A 22-year-old male patient presented with a chief complaint of swelling in the left side of the face in front of the ear since 5 years. The swelling was slow-growing but showed a rapid growth since last 2 months. On extraoral examination, the lesion was extending superiorly 0.5 cm in front of the pinna of the ear, inferiorly 1 cm below the inferior border of mandible and 2 cm anterior to the angle of the mandible [Figure 1]. The swelling was ovoid with well-defined borders, soft, fluctuant in nature and was not compressible. The lesion was surgically excised and the specimen received was soft, large cystic sac (5 cm × 4 cm in diameter) with external pebbly surface. The cut surface showed multiple blood-filled macrocysts with intersecting thick septae [Figure 2].

**INVESTIGATIONS**

**Ultrasonography**

Ultrasonography revealed a large cystic mass in the soft tissue of the left cheek region filled with turbid, hemorrhagic fluid, measuring 4.5 cm × 2.5 cm in diameter with septae in between. No extension or involvement of parotid gland was seen. Small, enlarged nodes were also seen in upper part of the neck. Overall findings were suggestive of cavernous hemangioma. Fine needle aspiration cytology aspirate was pale yellow and the smear showed macrophages on a hemorrhagic background, no malignant cells were seen.

**Provisional diagnosis**

- Cavernous hemangioma
- Lymphangioma.

**HISTOPATHOLOGY**

Hematoxylin and eosin stained section showed multiple large irregular cystic luminal spaces. The cystic spaces were lined by widely spaced thin endothelial cells [Figure 3]. The stroma supporting the cystic spaces was dense fibrocellular with proliferation of small variable size vascular channels filled with lymphocytes and occasional red blood cells [Figures 4 and 5]. Also there were lymphocyte aggregates [Figure 6], muscle bundles [Figure 7] and adipose tissue at the periphery.

**Final diagnosis**

Cystic lymphangioma.
Cystic lymphangioma

Figure 2: The excised specimen in toto. Inset showing multiple macrocysts with intersecting thick septae.

Figure 3: Multiple large irregular cystic luminal spaces (H&E stain, x40).

Figure 4: Cystic lumen lined by widely spaced thin endothelial cells (H&E stain, x100).

Figure 5: Supporting fibrocellular stroma with proliferation of small variable size vascular channels filled with lymphocytes and occasional red blood cells (H&E stain, x100).

Figure 6: Lymphocyte aggregates within the stroma (H&E stain, x100).

Figure 7: Dense fibrocellular stroma supporting the cystic spaces with muscle bundles (H&E stain, x100).
DISCUSSION

As with hemangiomas, it is often difficult to state whether lymphangiomas are true neoplasms, hamartomas or lymphangiectasias. In fact, this distinction is of little practical value because they are all benign lesions and therapy is largely dictated by their location and clinical extent. Lymphangiomas are mostly regarded as hamartomas arising from sequestration of lymphatic tissue that fails to communicate normally with lymphatic channel. Lymphangiomas are most common in the neck, where they typically lie in the supraclavicular fossa of the posterior cervical triangle or extend toward the crest of shoulder. Less frequently, they are located in the anterior cervical triangle just below the angle of jaw as was noted in our case. Rarely, multiple lesions are seen in infancy and childhood in “Lymphangiomatosis,” the lymphatic counterpart to angiomatosis of blood vessels and potentially life-threatening disease when visceral involvement occurs.

Histological classification of lymphangiomas into capillary, cavernous or cystic subtypes based on the size of the vessels is no longer used since this distinction is of little practical importance. There is a possibility that cystic lymphangioma is merely a long-standing cavernous lymphangioma in which the cavernous spaces have been converted to cystic spaces. Bill and Sumner suggested that histologic differences are attributable to differences in anatomic location. Cystic lymphangiomas arise in areas such as neck and axilla, where loose connective tissue allows expansion of the endothelial-lined channels; cavernous lymphangiomas develop in the mouth, lips, cheek, tongue or other areas where dense connective tissue and muscles prevent expansion.

Differential diagnosis

Cavernous hemangioma

Lymphangioma can be differentiated from cavernous hemangioma by the presence of lymphoid aggregates in the stroma and more irregular lumens with widely spaced nuclei whereas cavernous hemangiomas are more common during childhood and show large dilated blood-filled vessels lined by flattened endothelial cells. Vessels may be arranged in roughly lobular arrangement or diffuse haphazard pattern.

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Conflicts of interest

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