acellular core, surrounded by mucinous substance [Figures 2 and 3]. The periphery of a few globules showed a layer of viable flattened cells. There was

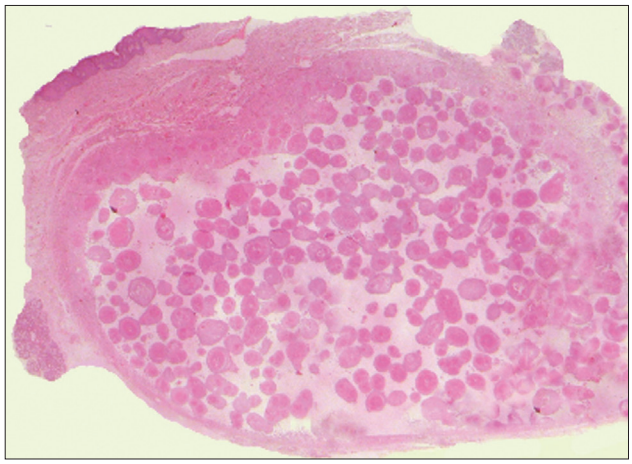

Figure 1: Photomicrograph showing cystic lumen in the submucosa with numerous globules, surrounded by granulation tissue wall and overlying epithelium (H and E stain, 15×)

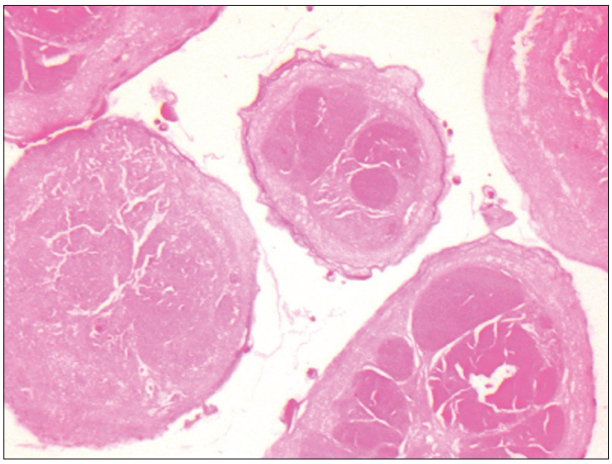

Figure 2: Photomicrograph showing cystic lumen with globules having central eosinophilic, amorphous and acellular core surrounded by mucinous substance (H and E stain, 200×)

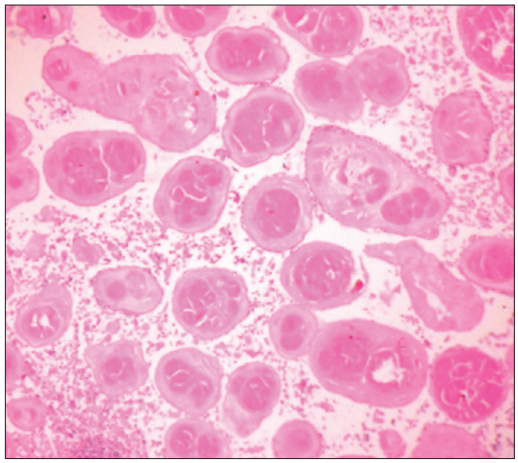

Figure 3: Photomicrograph showing cystic lumen, containing numerous globular organizations, spherical to polygonal in shape, with central eosinophilic, amorphous and acellular core surrounded by mucinous substance (H and E stain, 100×)

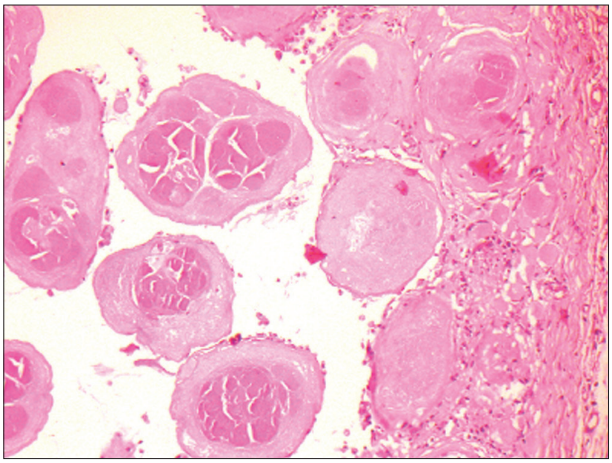

Figure 4: Photomicrograph showing some peripheral globules of the cystic lumen in contact with surrounding granulation tissue wall, and some within the granulation tissue wall. The globules within the granulation tissue wall are more cellular (H and E stain, 100×)

no evidence of calcification in these globules. The peripheral globules of the cystic lumen were in contact with the surrounding compressed granulation tissue wall, while some globules were also seen within the granulation tissue wall and the latter were more cellular compared to the centrally located globules in the lumen [Figure 4]. The globules showed negative results for periodic acid Schiff and Alcian blue staining. The granulation tissue forming the cystic wall was vascular, fibrous and cellular, comprising of fibroblasts, endothelial cells and chronic inflammatory cells, of which, muciphages were predominant. There were small mucinous pools surrounded by muciphages in the granulation tissue wall. The adjacent minor mucous salivary glands showed varying stages of degeneration like loss of architecture of acini, and mild chronic sialoadenitis [Figure 5].

Case 2

A 40-year-old male patient reported to our hospital with a chief complaint of painless swelling of lower lip near the right angle of mouth of 5 months' duration with a history of local trauma. No relevant medical history was elicited. On clinical examination, the swelling measured approximately 2 cm × 1.5 cm in size, was pale in color, sessile, dome-shaped, soft in consistency, smooth-surfaced and fluctuant. A provisional diagnosis of extravasation mucocele was done. Treatment included surgical excision of the lesion under local anesthesia and its submission for routine processing. Histopathological examination of the tissue revealed a cystic lumen in the submucosa containing mucin, few muciphages and a few unique mucinous globular organizations. These globules were spherical to polyhedral in shape, 5 in number, with each globule having a central, amorphous, acellular area surrounded by mucin. In 3 of these globules, peripheral cellular rimming with fibroblasts and muciphages was evident. Two of these globules were connected to the surrounding granulation tissue wall that formed the capsule. The granulation tissue was highly cellular, comprising of chronic inflammatory

cells (lymphocytes, macrophages and muciphages), plump fibroblasts, proliferating endothelial cells and numerous dilated and congested vascular channels [Figures 6 and 7]. Numerous mucin pooled areas were also evident. The cyst lacked epithelial lining. The overlying epithelium was orthokeratinized, stratified and squamous epithelium.

DISCUSSION

Mucoceles are the common non-neoplastic lesions of the oral minor salivary glands, of which, the majority are mucous extravasation cysts. Mucoceles develop due to the disruption of the flow of saliva from the secretory apparatus of the salivary glands. The lesions are most often associated with extravasation of mucus into the adjacent soft tissues caused by a traumatic ductal insult.[3]

In our present cases, the clinical features and histopathological findings are suggestive of an extravasation mucocele. Although diagnosis of extravasation mucoceles is non-challenging to the pathologists, the intraluminal globular organizations of mucin surrounded by the granulation tissue capsule are histologically very unusual.

The etiology and pathogenesis of these globular organizations are still unclear. Probstein and Lassar stated that in myxoglobulosis of the intestine or appendix, the speculative etiological factors are bacteria and necrotic epithelial debris which may act as nidus for concentric deposition of mucin.[4] Lubin and Berle proposed that the core of the globules' represents an organizing mass of mucin by granulation tissue originated from the appendical wall. These globules then break off by mechanical contractions and undergo necrosis.[1] Li *et al.* stated that the globules represent an attempt to organize the mucin by the granulation tissue capsule, which is then extruded into the lumen due to chronic frictional or mechanical forces.[5] Ide and Kusama stated that localized changes in the microenvironment may act as predisposing factors for formation of these florid globules. Myxoglobulosis of salivary mucocele probably occurs as a consequence of exuberant reparative process of the capsular granulation tissue wall in response to intraluminal accumulation of mucin.[6] This appears to be clearly the cause in our case reports also.

Lubin and Berle reported 2 cases of myxoglobulosis of the appendix, which is a rare variant of extravasation mucocele. Histologically, the lesions consist of intraluminal globules which measure about 0.2 cm to 1.0 cm in diameter, and consist of granulation tissue, mucin, varying degrees of secondary focal calcifications and cholesterol clefts.[1] In our present cases, the globules are similar to myxoglobulosis of appendix, except for their smaller size (ranging from 88 µm to 416 µm), absence of calcification and cholesterol clefts.

The globules in both of our cases are almost similar in size, shape and contents, but the number of globules seen in case 1 are more and the surrounding granulation tissue wall is moderately cellular and vascular. But in our case 2, the

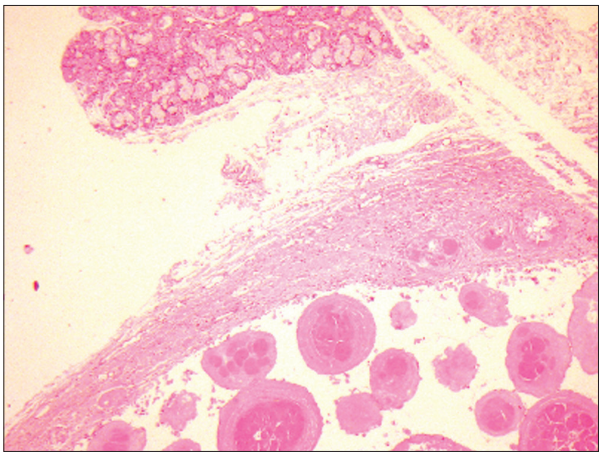

Figure 5: Photomicrograph showing cystic lumen, containing globular organizations surrounded by the granulation tissue wall, and minor mucous salivary glands showing varying stages of degeneration like loss of architecture of acini, and mild chronic sialoadenitis (H and E stain, 100×)

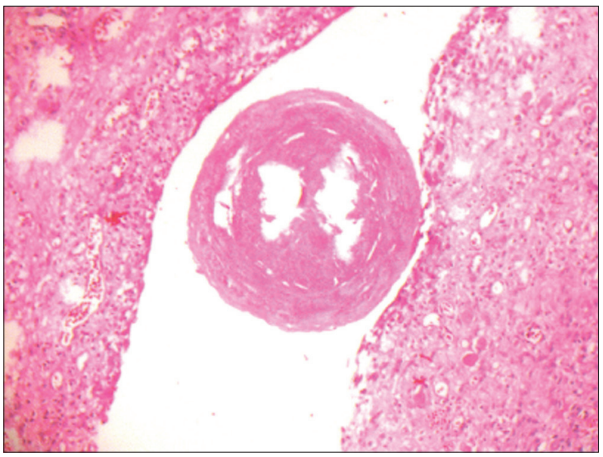

Figure 6: Photomicrograph showing cystic lumen containing globular organization that are spherical with surrounding cellular and vascular granulation tissue wall (H and E stain, 100×)

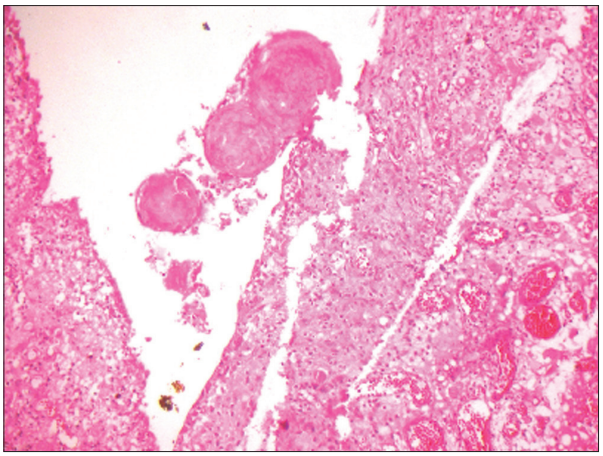

Figure 7: Photomicrograph showing cystic lumen containing globular organizations' in contact with surrounding granulation tissue wall (H and E stain, 100×)

globules are fewer in number and the surrounding granulation tissue wall is highly cellular and vascular.

In the present case reports, the globules in the granulation tissue wall are cellular but the globules in the lumen are acellular, which is in accordance with KA Shah, who stated that it is a time related phenomenon, with earlier globules being more cellular.[7]

We conclude that these present cases represent a unique organization of mucin by granulation tissue wall as an exaggerated reparative response and release of the globules into the lumen due to chronic mechanical forces. These globules in the lumen undergo necrosis due to absence of vasculature.

A somewhat similar analogy is seen in collagenous spherulosis that is frequently seen in benign and malignant salivary gland tumors, proliferative lesions of breast ductal epithelium, chondroid syringomas and schwannomas.[8] Differentiating collagenous spherulosis from myxoglobulosis is comparatively easy, as the globules in collagenous spherulosis are seen at the tissue level, are myoepithelial in origin, acellular in nature, and almost always seen in association with epithelial and myoepithelial cells.[9] They are generally positive for collagen, p63 and basement membrane, and generally negative for mucin stains,[10] whereas globules in myxoglobulosis are seen mostly in the lumen, are hyalinized, discrete, of variable cellularity and associated with granulation tissue wall. Their origin is from connective tissue and stains positive for acidic mucin and periodic acid Schiff stain.[5] Thus, it can be said that collagenous spherulosis and myxoglobulosis are the only morphologically related reaction patterns.[8]

Prognosis of this variant of extravasation mucocele will be better than other types of mucoceles with low recurrence rate due to good host response and globular organizations of mucin. However, follow-up of these cases and more such cases is required to confirm the prognosis. Study of many cases is needed to confirm the etiology, pathogenesis and biologic nature of this variant.

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