angiomyoma and its more widespread form, which is called lymphangiomyomatosis, are characterized by a proliferation of smooth muscle in the lymphatic structures of the mediastinum, retroperitoneum and the lung. These benign lesions affect only women. The term lymphangiopericytoma has been abandoned [14].

The collective term of 'angiosarcoma' has been suggested instead of lymphangiosarcoma, as these malignant tumors not only are composed of groups of endothelium-lined empty spaces suggesting lymphatics, but also contain areas resembling hemangiosarcoma [11, 12]. They are seen exclusively in patients with long-standing lymph stasis such as is seen after radical mastectomy (postmastectomy lymphangiosarcoma) or in chronic lymphedema of the lower extremities [12]. They are discussed in Chap. 16.

17.2 Lymphangioma

17.2.1 Classification and Clinical Behavior

Histologically, three types of lymphangioma have traditionally been described based on the size of the lymphatic channels. Capillary lymphangioma or simple lymphangioma (‘lymphangioma simplex’) is composed of small, capillary-sized endothelium-lined lymphatics, whereas cavernous lymphangioma is made up of larger lymphatic channels with adventitial coats. Cystic lymphangiomas or hygromas are multilocular masses, constituted of large macroscopic lymphatic spaces that possess investitures of collagen and smooth muscle [12]. Some authors have added a fourth entity to this classification system, which is called vasculolymphatic malformation [19, 50]. Although these classification systems are widely used, there are arguments for considering the group of lymphangiomas as a single clinical entity. The distinction between cavernous and cystic lymphangioma is not always clear-cut and is often arbitrary. Moreover, cystic and cavernous components of lymphangioma often coexist in the same lesion, suggesting that cystic lymphangioma can arise out of a long-standing cavernous lymphangioma in which the cavernous...
spaces have progressively widened to form cystic spaces [1]. It has been suggested by Bill and Sumner that the classically described histological differences are the reflection of their anatomical location [3]. This means that the morphology of the lesion is dictated by the histological composition of the surrounding tissues. This theory may explain why cystic lymphangiomas most frequently arise in the neck and axilla (Figs. 17.1, 17.2), where loose connective tissue allows for the expansion of the endothelium lined lymphatic channels. Conversely, cavernous lymphangiomas are encountered in the mouth, lips, cheek, tongue or other areas where dense connective tissue and muscle prevent expansion (Fig. 17.3) [6]. In the tougher dermal and epidermal elements, expansion is further limited, resulting in the formation of a capillary lymphangioma (e.g., lymphangioma circumscriptum) (Fig. 17.4) [50].

There has been a lot of discussion about the exact etiology of these lesions, including whether they are true neoplasms or represent developmental malformations. The ‘neoplastic’ aspect of these malformations has been made responsible for the local aggressive potential of some lesions [18]. Although lymphangiomas can arise on an obstructive basis after trauma, surgery, radiation or infection, it is now widely believed by most authors that lymphangiomas are congenital lymphatic malformations that are the result of noncommunication between sequestered lymphoid tissue and the peripheral lymphatic system [4, 12, 15, 16, 31, 40, 44, 50].

Three major theories have been proposed to explain this concept [48, 50]. Failure of the primordial lymphatic sacs to drain into the veins will result in enlargement of the isolated lymphatic channels. This may result in the formation of lymphangiommas, especially the more voluminous central cystic hygromas [48]. A second theory may help to explain the characteristics of the more peripherally located lesions, such as capillary and cavernous lymphangiomas, and makes it easier to understand them. When abnormal sequestration of lymphatic tissue occurs early in embryologic life, failure to join the normally developed central lymphatic channels will result in lymphatic malformations [41, 50]. Another theory explains the branching and permeative growth pattern into the surrounding anatomical structures seen in some cavernous lymphangiomas [25]. Aberrant buds of lymphatic material lose their connections with the lymphatic primordia and may give rise to lymph-filled cysts. These cysts keep their ability to branch and grow and do so in an uncontrolled, disorderly manner.

Compared with hemangiomas, lymphatic malformations are a rare finding. There is no clear sexual predilection. About 50–65% of these lesions are found at birth, and as many as 90% may be noticed within the first 2 years of life. Cystic lymphangioma or cystic hygroma of the head, neck and axilla is the most frequently encountered and best-known entity (Fig. 17.1). A cervical lobulated, fluctuating mass in the supraclavicular fossa, the posterior triangle or the axillary region, not attached to the skin but fixed to the deep tissues of the neck is seen in these patients [8, 40]. A cervical lobulated, fluctuating mass in the supraclavicular fossa, the posterior triangle or the axillary region, not attached to the skin but fixed to the deep tissues of the neck is seen in these patients [8, 40]. The mass is usually made up of a conglomerate of cysts and sheets of tissue, extending in various directions and separating nerves, vessels and fascial planes. A clear or straw-colored serous fluid can be aspirated out of the cystic com-